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Fellows Abstracts

AWIR Association of Women in Rheumatology

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Podium Abstract Presentations

Childhood Asthma to Cardiomyopathy: Unmasking EGPA in Elderly

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Abstract

Content Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare (1 case/million) vasculitis of small-to-medium vessels, commonly presenting with asthma, allergic rhinitis, peripheral eosinophilia, and multiorgan involvement (1). Despite being usually diagnosed in middle-aged adults, it can occur in older patients, where overlapping comorbidities often complicate the diagnosis. Notably, Cardiac involvement due to EGPA independently contributes to increased mortality risk, needing high clinical suspicion for early diagnosis.

We present a case of a 75-year-old female with with history of allergic rhinitis, early onset asthma, myocardial infarction (MI) needing cardiac stenting, heart failure with reduced ejection fraction (HFrEF)), hypertension, and CKD stage 3a, admitted with four weeks of generalized weakness and worsening bilateral lower extremity edema. She was noted to have purpuric lesions on the trunk and extremities, right-hand numbness, and intermittent chest pain. Laboratory results demonstrated an elevated WBC count (26.7μL), marked eosinophilia (20.9×1μ/L), and negative troponins. Transthoracic Echocardiography confirmed a reduced LVEF 30%, diastolic dysfunction, and mild pericardial effusion. With clinical suspicion for vasculitis, a rheumatologic workup was performed, which revealed negative ANCA, weakly positive ANA titer (1:80 cytoplasmic), and elevated serum Immunoglobulin E. A skin biopsy of the lower extremity purpuric lesions revealed leukocytoclastic vasculitis with a predominant eosinophilic infiltrate, raising the suspicion for EGPA.

Subacute worsening of pre-existing ischemic cardiomyopathy from EGPA created a diagnostic challenge. EGPA typically progresses through allergic, eosinophilic, and vasculitic phases, with possible extrapulmonary manifestations such as mononeuritis multiplex, which was suspected here as well. Hypereosinophilic syndrome, a common mimicker, was also excluded after a routine bone marrow biopsy. Our patient met the 2022 ACR/EULAR criteria (score ≥6) for diagnosis of EGPA with high eosinophil counts, asthma, and biopsy-confirmed eosinophilic infiltration. Guideline-driven management recommends pulse dose steroids and cyclophosphamide in patients with cardiac involvement (2). She was managed with a prednisone taper, with marked improvement in lower extremity edema, rash, hand numbness, and eosinophilic counts.

In conclusion, Cardiac involvement is common in ANCA-negative EGPA (3), often presenting as myocarditis or coronary arteritis, which may lead to myocardial infarction and progressive heart failure(2). Early recognition is paramount, considering steroid responsiveness. This case highlights the importance of considering evaluation for EGPA in older patients with unexplained eosinophilia, atypical skin lesions, and worsening cardiomyopathy—timely intervention can significantly affect prognosis and long-term outcomes (4).

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3. https://doi.org/10.1093/rheumatology/keab514
4. https://doi.org/10.1002/ehf2.14244

Dupilumab-associated inflammatory arthritis

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Abstract

Introduction: Dupilumab, a monoclonal antibody that inhibits interleukin (IL)-4 and interleukin-13 receptor signaling, is approved for several type 2 inflammatory diseases, including asthma. It has a rare association with musculoskeletal adverse effects. We report a case of a 62-year-old female who developed inflammatory arthritis following the initiation of dupilumab therapy.

Case Presentation: A 62-year-old female with epidermolysis bullosa (EB) had been on Vyjuvek therapy since October 2023. She developed generalized pruritus, suspected to be secondary to Vyjuvek or EB, and was subsequently initiated on dupilumab. Following two injections, she developed diffuse arthralgias and myalgias. Examination revealed right wrist swelling without synovitis elsewhere. Initially had positive ANA, but subsequently had negative ANA, RF, and anti-CCP antibody. Sedimentation rate 52 mm/hr and C-reactive protein 25 mg/L. Right wrist MRI demonstrated synovial thickening and enhancement with erosions. Whole-body joint scan showed increased right wrist uptake.

Despite discontinuation of dupilumab, her musculoskeletal symptoms persisted. She was started on upadacitinib for her skin symptoms. Notably, her dermatologic and musculoskeletal symptoms improved following initiation of upadacitinib, and had no further episodes of joint swelling.

Discussion: Dupilumab is a human IgG4 monoclonal antibody binding the alpha subunit of the IL-4 receptor, blocking IL-4 and IL-13 signaling. Dupilumab has been associated with musculoskeletal adverse effects, including seronegative arthritis, arthropathy (enthesitis and enthesopathy), and joint swelling. Onset of symptoms can range from days to months after initiation of therapy. Some patients have resolution of symptoms while continuing treatment, while others require discontinuation.

Although not fully understood, the mechanism of dupilumab-associated arthritis appears to be related to inhibition of IL-4 and IL-13 pathways. IL-4 and IL-13 are important in the regulation of IL-23 in entheseal tissue. Blockade of IL-4 and IL-13 by dupilumab leads to upregulation of IL-23, which in turn promotes IL-17-driven inflammation. This pathway is implicated in the pathogenesis of spondyloarthropathies, including enthesitis and arthritis.

Conclusion: Management should be individualized based on the severity of symptoms. NSAIDs may be sufficient in mild to moderate cases, allowing for continued dupilumab therapy. In more severe or persistent cases, treatment with Janus kinase inhibitors is effective in controlling both cutaneous and musculoskeletal symptoms.

References:

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An Uncommon Manifestation of Hydroxychloroquine Toxicity: Myopathy with Concurrent Nephropathy

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Abstract

Introduction

Hydroxychloroquine (HCQ) is an antimalarial agent with immunomodulatory properties that has become a cornerstone in the management of autoimmune diseases. Although retinal toxicity is the most widely recognized adverse effect, long-term use has been associated with rare adverse effects, including myopathy and nephropathy that significantly impact patient outcomes.

Case Presentation

A 63-year-old female with positive ANCA without vasculitis and six-year history of systemic lupus erythematosus (anti-dsDNA+ by Crithidia, low C4) with musculoskeletal complaints and oral ulcers was initially treated with low-dose steroids and was weaned using hydroxychloroquine (HCQ), with symptomatic improvement. After self-discontinuing HCQ, she developed progressive muscle weakness, joint pain, and recurrent oral ulcers. Resuming HCQ provided slight relief, but symptoms worsened over time, leading to severe myalgia, weakness, and sudden kidney dysfunction.

Outpatient labs showed positive ANA, Creatinine kinase of 1767 U/L, high erythrocyte sedimentation rate and aldolase at 12 U/L. Electromyography confirmed active myositis. Despite prednisone therapy, the patient remained refractory to treatment. She was shortly after hospitalized for worsening weakness and suspected ANCA vasculitis reactivation (P-ANCA 1:640, new proteinuria, hematuria). A kidney biopsy confirmed HCQ-induced nephropathy (Figure 1).

Neurologic evaluation revealed proximal weakness, hypoactive reflexes, and mild atrophy. Abdominal fat pad biopsy ruled out amyloidosis, and thigh muscle biopsy showed chronic active myopathy with atrophy, necrosis, regeneration, and vacuolar changes.
HCQ was discontinued, and she was started on IVIG outpatient and corticosteroids (Medrol 16 mg alternating with 8 mg daily). Despite initial worsening, she gradually improved in muscle strength and renal function.

Discussion

HCQ-induced myopathy is an insidious condition that can mimic inflammatory myopathies. It is characterized by proximal muscle weakness and elevated muscle enzymes, with biopsy findings demonstrating vacuolar myopathy and curvilinear bodies. The pathogenesis remains unclear but is thought to involve lysosomal dysfunction. HCQ-induced nephropathy, although rare, has been reported in patients with long-term exposure. The mechanism involves lysosomal accumulation within renal podocytes and tubular cells, leading to proteinuria and renal impairment. However, the simultaneous occurrence of HCQ-induced myopathy and nephropathy is exceptionally rare, making this case particularly unique. Early recognition of these toxicities is crucial, as withdrawal of HCQ often results in symptom resolution. Corticosteroids and IVIG may be considered in severe cases to aid recovery.

Conclusion

This case highlights the critical importance of being vigilant for rare toxicities associated with long-term HCQ therapy. While HCQ is widely used in the management of autoimmune diseases like SLE and vasculitis, it is essential to recognize that rare but serious adverse effects, such as myopathy and nephropathy, can occur, especially when the drug is used over extended periods. In this patient's case, the concurrence of both myopathy and nephropathy, coupled with an elevated CK level and worsening renal function, raised suspicion for HCQ-induced toxicity. This made it difficult for the rheumatologists to initially recognize the full extent of her condition, as they were managing the complexities of both the lupus and the HCQ-induced toxicity.

Evaluating the Impact of Long-Term Glucocorticoid Use on Cancer Risk in Patients with Rheumatologic Diseases: A Retrospective Cohort Study

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Abstract

Background:

Long-term glucocorticoid (GC) therapy is widely used in the management of rheumatologic diseases, but its immunosuppressive effects have raised concerns about a potential increased risk of cancer. The association between long-term steroid use in patients with rheumatologic diseases and the development of cancer remains unclear. This study aims to evaluate cancer prevalence in patients with rheumatologic conditions who are on long-term steroids compared to those who are not.

Methods:

A retrospective cohort study was conducted involving 35,272 patients with rheumatologic diseases between 2011 and 2024. Patients with rheumatoid diseases were categorized into two groups: those receiving long-term GC therapy (≥30 days; n=5521) and those not on GC therapy (n= 29,751). Cancer prevalence was assessed in both groups as well as their subgroups based on sex, and age. Prevalence rates and odds ratios (OR) were calculated to determine the association between steroid use and cancer.

Results:

Cancer prevalence was significantly higher among GC users (34.2%) compared to non-GC users (20.8%). The odds ratio for cancer in GC users was 1.97 (95% CI: 1.95–2.02), indicating 97% increased odds of cancer for GC users. Stratified by sex, female GC users had a cancer prevalence of 32.6%, compared to 19.9% in non-GC users, with an odds ratio of 1.96 (95% CI: 1.48–2.59), suggesting a nearly two-fold increase in cancer risk. In male patients, cancer prevalence in GC users was 38.2%, compared to 23.4% in non-GC users, with an odds ratio of 2.02 (95% CI: 1.72–2.41), indicating a 102% increase in cancer risk. In those aged over 18 years, cancer prevalence in GC users was 34.5%, compared to 21.3% in non-GC users, with an odds ratio of 1.94 (95% CI: 1.75–2.14), suggesting a significantly higher cancer risk for those on long-term steroid therapy.

Conclusion:

Long-term steroid use in patients with rheumatologic diseases is associated with a higher prevalence of cancer. This association was observed across both sexes and age groups. The substantial cancer risk observed in patients without a personal history of cancer underscores the need for cautious GC prescribing and regular cancer screening. Further investigations should focus on additional factors, including race and comorbidities, to better understand the modulating factors behind this association and refine clinical decision-making.

Exploring the diagnosis of IgG4-related disease (IgG4-RD) in a patient with monoclonal gammopathy of undetermined significance and pancreas divisum

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Abstract

This case describes a 70-year-old female with a history of Sjogrens disease, pancreas divisum, and monoclonal gammopathy of undetermined significance, admitted for expedited evaluation of progressive pancytopenia.

Bone marrow biopsy demonstrated elevated kappa restricted plasma cells (10%) and PET-CT demonstrated a hypermetabolic pancreatic head mass, prompting biopsy which demonstrated IgG4 to IgG positive plasma cell ratio of 0.4%. Further hematopathology evaluation found that the cells were polytypic, favoring a diagnosis of IgG4 related disease despite the lack of typical histologic findings. The patient was treated with induction therapy of prednisone for fourteen days and two infusions of rituximab. Serum IgG4 levels two weeks after induction were reduced from 818 mg/dL to 501 mg/dL.

Two weeks after tapering a maintenance steroid regimen, the patient developed acute pancreatitis. After common causes of acute pancreatitis were excluded, there was raised suspicion for IgG4-RD activity, given recent steroid taper and being less than six weeks from completion of rituximab therapy. While etiology of this acute episode of pancreatitis could not be confirmed as a manifestation of IgG4- RD, improvement in symptoms after increasing steroid dosage supports this diagnosis.

IgG-4 related disease was initially described in 1961 as a case of pancreatitis with elevated serum IgG4, while later identification of extra-pancreatic lesions of similar histopathology led to the systemic disease classification. The gold-standard diagnostic criteria for autoimmune pancreatitis (AIP) include histopathologic criteria of lymphoplasmacytic infiltrate, storiform fibrosis, and obliterative phlebitis (Nambiar & Oliver, 2023).

The most accessible and evidence-based markers of disease activity in IgG-4 RD include serum IgG4, IgG2, IgE, C3, C4, IL-2R, CRP, ESR, and eosinophil levels (Iaccarino et al., 2022). While a clinical scale for disease activity does not currently exist, a tool used for research purposes called the IgG4 Responder Index accounts for symptoms of various organs, IgG4 levels, and steroid regimens in the last twenty-eight days to guide understanding of treatment response (Orozco-Galvez et al., 2023).

This case presented a diagnostic challenge due to a nuanced medical history, prompting clinicians to rely on histopathologic and serologic markers in relation to symptom presentation to guide management.

Comparative Cardiovascular Outcomes of GLP-1 Receptor Agonists versus SGLT2 Inhibitors in Patients with Type 2 Diabetes and Inflammatory Arthritis

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Abstract

Background: Both GLP-1 receptor agonists and SGLT2 inhibitors have demonstrated cardiovascular benefits in type 2 diabetes (T2DM), but their comparative cardiovascular profiles in patients with comorbid inflammatory arthritis remain unclear. We aimed to evaluate the differential cardiovascular risks between these medication classes in this population.
Methods: Using the TriNetX global federated health research network, we conducted a retrospective cohort study of adults with T2DM and inflammatory polyarthropathy who received either GLP-1 receptor agonists (n=2,838) or SGLT2 inhibitors (n=2,838) after propensity score matching. Primary outcomes included mortality, cardiac arrest, heart failure, and major adverse cardiovascular events over a 5-year follow-up period.

Results: Patients receiving SGLT2 inhibitors demonstrated significantly higher mortality compared to those on GLP-1 receptor agonists (7.7% vs 5.5%, HR 1.678, 95% CI 1.308-2.021, $p<0.001$). SGLT2 inhibitor users showed increased risk of cardiac arrest (1.4% vs 0.9%, HR 1.730, 95% CI 0.902-3.320, $p=0.095$), sepsis (12.4% vs 8.3%, HR 1.771, 95% CI 1.617-1.939, $p<0.001$), and heart failure (33.4% vs 16.3%, HR 2.462, 95% CI 2.203-2.751, $p<0.001$). The SGLT2 group also showed higher rates of atrial fibrillation (10.8% vs 6.2%, HR 1.876, 95% CI 1.728-2.036, $p<0.001$) and ventricular tachycardia (4.8% vs 2.2%, HR 2.182, 95% CI 1.808-2.595, $p<0.001$).

Conclusions: In patients with T2DM and inflammatory arthritis, treatment with GLP-1 receptor agonists was associated with significantly lower risk of cardiovascular events compared to SGLT2 inhibitors. These findings suggest GLP-1 receptor agonists may represent a preferred option for glycemic control in this specific high-risk population. Prospective randomized trials are needed to confirm these observations

Rapid Improvement of Deep Ulcerative Lesions in case of Necrotizing Vasculitis with Upadacitinib: A Case Report

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Abstract
Introduction

Janus Kinase (JAK) inhibitors are emerging as promising steroid-sparing agents in various rheumatologic diseases.[1] Their use in medium and small vessel vasculitis is being described in case reports and series. We present a 58-year-old male with leukocytoclastic vasculitis on punch biopsy, though clinical signs suggested medium vessel involvement. Upadacitinib led to rapid improvement in his deep ulcerative skin lesions.

Case Description

A 58-year-old man with type 2 diabetes mellitus presented with a diffuse papular rash and necrotic ulcerative lesions on both lower extremities (Figure 1). The rash began after a spider bite during a hunting trip. A punch biopsy showed leukocytoclastic vasculitis with fibrinoid necrosis. He was treated with doxycycline and linezolid, then started on 60 mg/day prednisone. After tapering to 10 mg, methotrexate 15 mg/week was added, but his lesions worsened within six days (Figure 2). A deeper biopsy was non-diagnostic. Due to worsening glucose control and suspicion for medium vessel vasculitis, upadacitinib was trialed due to availability. He was later hospitalized for recurrent cellulitis; during which he was treated with antibiotics and upadacitinib was held, and prednisone increased to 40 mg. Following discharge, methotrexate and upadacitinib were reintroduced and later increased due to lesion recurrence. At 30 mg of upadacitinib, methotrexate 20mg/ week and 12.5 mg prednisone, lesions significantly improved. Insurance approval was obtained for rituximab as a long-term steroid-sparing option.

Laboratory Test at presentation	Value
Rheumatoid factor	Negative
ANA	Negative
C3 and C4	Within normal limits
Cryoglobulins	Negative at 72 hours
C-ANCA and P- ANCA	Negative
MPO and PR-3 antibodies	Negative

Figure 1- Initial presentation of necrotic ulcerative lesions on bilateral lower extremities.

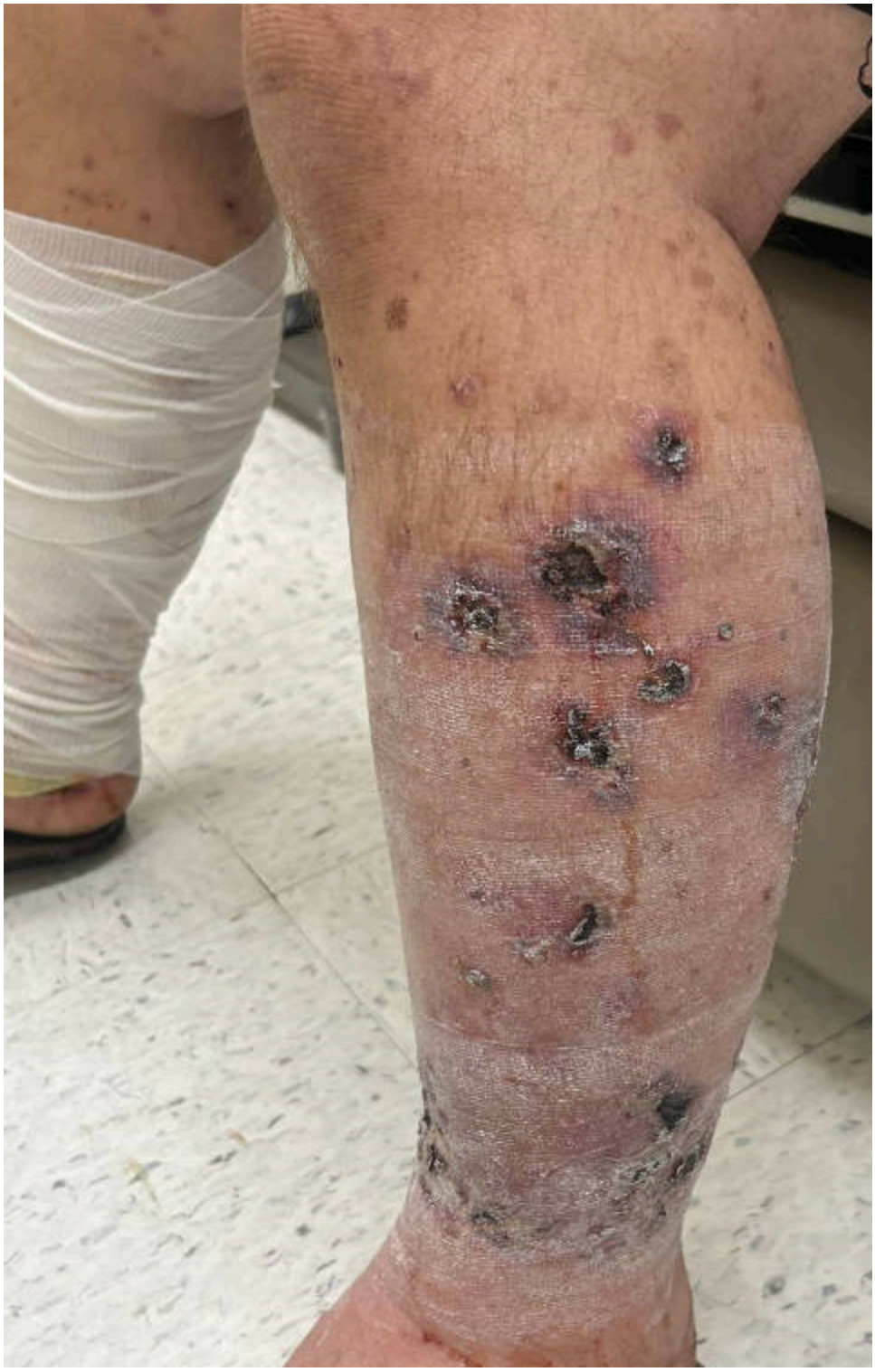


Figure 2- Worsening of ulcerative lesions on bilateral lower extremities with oozing of blood upon tapering steroids.

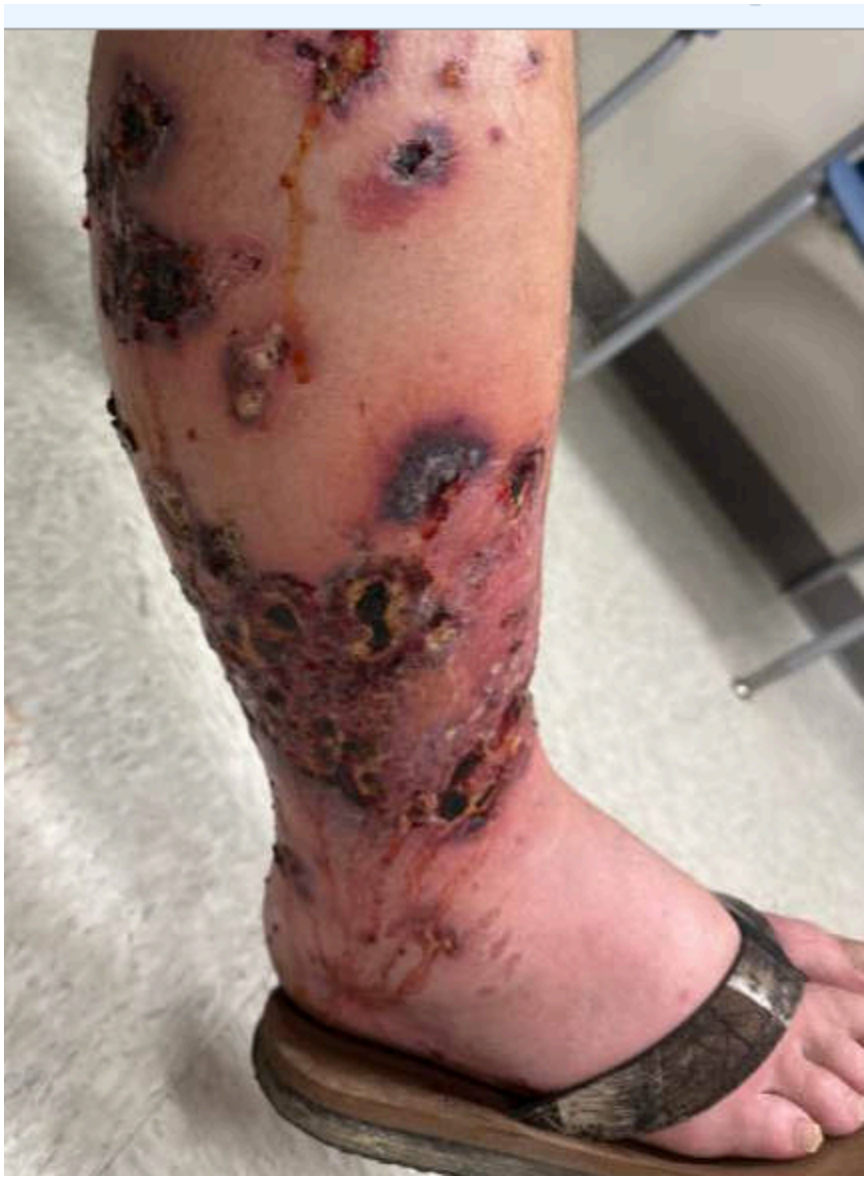
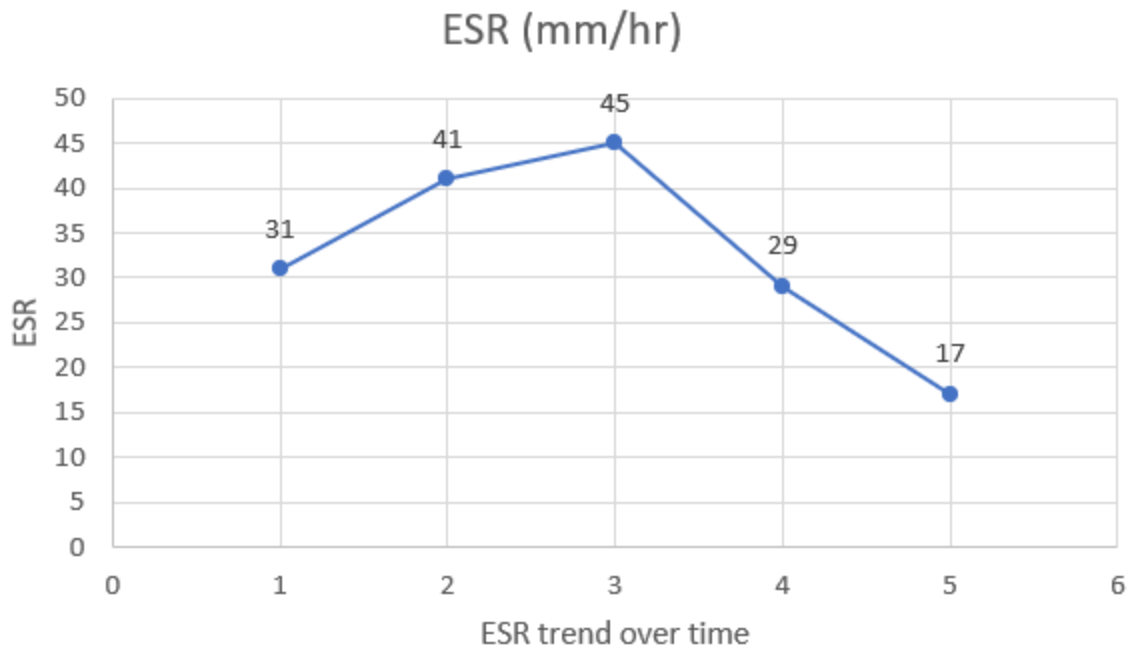


Figure 3- ESR trend over time



Discussion

Upadacitinib is a selective oral JAK-1 inhibitor approved for rheumatoid arthritis, psoriatic arthritis, and ankylosing spondylitis.[1] Its role in vasculitis is under investigation. Loricera et al reported JAK-1 inhibitor efficacy in 35 patients with giant cell arteritis.[2] One case of biopsy-proven PAN showed rapid improvement with upadacitinib after failure of multiple immunosuppressants.[3] Roy et al described four cutaneous PAN cases responding to tofacitinib.[5] Although our patient's biopsy indicated small-vessel vasculitis, clinical features suggested medium vessel involvement. His response to upadacitinib was promising. Reported side effects include infections and herpes zoster (<1%).[6] Larger trials are needed to confirm efficacy and safety in vasculitis.

Conclusion

JAK-1 inhibitors show promise as steroid-sparing agents in vasculitis. Further trials are needed to clarify their role compared to standard therapies.

Poster Abstract Presentations

“Co-occurrence of Castleman’s Disease and Systemic Lupus Erythematosus: A Rare Clinical Conundrum”

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Abstract

Introduction: Castleman disease (CD) is a rare, nonclonal lymphoproliferative disorder affecting lymph nodes and mimicking benign and malignant conditions. Systemic lupus erythematosus (SLE) is a chronic, multisystem autoimmune disease affecting various organs. CD can present with overlapping autoimmune features, complicating diagnosis and management. This case highlights a patient with idiopathic multicentric Castleman Disease (iMCD) and serologic findings suggestive of SLE without any overt clinical manifestations of lupus.

Case description: A 42-year-old male with no prior medical history presented with fatigue, a 30 lbs unintentional weight loss, dry mouth, difficulty swallowing meat, and daily mucosal bleeding for 3–4 weeks. He had 2 episodes of epistaxis, petechiae on both feet that self-resolved, and right thumb discomfort for 2 weeks. Denies fever, joint pain or swelling, Raynaud’s, dry eyes, and skin changes. No family history of autoimmune disease. Examination revealed bilateral submandibular lymphadenopathy, diffuse cervical lymphadenopathy, and spontaneous gum bleeding. Initial labs showed anemia, mild thrombocytopenia, lymphopenia, elevated inflammatory markers, and polyclonal gammopathy. Serologic tests revealed ANA of 1:2560, low C3 and C4, positive RF, dsDNA titer of 1:320, and negative SSA, SSB, RNP Ab, direct Coombs, and spherocytes. CT neck, chest, abdomen, and pelvis showed diffuse lymphadenopathy. Lymph node excisional biopsies revealed polytypic plasmacytosis and regressed germinal centers. PET scan showed hypermetabolic lymph nodes above and below the diaphragm. Bone marrow biopsy revealed hypercellular marrow with trilineage hematopoiesis, polytypic plasmacytosis, and reticulin fibrosis. Given these biopsy findings suggestive of idiopathic multicentric Castleman’s disease started on prednisone 60 mg daily. At follow-up, the patient was started on rituximab for CD and was started on hydroxychloroquine to prevent renal involvement for meeting 2019 EULAR criteria for SLE (>10 points). He showed symptomatic improvement.

Discussion: This case underscores the complexity of differentiating iMCD from autoimmune conditions like SLE due to overlapping serologic markers like ANA, ESR/CRP, dsDNA Ab. Given serologic abnormalities without overt SLE symptoms, ongoing monitoring remains essential. Additionally, Castleman’s disease can present with autoimmune-like features such as positive ANA and thrombocytopenia, further complicating differentiation. While corticosteroids benefit both conditions, a multidisciplinary approach is required to optimize management while avoiding unnecessary immunosuppression.

The Vitamin D Puzzle in Tolosa-Hunt Syndrome: A Granulomatous Insight

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Abstract

Introduction

Tolosa-Hunt Syndrome (THS) is a rare, idiopathic granulomatous inflammation of the cavernous sinus, superior orbital fissure, or orbit, presenting with painful ophthalmoplegia. It is a clinical diagnosis of exclusion, supported by neuroimaging, clinical course, and steroid responsiveness. We present a case of THS associated with elevated 1,25-dihydroxyvitamin D, raising the question of its potential diagnostic value.

Case Presentation

A 70-year-old woman with a history of Takotsubo cardiomyopathy, transient ischemic attack, and chronic headaches presented in 2020 with right-sided headache and diplopia. MRI of the orbits revealed right internal carotid artery (ICA) occlusion without signs of inflammation. ESR was elevated at 62 mm/hr. Symptoms resolved spontaneously but recurred in 2022. Repeat MRI showed new inflammation in the right cavernous sinus and orbital apex with persistent ICA occlusion. She responded well to corticosteroids. Biopsy was deferred due to proximity to vital vasculature.

Laboratory studies revealed elevated 1,25-dihydroxyvitamin D (93.2 pg/mL) with negative vasculitis and autoimmune markers (C-ANCA, P-ANCA, PR3, MPO, soluble IL-2 receptor, aquaporin-4 antibody), low ACE, and normal IgG4. CT angiography of the chest, abdomen, and pelvis showed no evidence of large-vessel vasculitis. Based on the clinical presentation, MRI findings, and steroid response, neuroimmunology diagnosed THS. The patient was treated with prednisone and mycophenolate mofetil, achieving sustained remission on follow-up MRIs through 2025.

Discussion

THS diagnosis is often delayed due to its rarity and the need to rule out multiple alternative diagnoses. In this case, the delayed appearance of inflammatory changes and steroid responsiveness supported the diagnosis. Notably, the patient had elevated 1,25-dihydroxyvitamin D—despite normal ACE and no systemic involvement—suggesting granulomatous activity, possibly via macrophage-driven extra-renal conversion of 25-hydroxyvitamin D, as seen in sarcoidosis. Although nonspecific, this marker may provide adjunctive value in select THS cases.

Conclusion

In diagnostically complex cases of painful ophthalmoplegia, where biopsy is contraindicated, neuroimaging and steroid response are key. Elevated 1,25-dihydroxyvitamin D may serve as a potential biomarker for Tolosa-Hunt Syndrome and warrants further investigation.

Progressive Pseudo-Rheumatoid Dysplasia: A Genetic Twist to a Juvenile Idiopathic Arthritis Mimic

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Abstract

Background

Progressive Pseudo Rheumatoid Dysplasia (PPRD) is a rare genetic disorder that affects the skeletal system, primarily causing progressive degeneration of articular cartilage, which leads to significant disability. It typically manifests in children between the ages of 3 to 8 years and may, therefore, be confused with juvenile idiopathic arthritis (JIA). The main clinical presentation includes swelling of the small joints of the hands, which gradually progresses to affect larger joints, leading to abnormal posture and gait abnormalities. Although imaging can help differentiate between the PPRD and JIA, a definitive diagnosis relies on genetic testing. PPRD is associated with mutations in the CCN6 gene, formerly known as Wnt1-inducible signalling pathway protein 3 (WISP3). This gene encodes a protein involved in bone and cartilage development. Accurate molecular diagnosis of PPRD is crucial to prevent unnecessary treatments directed at inflammatory arthropathies.

Cases

We present a case series of six paediatric patients diagnosed with PPRD through clinical evaluation and whole exome sequencing (WES) as shown in Table 1. All cases demonstrated hallmark features, including joint swelling, stiffness, and limited range of motion predominantly in the small joints of the hands. Radiographic findings revealed epiphyseal expansion and metaphyseal widening, consistent with PPRD. Laboratory parameters, including inflammatory markers were unremarkable in all cases, aiding differentiation from JIA.

Table 1: Characteristics of the cases

Subject	**Case 1**	**Case 2**	**Case 3**	**Case 4**	**Case 5**	**Case 6**
Gender	Male	Male	Male	Male	Female	Male
Age of onset (years)	10	5	4	4	4	6
Age at diagnosis (years)	13	6	5	7	10	8
Consanguinity	Non-consanguineous	2nd degree	2nd degree	2nd degree	2nd degree	3rd degree
Joints Involved	PIP, DIP, Elbow, Hip, Knee	PIP, DIP, Knee	PIP, DIP, Knee	PIP, DIP	Cervical spine, PIP, DIP, Wrist, Elbow, Knee, Ankle	PIP, DIP, Knee
WES Report = Mutation in the CCN6 gene						
Variant	c.1010G>A p.Cys337Tyr	c.83T>C p.Leu28Pro	c.83T>C p.Leu28Pro	c.210C>A c70Stop	c.376C>T p.Gln126Ter	c.156C>A p.Cys52Ter
Zygosity	Homozygous	Homozygous	Homozygous	Homozygous	Homozygous	Homozygous
Location	Exon 5	Exon 1	Exon 1	Not available	Exon 3	Exon 2

*PIP = Proximal interphalangeal joint, DIP = Distal interphalangeal joint
Case 2 and 3 are siblings with affected parents (unaffected heterozygous carriers)

As seen in Table 1, the variants of the mutations seen in the CCN6 gene of cases 1,4 and 6 have been reported in literature. However, the variants seen in cases 2,3 and 5 have been reported newly in our study to the best of our knowledge.

Learning points:

1. PPRD should be considered in paediatric patients presenting with progressive joint symptoms and normal inflammatory markers to avoid misdiagnosis as JIA.
2. Genetic testing by WES, is pivotal in confirming PPRD and preventing unnecessary immunosuppressive therapy.
3. Supportive management, including physiotherapy and analgesia, remains the mainstay of treatment, as curative options are unavailable.
4. Awareness of consanguinity as a potential risk factor can guide early diagnosis and genetic counselling.

A prospective cohort study to assess incidence of persistence and its predictors in epidemic post-viral arthritis

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Abstract

Background: Post viral arthritis follows a presumed or confirmed viral infection. Common causes are chikungunya, dengue, parvovirus B19, rubella, hepatitis C virus, human T-cell lymphotropic virus type 1, and human immunodeficiency virus (HIV). The confirmation of the infecting virus is not feasible in many of such cases. Short course of glucocorticoids and non-steroidal anti-inflammatory drugs (NSAIDs) are standard treatments, while the role of hydroxychloroquine (HCQ) remains debated. Retrospective studies exist for confirmed viral arthritis, but data on presumed post-viral arthritis, typically seen in epidemics, are scarce.

Objectives:

1. To assess the proportion of cases progressing to chronic arthritis beyond six weeks.
2. To compare the rate of clinical recovery between patients treated with or without HCQ.
3. To investigate potential predictors of progression to chronic arthritis, including inflammatory markers: erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and the type of virus (dengue and chikungunya)

Method: A prospective cohort study was conducted in Pune, India during a 2024 outbreak. Consecutive cases without any pre-existing arthritis were included. Data on demographics, joint involvement, inflammatory markers (ESR, CRP), rheumatoid factor (RF), anti-cyclic citrullinated peptide (Anti-CCP) antibody, viral serology and treatment details were recorded. Pain severity was assessed using a visual analogue scale (0–10). Outcomes at six weeks were classified as recovery (pain-free, off glucocorticoids) or persistence. Statistical analysis was done using SPSS v29 (χ^2 test, $p<0.05$ significant).

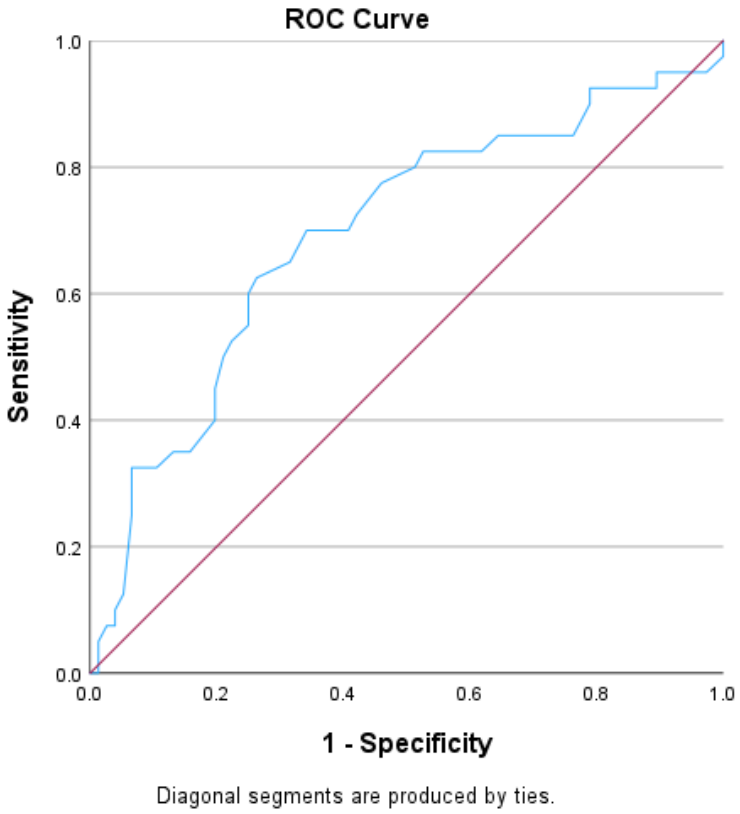
Results: Among 178 cases, viral serology (available for 129) confirmed chikungunya in 78 (68.5%) and dengue in 29 (22.5%). At six weeks, 151 cases were evaluated—98 (64.9%) recovered, while 53 (35.1%) had persistent arthritis. ESR elevation was more common in the persistence group (82.5% vs. 60.5%, p=0.016). ESR ≥31 mm at baseline predicted persistence (AUC=0.695). RF positivity was higher in persistence cases (21.9% vs. 4.4%, p=0.029). Of 151 follow-ups, 5 stopped all treatment, 11 used alternative medicine. HCQ use (39.1%) did not significantly impact recovery (HCQ: 38.3% vs. non-HCQ: 59.7%, p>0.99).
Conclusion: One-third of post-viral arthritis cases during an epidemic persisted beyond six weeks. Elevated ESR at baseline may predict an increased risk of persistence. The addition of HCQ to treatment did not significantly affect the recovery rate compared to glucocorticoids alone.

Table 1: Demographic data as per the groups

Parameter	Total (n)	Recovered	Persistent	P value
Count	151	98 (64.9%)	53 (34.1%)	

Age	151	46.27±16.75	44.04±12.32	0.40
Sex – Male/Female	151	43/55	19/34	0.388
Duration of complaints in days	151	27.24±17.20	30.45±19.73	0.301
Pain score by VAS	151	7.54±1.77	7.96±1.48	0.141
Polyarthritis	121	76 (62.80%)	45 (37.2%)	0.280
Oligoarthritis	30	22 (73.33%)	8 (26.67%)	0.280
Chikungunya positive	78	51 (65.39%)	27 (34.61%)	0.849
Dengue positive	29	21 (72.42%)	8 (27.58%)	0.51
Rheumatoid factor positive	9	2 (22.22%)	7 (77.78%)	0.029
ESR (>31mm in the 1st hour)	45	25 (55.56%)	20 (44.44%)	<0.001
Elevated CRP (>6mg/dL)	85	57 (67.06%)	28 (32.94%)	0.579
HCQ received	59	38 (64.41%)	21 (35.59%)	>0.99

Figure 1: Receiver operator curve showing the predictability of persistent arthritis using baseline ESR



Development of MOGAD in a patient with Granulomatosis with Polyangiitis and Giant Cell Arteritis Overlap Syndrome

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Abstract

Objectives

Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is a rare CNS autoimmune disorder. Characterized by antibody-mediated demyelination, a common initial manifestation is visual disturbance and vision loss due to optic neuritis. In this unique case study, we present a rare instance of MOGAD-related visual changes in a patient with an established history of both GPA (granulomatosis with polyangiitis) and GCA (giant cell arteritis) (Figure 1).

Case

A 77-year-old male with GCA with PMR and GPA (both in remission) presented in May 2024 with new-onset left eye pain with reduced vision and visual white out immediately following scheduled cataract surgery. He was seen by ophthalmology who suspected optic neuritis with MRI showing enhancement of the left optic nerve. CTA head was normal. He was initiated on methylprednisolone 1g IV for 3 days and his vision recovered to baseline by day 3 of therapy. He was continued on a course of prednisone 30mg po qd. Due to the atypical nature of his presentation, MOG and NMO antibodies were sent. MOG antibody returned as highly positive which suggested MOGAD After Rheumatology review, there was no evidence of vasculitis flare. He was continued on prednisone without relapse with a gradual taper over 3 months.

Results

Isolated case reports exist for MOGAD presentation in patients with GCA or GPA uniquely, but to our knowledge there has been no report of MOGAD in a patient with both forms of vasculitis. While no treatment guidelines exist for MOGAD, it is highly responsive to steroid therapy. While there are no Health Canada approved maintenance therapies for MOGAD, immunomodulation is recommended if there is disease recurrence or high-risk disease features. This patient is currently on methotrexate for GCA/PMR remission maintenance which has some evidence of efficacy for MOGAD and GPA. Stronger immunosuppression is limited due toa recent diagnosis of prostate cancer and previous cancer history (IPMN,? SCC).

Timeline:

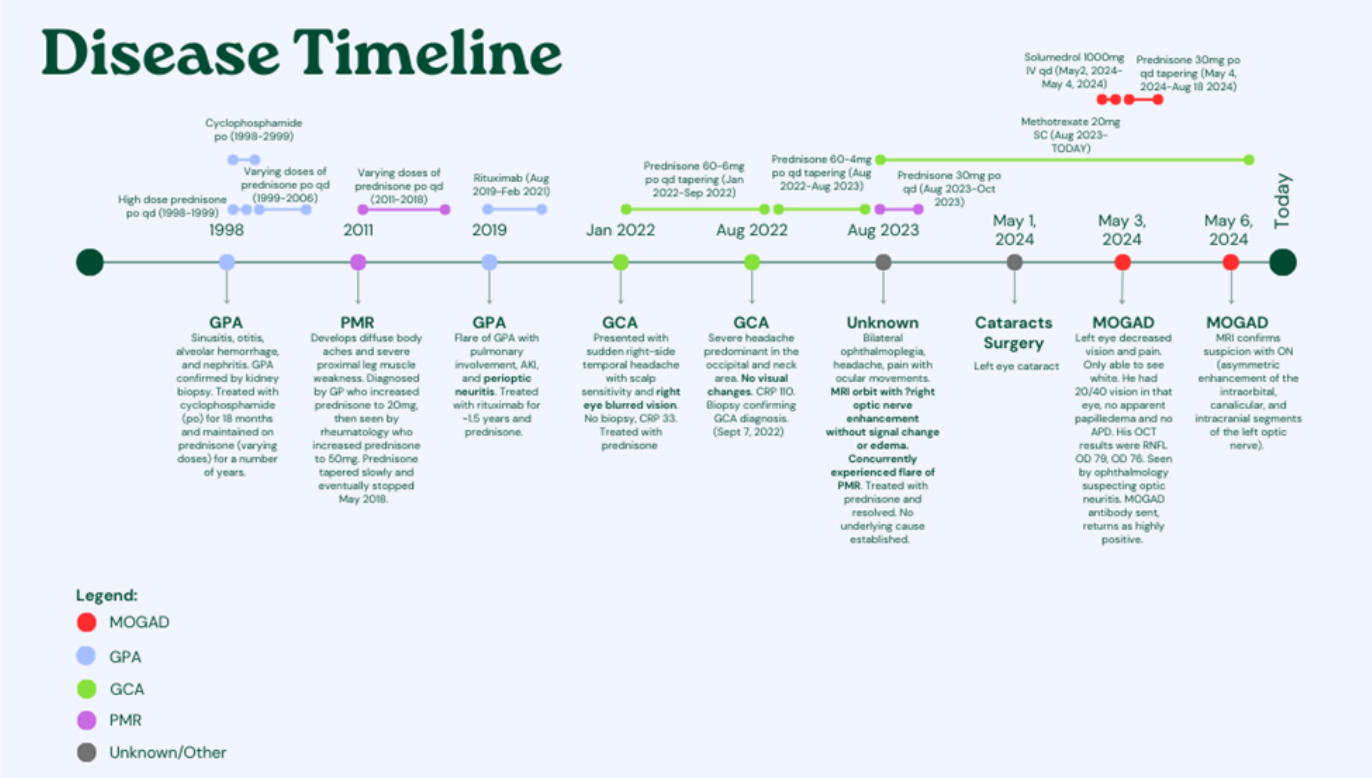

Figure 1. Disease timeline of patient with overlapping presentation of GCA, GPA, and MOGAD

Roses are red, violets are blue, eosinophils and hands can be too

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Abstract

This case describes a patient with Raynaud's phenomenon and digital gangrene as the presenting feature of Hypereosinophilic Syndrome (HES). Although reported, Raynaud's and digital ischemia in HES are rare features. Furthermore, distinguishing HES from other autoimmune disorders, particularly eosinophilic granulomatosis with polyangiitis (EGPA), can be challenging due to overlapping clinical features. This case underscores the importance of considering HES as a potential underlying cause of Raynaud's phenomenon and digital ischemia.

A 57-year-old male with history of tobacco abuse presented with new onset Raynaud's phenomenon, which progressively worsened to gangrenous digits. Examination revealed an erythematous maculopapular rash on his trunk and extremities, along with digital gangrene. Labs showed an absolute eosinophil count of 11.14 k/μL (.04-.54 k/uL), ESR of 40 mm/hr (0-30 mm/hr), CRP of 44.9 mg/L (0-10 mg/L), and IgE of 3122 IU/mL (0-200 IU/mL). Initial P-ANCA was positive (>1:160) with negative MPO/PR-3; subsequent P-ANCA, C-ANCA, MPO, and PR-3 were negative. Urinalysis and renal function were normal. Bone marrow biopsy revealed normocellular marrow with hyperplastic myeloid series and 4% eosinophils. Upper extremity angiogram revealed occlusive thrombi in the distal ulnar arteries. Cytogenetic testing was negative. Skin biopsy demonstrated sparse superficial perivascular lymphocytic inflammation with dermal eosinophils without vasculitis.

The patient was advised to stop smoking and started on sildenafil and prednisone 60 mg daily, leading to improvement in his eosinophilia. Prednisone was gradually tapered as Mepolizumab was initiated. The patient achieved sustained remission; however, he required surgical amputation of multiple digits.

Extensive evaluation confirmed the diagnosis of idiopathic HES. The patient lacked hallmark features of EGPA, such as asthma, sinusitis, rhinitis, or evidence of vasculitis, making EGPA unlikely. Endothelial injury and microvascular thrombosis secondary to eosinophils, coupled with activation of the coagulation system, have been hypothesized as the underlying cause of Raynaud's phenomenon and digital necrosis in HES. This case highlights that HES can present with significant vascular complications. Clinicians should maintain a high index of suspicion for HES, particularly in patients presenting with new-onset Raynaud's phenomenon.

Reactive Infectious Mucocutaneous Eruption (RIME) or Reason?

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Abstract
The differential diagnosis of mucocutaneous exanthems is challenging because they can range from toxic drug reactions to multisystem inflammatory syndromes. Kawasaki disease and Reactive Infectious Mucocutaneous Eruption (RIME) are both conditions that cause inflammation of the skin and mucous membranes and present with similar clinical manifestations but differ significantly in their treatments. We report a case of a viral infection triggering eruption of prominent mucositis with Kawasaki disease-like features.

An 18-year-old male with no past medical history presented with four days of fevers, sore throat, conjunctivitis, and a papular rash over the trunk and dorsum of his hands. He recently returned from Estonia a week before symptom onset. He was started on azithromycin and solumedrol, and subsequently developed oral ulcers and urethritis. Due to worsening symptoms, he was admitted to the hospital.

On physical exam, he had erosions of the upper and lower vermilion and mucosal lips, buccal mucosa, and hard palate (Figure 1a). He had ulcerations on the glans penis, conjunctival injection and scattered deep pink papules of the dorsal hands (Figure 1b).
Bloodwork showed WBC of 8.3, ESR 50, CRP 183.3. Infectious workup revealed negative blood cultures. Since Kawasaki disease criteria were met with conjunctivitis, rash, fever, and mucosal involvement, he was started on aspirin and IVIG.

Mycoplasma nasopharyngeal swab was found to be positive, steering the diagnosis towards RIME. He began a two week course of oral cyclosporine. A few weeks after discharge, he had significant improvement in his oral lesions (Figure 1c).

We report a case of acute onset mucosal predominant eruption induced by mycoplasma infection which was consistent with RIME but simultaneously fulfilled criteria for Kawasaki disease. Although both diseases have similar prodromal symptoms and mucosal involvement, they differ in rash pattern and laboratory findings. Supportive care is the treatment for RIME, and systemic treatment should be considered when the diagnosis is uncertain. Clinicians should be vigilant of RIME in cases of severe mucositis, and determine whether treatment for vasculitis is warranted.

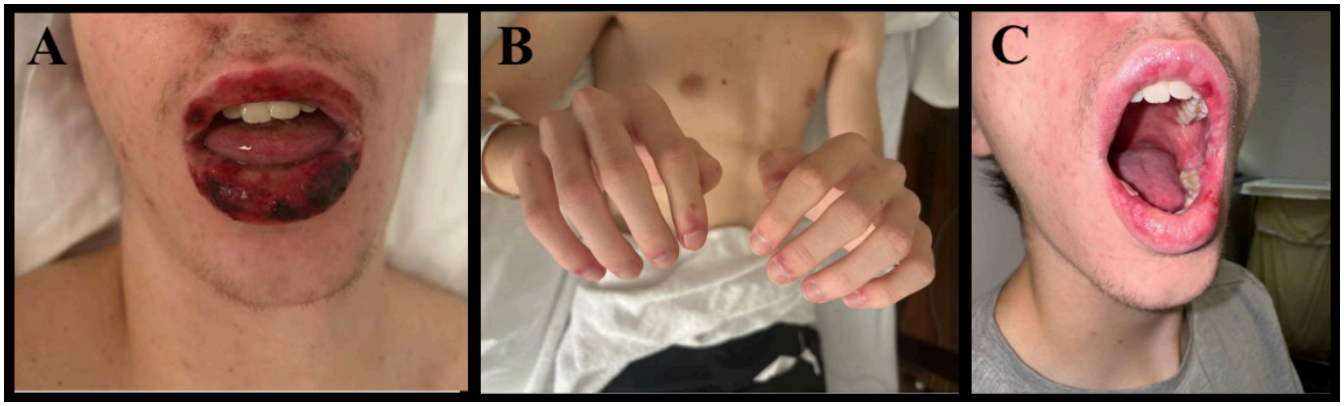

Figure 1
Clinical features of case. (A) Oral erosions at symptom onset. (B) Scattered papules of dorsal hands. (C) Resolving oral ulcerations.

CLINICAL SIGNIFICANCE OF THERAPEUTIC DRUG LEVEL MONITORING FOR MYCOPHENOLATE IN PATIENTS WITH EXTRA-RENAL SYSTEMIC LUPUS ERYTHEMATOSUS – A SYSTEMATIC REVIEW & META-ANALYSIS

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Abstract

Background/Purpose

Clinical response to Mycophenolic acid (MPA) is highly heterogeneous; thus, therapeutic drug monitoring (TDM) of MPA could help improve treatment efficacy. Our objective in this systematic review and meta-analysis was to examine therapeutic ranges for MPA levels that are associated with better outcomes and safety in patients with SLE, particularly those with extra-renal SLE manifestations.

Methods

We performed a comprehensive search of studies measuring an association between MPA levels and clinical SLE response. Using forest plots, we calculated pooled odds of clinical response by MPA levels. Additionally, we measured the weighted mean difference in MPA levels across clinical outcomes. Analysis was performed in all patients with SLE, then separately in patients with extra-renal SLE manifestations.

Results

Among 459 reviewed abstracts, 24 met inclusion. Summarized evidence supported maintaining MPA AUC0-12 levels within 30-60 mg h/L or MPA C12 within 1-2.5 mg/L; these levels were not associated with higher adverse events. Therapeutic MPA levels were associated with 12-fold higher odds (95% CI 5.44-27.35, $P < 0.0001$, $I2 = 41\%$) of overall clinical SLE response, and 15-fold higher odds (95% CI 4.74–46.89); $p < 0.0001$; $I2=61\%$) of clinical response in patients with extra-renal manifestations. Additionally, MPA levels were 32 units higher (95% CIs 17.35 45.67) in overall SLE responders, and 39 units (95% CI 15.05-62.53) higher in patients with extra-renal manifestations.

Conclusions

This study highlights the clinical utility of TDM to guide precise MPA dosing to balance efficacy vs. safety in all patients with SLE, including those with extra-renal manifestations.

Insights on Idiopathic Inflammatory Myopathies Through the Lens of Gender Differences: A Single Center Study

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Abstract

Background/Purpose: Idiopathic inflammatory myopathies (IIM) are systemic inflammatory disorders that classically affects the skin and muscles as well as other organs, including the respiratory tract, cardiac system, and gastrointestinal tract. While gender differences have been noted in other systemic autoimmune rheumatic diseases, data on gender-related variations in IIM remain limited. Our aim was to evaluate clinical differences between female and male patients with IIM.

Methods: Between April 2015 and March 2023, all patients diagnosed with IIM based on EULAR/ACR 2017 criteria were retrospectively analyzed at a single center in the United Arab Emirates. Patient characteristics and clinical data were collected for female and male patients. Statistical significance was assessed using unpaired t-test and two-tailed Fisher's test with a level of significance set at $p<0.05$.

Results: Of the 76 patients diagnosed with IIM, there were 45 female patients and 31 male patients. Females were diagnosed at a later age (45.88±14.29 vs. 40.46±13.92 years, p=0.1048) and had a longer disease duration (7±5.08 vs. 6±3.80 years, p=0.3551). Smoking history was more prevalent in males (35% vs. 9%, p=0.0072). Females had a higher frequency of hypothyroidism (29% vs. 0%, p=0.0005), while other comorbidities were comparable. In terms of diagnoses, anti-synthetase syndrome was more frequent in males (39% vs. 31%, p=0.6235), while overlap myositis was more common in females (24% vs. 19%, p=0.7805). Notably, inclusion body myositis and immune-mediated necrotizing myositis were only observed in females (4% and 2%, respectively) but not in males. Neurological manifestations were more common in females (64% vs. 39%, p=0.0359) and the prevalence of other manifestations such as dermatological manifestations (56% vs. 61%, p=0.6446) and interstitial lung disease (40% vs. 52%, p=0.3548) were similar. Myositis-specific antibodies showed no significant gender differences, except for a trend toward higher anti-MDA5 positivity in males (13% vs. 2%, p=0.152). Treatment patterns were largely comparable.

Conclusions: Our findings highlight statistically significant gender differences in smoking prevalence, association with hypothyroidism, and prevalence of neurological manifestations. Future studies with larger cohorts and longitudinal follow-up are needed to better understand the implications of these differences on disease progression and treatment response.

	Female (n = 45)	Male (n = 31)	p-value
Demographic data			
Emirati, n (%)	*26 (58)*	*22 (71)*	0.3338
Non-Emirati, n (%)	*19 (42)*	*9 (29)*	0.3338
Mean age at myositis diagnosis, n (%)	*45.88 ± 14.29*	*40.46 ± 13.92*	0.1048
Mean disease duration, years ± SD	*7 ± 5.08*	*6 ± 3.80*	0.3551
Ever smoker, n (%)	*4 (9)*	*11 (35)*	**0.0072**
Mortality, n (%)	*0 (0)*	*1 (3)*	0.4079
Comorbidities, n (%)			
Malignancy	*4 (9)*	*2 (6)*	1
Diabetes mellitus	*8 (18)*	*6 (19)*	1
Hypertension	*15 (33)*	*5 (16)*	0.1166
Hyperlipidemia	*16 (36)*	*10 (32)*	0.81
Coronary artery disease	*3 (7)*	*2 (6)*	1
Hypothyroidism	*13 (29)*	*0 (0)*	**0.0005**
Hyperthyroidism	*1 (2)*	*0 (0)*	1
Diagnosis, n (%)			
Juvenile dermatomyositis	*4 (9)*	*4 (13)*	0.7087
Anti-synthetase syndrome	*14 (31)*	*12 (39)*	0.6235
Dermatomyositis	*11 (24)*	*7 (23)*	1
Polymyositis	*2 (4)*	*2 (6)*	1
Overlap myositis	*11 (24)*	*6 (19)*	0.7805
Inclusion body myositis	*2 (4)*	*0 (0)*	0.5105
Immune-mediated necrotizing myositis	*1 (2)*	*0 (0)*	1
Disease manifestations, n (%)			
Dermatological manifestations	**25 (56)**	**19 (61)**	0.6446
Heliotrope rash	*6 (24)*	*6 (32)*	0.735
Shawl sign	*6 (24)*	*3 (16)*	0.7095
Gottron's papules	*10 (40)*	*9 (47)*	0.7609
Calcinosis	*2 (8)*	*1 (5)*	1
Mechanic's hands	*7 (28)*	*3 (16)*	0.4744
Neurological manifestations	**29 (64)**	**12 (39)**	**0.0359**
Ptosis	*1 (3.45)*	*0 (0)*	1
Dysphagia	*13 (45*	*9 (75)*	0.567
Parasthesia	*2 (7)*	*2 (17)*	0.0977
Muscle weakness	*29 (100)*	*12 (100)*	1
Cardiac manifestations	**4 (9)**	**0 (0)**	0.1407
Myocarditis	*3 (75)*	*0 (0)*	1
Pericarditis	*1 (25)*	*0 (0)*	1
Other manifestations			
Arthritis	*16 (36)*	*15 (48)*	0.3432
Interstitial lung disease	*18 (40)*	*16 (52)*	0.3548
Gastroesophageal reflux disease	*16 (36)*	*12 (39)*	0.8125
Myositis panel available, n (%)	26 (58)	18 (58)	**0.0486**
Myositis-specific antibodies, n (%)			
Anti-Jo-1	*6 (13)*	*6 (19)*	0.5328
Anti-OJ	*0 (0)*	*1 (3)*	0.4079
Anti-P155/140	*1 (2)*	*0 (0)*	1
Anti-PL7	*2 (4)*	*1 (3)*	1
Anti-PL12	*5 (11)*	*2 (6)*	0.6937
Anti-Mi-2	*2 (4)*	*1 (3)*	1
Anti-TIF1γ	*2 (4)*	*0 (0)*	0.5105
Anti-NXP2	*1 (2)*	*0 (0)*	1
Anti-MDA5	*1 (2)*	*4 (13)*	0.152
Anti-SRP	*0 (0)*	*0 (0)*	1
Anti-cN1A	*0 (0)*	*3 (10)*	0.0639
Myositis-associated antibodies, n (%)			
Anti-PMScl	*3 (7)*	*2 (6)*	1
Anti-U1-snRNP	*0 (0)*	*1 (3)*	0.4079
Anti-Ku	*2 (4)*	*1 (3)*	1
ANA	*19 (42)*	*13 (42)*	1
Anti-RNP	*3 (7)*	*2 (6)*	1
Anti-sm/RNP	*2 (4)*	*3 (10)*	0.6438
Anti-centromere	*0 (0)*	*0 (0)*	1
Anti-SCL70	*0 (0)*	*0 (0)*	1
Anti-SSA (Ro)	*13 (29)*	*14 (45)*	0.2224
Anti-SSB (La)	*0 (0)*	*1 (3)*	0.4079
Laboratory investigations, mean ± SD			
Creatine kinase (CK)	1483 ± 3307	1631 ± 3365	0.8495
Aldolase	0.47 ± 1.30	0.22 ± 0.22	0.2934
C-Reactive protein (CRP)	16.44 ± 21.40	18.95 ± 34.94	0.699
Erythrocyte sedementation rate (ESR)	54.85 ± 35.42	43.72 ± 36.25	0.1865
Muscle Biopsy, n (%)	11 (24)	5 (16)	0.568
Myopathic changes on biopsy, n (%)	*8 (73)*	*2 (40)*	0.1854
Medications, n (%)			
Corticosteroids	*35 (78)*	*22 (71)*	0.5929
Azathioprine	*12 (27)*	*6 (19)*	0.5861
Methotrexate	*12 (27)*	*8 (26)*	1
Cyclosporin	*2 (4)*	*3 (10)*	0.3928
Hydroxychloroquine (HCQ)	*8 (18)*	*6 (10)*	1
Mycophenolate mofetil (MMF)	*16 (36)*	*6 (19)*	0.4766
Tacrolimus	*1 (2)*	*2 (6)*	0.5634
Rituximab	*14 (31)*	*11 (35)*	0.8048
Tofacitinib	*3 (7)*	*1 (3)*	0.6412
Baricitinib	*1 (2)*	*0 (0)*	1
Upacitinib	*1 (2)*	*0 (0)*	1
Intravenous immunoglobulins (IVIG)	*10 (22)*	*8 (26)*	0.787
Plasma exchange	*1 (2)*	*1 (3)*	1

Table 1. Demographic data, clinical characteristics and treatment of patients with myositis. P-values comparing between female and male patients in the registry.

CAR-T cell Therapy Inducing Rheumatoid Arthritis Remission - A Case Report

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Abstract
CAR-T cell Therapy Inducing Rheumatoid Arthritis Remission - A Case Report
Guru Prasad Parthiban MD., Tanmayee Bichile MD.

A 56-year-old Caucasian female was diagnosed with Rheumatoid arthritis (RA) in 2007 based on inflammatory polyarthritis, rheumatoid factor of 68 (<20 IU/ml), anti-cyclic citrullinated peptide (anti-CCP) level of 294 (< 17 U/ml). Treatment included prednisone followed by methotrexate 15 mg oral weekly and folic acid 1 mg daily. She did not tolerate adalimumab and failed golimumab. Subsequently, etanercept 50 mg SC weekly was added in 2013. Despite etanercept and methotrexate combination from 2016 to 2020, RA was active with flares requiring NSAIDs, oral and intramuscular Glucocorticoids.

In 2021, she was diagnosed with Diffuse Large B-cell Lymphoma (DLBCL), treated with 6 cycles of Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone (CHOP) chemotherapy. In 2022, she received lymphodepleting therapy and Chimeric Antigen Receptor -T cell therapy (CAR-T). Both etanercept and methotrexate were stopped in 2021. Lymphoma has been in remission as per the last PET/CT in 2024.

At the rheumatology follow-up in 2024, RA was in clinical remission with Clinical Disease Activity Index (CDAI) score of 0, despite discontinuation of all RA medications. A decision was made to monitor the patient off RA treatments.

In one case series, 3 difficult to treat RA patients were successfully treated with CAR-T cell therapy with a significant reduction in their CDAI scores (23 to 1.5 points in patient 1, 18 to 2 points in patient 2, and 28 to 2 points in patient 3) at 6-month follow up. RF disappeared and anti-CCP levels had significantly reduced in all 3 patients (1). Similar improvement in RA activity (clinical and serologic) is reported in 2 other patients who received CAR-T therapy (2,3).

Our case emphasizes the efficacy of CAR-T therapy in RA with sustained remission at 2 years highlighting the need for further studies to assess the effectiveness and safety of this intervention in RA.

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A Rare Presentation of Drug Induced Lupus

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Abstract

Cryptococcoid Sweet Syndrome is a rare, newly discovered variant of Sweet syndrome that is characterized by yeast-like structures on histopathologic analysis. Sweet Syndrome, also known as febrile neutrophilic dermatosis, is an inflammatory skin condition that results in skin lesions (tender plaques or nodules) with associated systemic symptoms, ex. fever, arthralgias, ocular inflammation. In some cases, Sweet Syndrome can be the presenting finding for drug-induced lupus, although also relatively rare. We present a case of a 67-year-old female with a past medical history of hypertension, diabetes, end stage renal disease (ESRD) on peritoneal dialysis (PD) presenting to the emergency department with acute pulmonary embolism, exudative conjunctivitis, oral ulcerations, and new onset skin lesions involving the face and elbows. Of note; she had been on hydralazine for two years prior to presentation. Upon biopsy of one of the skin lesions; findings were remarkable for marked papillary dermal edema resulting in a subepidermal blister, with diffuse neutrophilic inflammation associated with pseudo-cryptococcoid structures, leukocytoclasis and focal necrosis, consistent with cryptococcoid Sweet Syndrome. Lab work was also notable for high titer positive antinuclear antibody (ANA), positive double stranded DNA (dsDNA) antibody, elevated anti-MPO titer, low C3, and elevated anti-histone antibody. The clinical picture raised concern for hydralazine-induced lupus erythematosus. The patient was treated with high-dose systemic glucocorticoids (1 mg/kg prednisone) with a taper and transitioned to hydroxychloroquine 300 mg daily with eventual healing of her skin lesions. Given the rarity of documented cases of hydralazine-induced cryptococcoid sweet syndrome associated with drug-induced lupus, our goal is to add to the existing literature this rare variant of the disease.

Non-arteritic anterior ischemic optic neuropathy (NAION) associated with semaglutide use mimicking giant cell arteritis (GCA): A diagnostic challenge

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Abstract

Introduction:
Non-arteritic anterior ischemic optic neuropathy (NAION) and giant cell arteritis (GCA) both cause acute painless monocular vision loss in adults older than 50, making their diagnosis challenging. We present a case of NAION mimicking GCA.

Case presentation:
A 59-year-old male with history of type 2 diabetes mellitus on semaglutide and hyperlipidemia presented with sudden onset painless left visual loss. Outpatient work up showed elevated ESR (48 mm/hr), normal CRP, and MRI brain/orbit with enhancement of left optic disc. Ophthalmology evaluation reported left optic disc edema so prednisone 60 mg was started. At our facility, inflammatory markers were normal and temporal artery ultrasound showed no halo sign. Eye exam revealed bilateral optic disc edema and pallor with superior-temporal visual field defect. Imaging and lumbar puncture showed no evidence of demyelinating neuropathy. The patient had no systemic symptoms and scored 3 points on the ACR/EULAR criteria. He was thought to have NAION and was discharged on prednisone taper, however he returned 3 days later with persistent vision loss, frontotemporal headache and tenderness. Given high suspicion of GCA, IV methylprednisolone 1 gm daily was administered for 3 days followed by prednisone 1 mg/kg/day. Repeat inflammatory markers and temporal artery ultrasound were normal. Infectious work up was negative for Lyme, HSV, VZV, CMV, TB, HCV, HBV, and syphilis. ANCA panel, ANA, MOG, NMO, and antiphospholipid antibodies were negative. IgG4 level was normal. Given persistent symptoms and optic disc edema, temporal artery biopsy was pursued that showed no evidence of arteritis. PET scan revealed no metabolic evidence of large vessel vasculitis. Extensive work up ruled out a diagnosis of GCA and the patient's vision loss was attributed to NAION exacerbated by semaglutide use. Visual Evoked Potential was severely reduced in the left eye with normal latency further confirming a diagnosis of NAION.

Conclusion:
Eye exams are crucial to distinguish NAION from GCA with evidence of disc hyperemia in NAION and pale optic disc in GCA. Our case demonstrated an atypical NAION presentation mimicking GCA resulting in unnecessary steroid use. Comprehensive multidisciplinary evaluation is required for definitive diagnosis with increased awareness of risk of NAION with semaglutide use.

Clinicians' Perspectives on Gout: Confidence, Knowledge, and Management Practices

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Abstract

Gout, the most prevalent inflammatory arthritis in adults, results from monosodium urate crystal deposits in joints and tissues, causing significant joint swelling and pain. If inadequately treated, it can lead to tophi and affect other organs. Given its widespread occurrence, gout is managed by various healthcare providers across different clinical settings. This study explores clinicians' perceptions of gout, particularly focusing on their comfort with diagnosing, treating both acute and chronic gout, and making specialist referrals.

A survey was conducted among clinicians from rheumatology, nephrology, podiatry, primary care, hospitalists, and emergency medicine across the United States using REDCap, with IRB approval from Wake Forest. The 26-question online survey targeted physicians and advanced practitioners involved in gout management, assessing confidence, awareness, comfort, and satisfaction on a 10-point scale. Multiple-choice questions explored the impact of comorbidities on treatment plans and other aspects of gout care.

Among 155 participants, 146 treated gout with varying frequency. While 41.3% felt extremely comfortable diagnosing gout, awareness of classification criteria was notably lower, with 22.4% reporting no awareness. Confidence in managing acute gout flares was high (43% scoring a 10), but comfort with prescribing uric acid-lowering therapy (ULT) varied, with 34.5% feeling extremely confident and 11% uncomfortable. Satisfaction with treating acute and chronic gout was low, reported by 14.2% and 6.4% respectively. Comorbidities, particularly chronic kidney disease, obesity, and cardiovascular disease, significantly influenced treatment decisions. Allopurinol was the most prescribed ULT (98.6%), followed by febuxostat and probenecid. For acute episodes, NSAIDs, colchicine, and oral prednisone were predominantly used. Regarding ULT, 73.3% continued therapy once gout was controlled, while 10.7% reduced the dose, and 2% discontinued it. 48.9% referred patients to specialists regularly.

This study reveals gaps in clinicians' knowledge about gout classification and treatment, especially with comorbidities. While most continue ULT once gout is controlled, some reduce or stop therapy, indicating a need for clearer long-term management guidelines. The variability in specialist referral practices suggests an opportunity to standardize criteria for consistent care. Addressing these variations could improve treatment consistency and patient outcomes. Future analyses will assess differences in perceptions across subspecialty groups.

Chronic calcium pyrophosphate arthritis presenting with reducible ulnar drift mimicking seronegative rheumatoid arthritis

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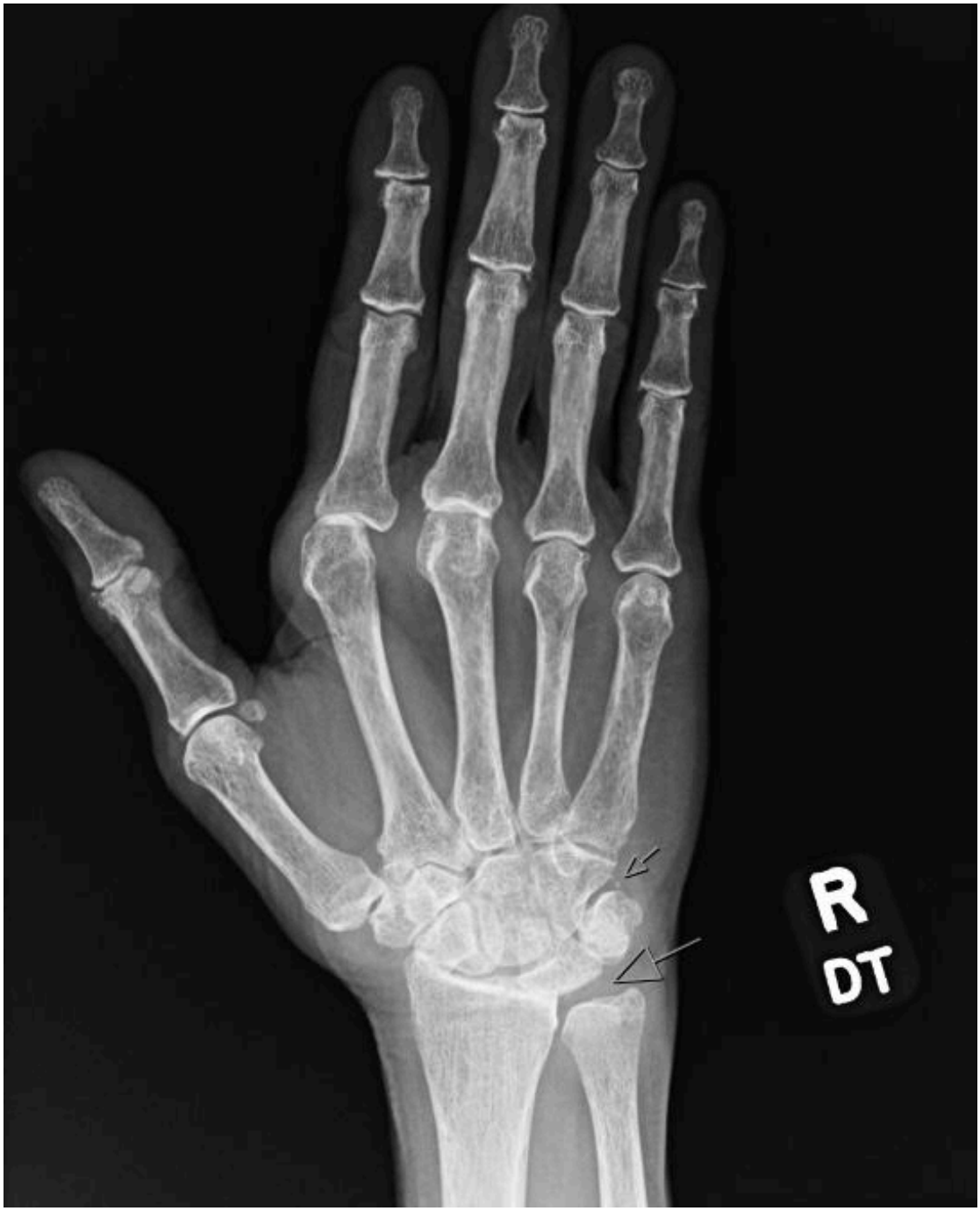
Abstract

Introduction

Chronic calcium pyrophosphate (CPP) arthritis is characterized by accumulation of calcium pyrophosphate dihydrate (CPP) crystal deposits in articular tissues, resulting in inflammatory arthritis. It is also known as pseudo-rheumatoid arthritis (pseudo-RA) because its clinical presentation can closely resemble that of rheumatoid arthritis, often leading to misdiagnosis and inappropriate management. We report the case of a 79-year-old female who presented with pseudo-RA.

Case presentation

A 79-year-old female with past medical history of osteoarthritis, spinal stenosis and right rotator cuff tear, was referred to rheumatology clinic for evaluation of joint pain. She was told by her PCP that her hands look like she has rheumatoid arthritis. She had bilateral hand pain for many years, aggravated with knitting, relieved with meloxicam, morning stiffness for 1.5 hrs. Recent onset tingling in right hand and right foot numbness also present. Physical examination revealed bilateral reducible ulnar drift, prominence of MCPs but no appreciable synovitis. Laboratory studies showed negative RF, ANA and ACPA. X-Ray bilateral hands showed bilateral scapholunate dislocation. Right 2nd and 3rd distal interphalangeal osteoarthritis. Soft tissue mineralization of right triangular fibrocartilage complex. Left 2nd, right 2nd, and 3rd metacarpophalangeal joint space narrowing. Questionable left 2nd metacarpophalangeal periarticular mineralization. She was diagnosed with chronic CPP arthritis.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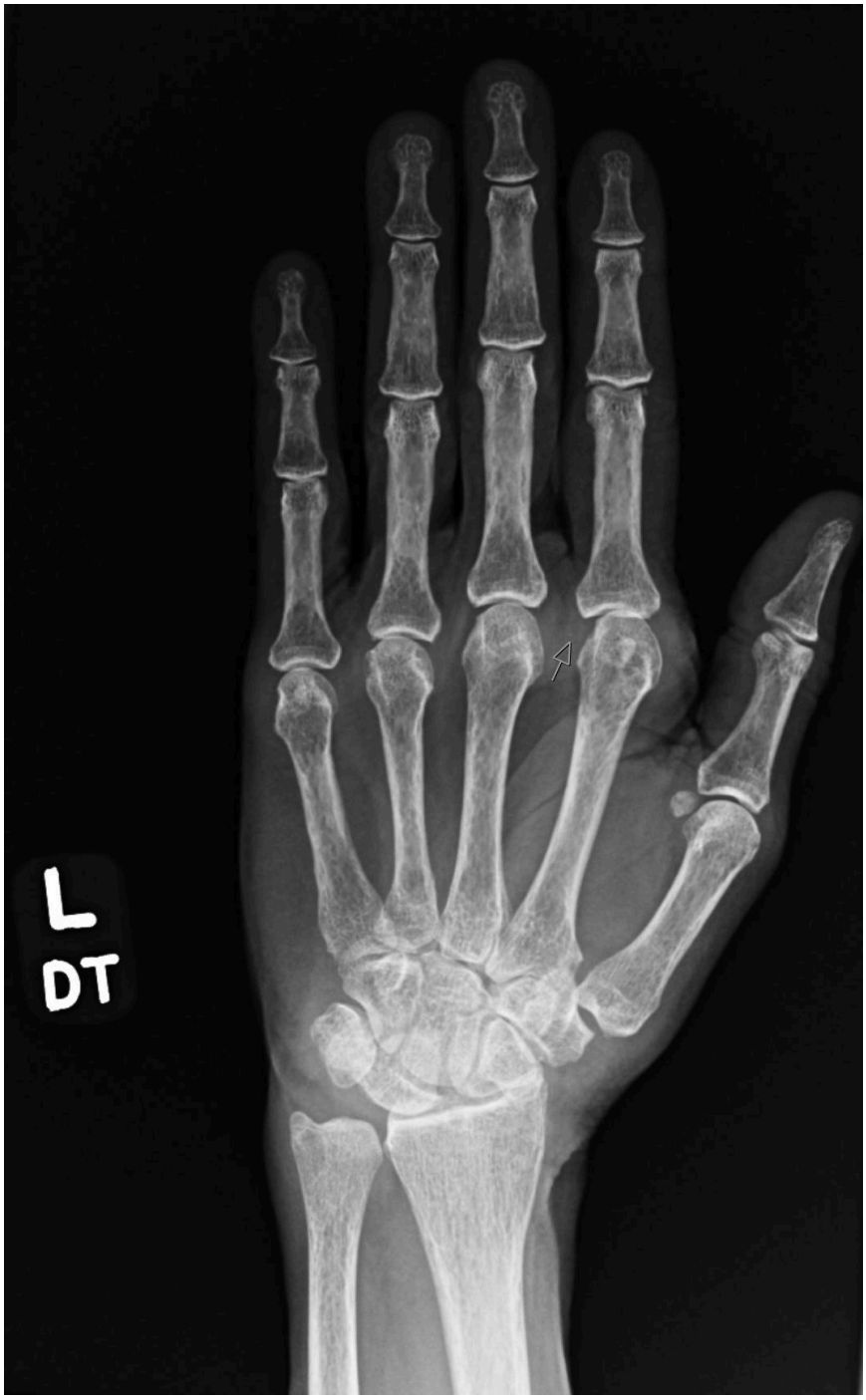

Discussion

Chronic CPP arthritis is frequently misdiagnosed as seronegative rheumatoid arthritis. Diagnostic challenges increase when patients are in early stages, as radiographic findings may be absent. Unlike our case, early CPP arthritis may not reveal chondrocalcinosis on imaging. More typical radiographic findings include joint space narrowing in second and third MCP joints and chondrocalcinosis of triangular fibrocartilage complex in the wrist. These observations underscore the limited sensitivity of plain radiographs in detecting CPP arthritis. Similarly,

synovial fluid analysis may also yield negative results, further complicating the diagnosis. In such cases, musculoskeletal ultrasound can serve as a valuable tool. While ulnar drift is a rare manifestation in CPP arthritis, recognizing this deformity is crucial to avoid misdiagnosis.

Conclusion

Physicians should maintain a high index of suspicion for chronic CPP arthritis to avoid misdiagnosing it as rheumatoid arthritis and thereby prevent unnecessary treatment with DMARDs.

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A Rare Glimpse: An Atypical Case of Ocular Sarcoidosis

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Abstract

OBJECTIVES

To describe a rare presentation of ocular sarcoidosis, emphasizing the diagnostic approach, clinical features, and the role of timely intervention in preventing long-term ocular complications.

METHODS

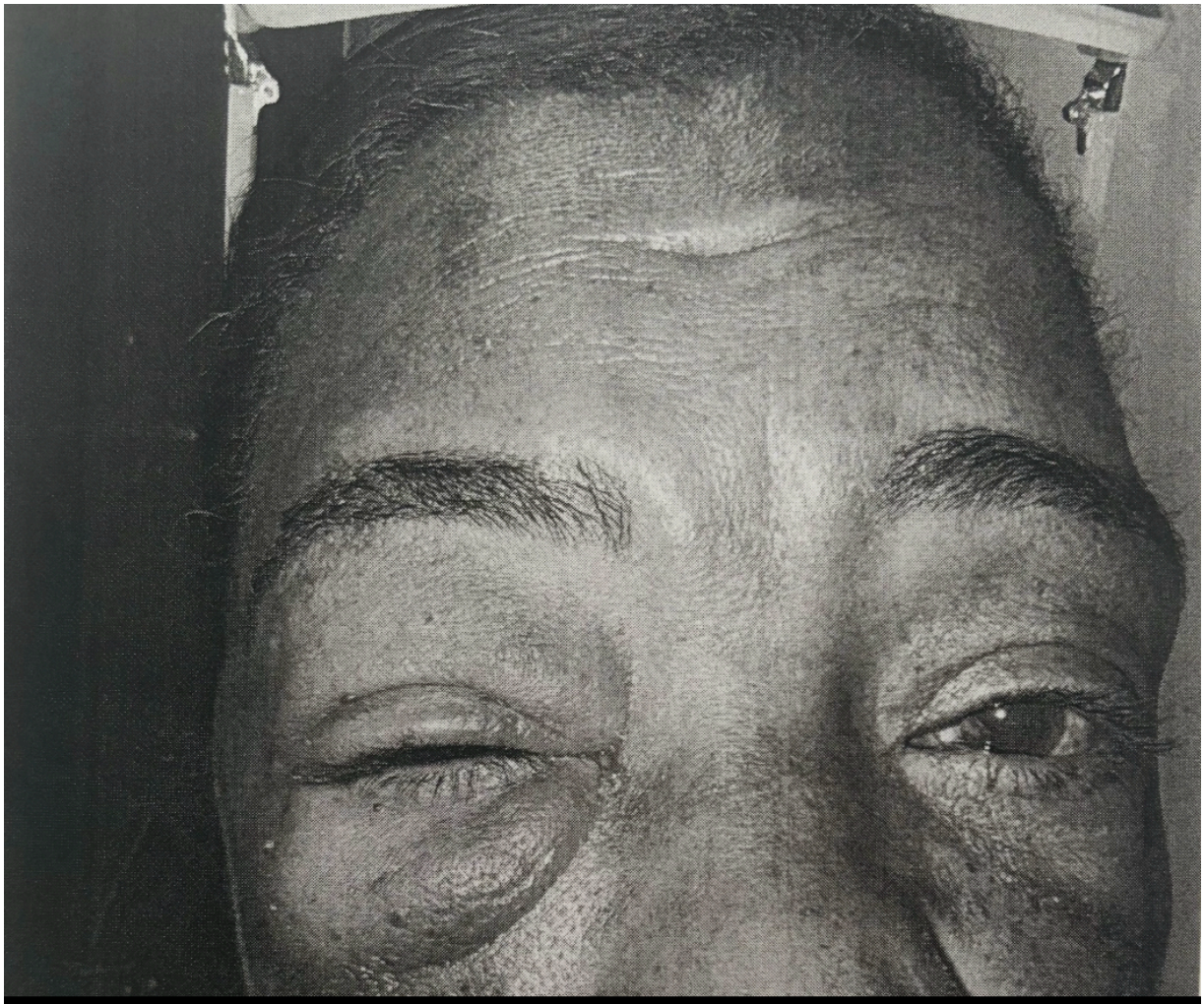
A comprehensive literature search was conducted through PubMed/Medline, BioMed Central, British Medical Journal, the American College of Rheumatology, and the American Academy of Ophthalmology up to March 2025. Case reports describing ocular sarcoidosis were included, and data on diagnostic methods, clinical presentation, systemic involvement, treatment strategies, and patient outcomes were analyzed.

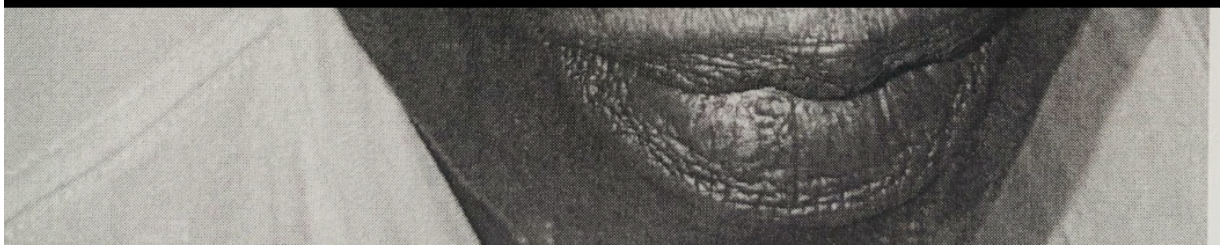
RESULTS

Ocular sarcoidosis remains a rare but significant manifestation of systemic sarcoidosis, occasionally presenting without pulmonary involvement. Uveitis was identified as the most common ocular feature, often preceding systemic diagnosis. Diagnostic confirmation relies on clinical signs, laboratory markers, and histopathological evidence of noncaseating granulomas. The IWOS criteria provide a structured diagnostic framework, though limitations exist in adnexal presentations. Corticosteroids remain first-line therapy, with immunomodulators reserved for refractory or steroid-intolerant cases.

CONCLUSION

Orbital sarcoidosis, while a rare and often underdiagnosed manifestation of systemic sarcoidosis, presents a complex challenge, as its clinical features can easily overlap with other inflammatory and infectious conditions. Timely and accurate diagnosis remains crucial, requiring a multidisciplinary approach that integrates rheumatologists, ophthalmologists, and pathologists for comprehensive evaluation. A personalized, multidisciplinary approach is critical for managing ocular sarcoidosis effectively and preventing long-term complications, with the ultimate goal of preserving vision and improving the patient's quality of life.





A Case Conundrum: Lupus Nephritis presenting with Severe Multiple Thrombosis

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Abstract
A Case Conundrum: Lupus Nephritis presenting with Severe Multiple Thrombosis
Background:
Lupus Nephritis (LN) is a common manifestation that occurs in around 40% of those with systemic lupus erythematosus. We present a case of LN that presented as extensive renal vein and inferior vena cava (IVC) thrombosis that complicated the confirmation of the diagnosis via renal biopsy.
Case:
Previously healthy 22 year old female presented with dyspnea and bilateral lower extremity 3+ pitting edema up to her thighs. Imaging demonstrated bilateral renal vein thrombosis with extension into the IVC, anasarca, and bilateral moderate pleural effusions. While an autoimmune cause was suspected and a renal biopsy was desired, the patient needed therapeutic anticoagulation and urgent suction thrombectomy. Initial workup confirmed nephrotic range proteinuria and decreased complement levels but a negative ANA screen, dsDNA, and ANCA. Hematologic studies were INR 0.95, PT 10.4, PTT 34.4, elevated Kaolin activated clotting time 239, negative cardiolipin and beta 2 glycoprotein antibodies. With suspicions still high, repeated studies returned with elevated ANA 1:1280 nuclear, speckled pattern, positive Smith, Smith/RNP, and Chromatin antibodies. Regarding renal biopsy, discussions were had given apprehension of suspending anticoagulation long enough post biopsy to reduce risk of bleeding in the setting of the active thrombosis. The decision was made to start high dose steroids and transfer to a higher level of care with inpatient rheumatology where the renal biopsy was performed showing WHO Class V membranous glomerulopathy.
Discussion:
Ultimately, the confirmatory renal biopsy was delayed due to the balancing act needed with treating her extensive thrombosis and need for anticoagulation. It is recommended to hold anticoagulation perioperatively for renal biopsy due to its increased bleeding risk with 11% of cases developing hematoma and 1.6% requiring blood transfusion. Her risk of thrombosis was also concurrently elevated with her hypoalbuminemia, worsening renal function, increased inflammatory state, and nephrotic syndrome which independently has a 25% incidence of thromboembolic events. Risks and benefits must be weighed since renal biopsy is still needed to guide treatment as quickly as possible to reduce needs for dialysis or transplant.
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Polymyalgia Rheumatica Secondary to Immune Checkpoint Inhibitor: A Case Report

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Abstract

Introduction

Polymyalgia rheumatica (PMR) is a rare immune-related adverse event associated with immune checkpoint inhibitors (ICIs), such as pembrolizumab. While effective in enhancing immune response against malignancies, ICIs may trigger autoimmune complications, including PMR-like symptoms. Recognizing these manifestations is critical for timely management and ensuring the continuation of cancer therapy when possible.

Case Presentation

An 83-year-old male with a history of malignant melanoma treated with pembrolizumab presented with bilateral shoulder pain, morning stiffness lasting over an hour, difficulty getting up from the chair without the use of his arms and significant difficulty lifting his arms above his head. Laboratory findings included elevated inflammatory markers and positive ANA, with no clinical features of giant cell arteritis or rheumatoid arthritis. Patient was started on low-dose prednisone (15mg/day) with a dramatic clinical improvement within one week. PMR secondary to pembrolizumab was diagnosed. Prednisone was subsequently tapered down to 10mg/day. Follow-up demonstrated sustained remission on a tapered steroid regimen of 1mg every 2 weeks

Discussion

PMR secondary to ICIs is uncommon but increasingly recognized as ICIs are more widely used, especially in treatment of malignancies. Symptoms can mimic classic PMR, but the temporal association with ICI therapy and exclusion of other inflammatory or autoimmune conditions are key to diagnosis. Prompt initiation of corticosteroids typically leads to symptom resolution and may allow for the continuation of ICI therapy in non-severe cases.

Conclusion

PMR should be considered in the differential diagnosis of new-onset musculoskeletal symptoms in patients receiving ICIs. Early identification and management with corticosteroids can significantly improve outcomes while preserving oncologic treatment goals.

Systemic Lupus Erythematous and Antiphospholipid Syndrome Synergy: Aortic Dissection

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Abstract

Introduction:
Systemic lupus erythematosus (SLE) is an autoimmune rheumatological disease that increases chronic inflammation, secondary hypertension, and immune complex deposition. Of these patients, 10-20% of them eventually develop antiphospholipid syndrome (APS), a disease of hypercoagulability in the setting of antiphospholipid antibodies. These two diseases in tandem can increase vascular complications significantly including aortic aneurysms and aortic dissections.

Case Presentation:
We present a 29 year female with a past medical history of SLE (on Azathioprine and Prednisone), focal segmental glomerulosclerosis (FSGS), APS on Coumadin, history of cerebrovascular accident, hypertension, history of abdominal aortic aneurysm with repair as well as history of type B aortic dissection that had presented for chest pain. CT angiogram of the chest showed a type B aortic dissection that had progressed to type A dissection that involved both the ascending aorta and aortic root, for which she had a Bentall procedure.

Importance:
SLE and APS are both well known and documented, but in conjunction with aortic dissection it is very rare. Literature research shows there are few patient cases with this triad. About 10-20% of patients with lupus end up developing APS. However, less than 40% of these patients have thrombosis. Each of these diseases individually increase the risk of a patient developing an aortic aneurysm and subsequent dissection. SLE and APS can have a synergistic effect leading to chronic inflammation and thrombosis secondary to APS. The result is vascular damage like aortic aneurysm and eventual aortic dissection. The accelerated progression of our patient from AAA to type B aortic dissection and eventual type A dissection is likely due to medical noncompliance and these two conditions working hand in hand. Lastly, while this triad of pathologies is novel in and of itself, this case is also interesting because the patient is of a younger demographic.

A RARE CASE OF PSEUDO-PSEUDO MEIGS SYNDROME WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Abstract
Background:

Pseudo-pseudo Meigs syndrome (PPMS), also known as Tjalma Syndrome, is a rare manifestation of systemic lupus erythematosus, marked by pleural effusions, ascites, and elevated CA-125 in the absence of ovarian tumors. We present a case of a 26-year-old female who's initial SLE presentation, including lupus nephritis, featured progressive ascites and other atypical findings.
Case Presentation:

A 26-year-old woman with a history of idiopathic thrombocytopenic purpura presented with two months of progressive abdominal distension and nausea. The exam revealed decreased breath sounds at the lung bases and massive ascites. Labs showed normocytic anemia, mild hyponatremia, elevated creatinine, hypoalbuminemia, elevated ESR, CRP, and CA-125 (83 U/mL). LFTs, beta-HCG, and hepatitis panel were unremarkable. Paracentesis drained 2 liters of clear fluid with low SAAG and negative cytology. Imaging showed moderate ascites, mild bilateral pleural effusions, and no pelvic masses. Urinalysis revealed 3+ proteinuria and microscopic hematuria, prompting further immunologic evaluation.
Autoimmune workup showed elevated Anti-dsDNA, Anti-Smith, Anti-SSA, Anti-RNP antibodies, and low C3/C4 levels. ANA, ANCA, Anti-CCP, and APL antibodies were negative. Renal biopsy confirmed class V lupus nephritis. The patient was treated with IV steroids and discharged on mycophenolate mofetil, hydroxychloroquine, and oral steroids, with significant improvement.
Discussion:

PPMS is a rare SLE complication often misinterpreted as gynecologic malignancy due to ascites and raised CA-125. Unlike classic Meigs syndrome, PPMS lacks an ovarian tumor. Misdiagnosis may lead to unnecessary investigations or surgeries. In this case, thorough immunologic testing and imaging ruled out malignancy and led to the correct diagnosis of SLE with PPMS. Treatment is directed at managing SLE itself, with no need for special interventions for PPMS.
Conclusion:

This case underscores the importance of considering SLE in patients with unexplained ascites, pleural effusions, and elevated CA-125. Early recognition prevents misdiagnosis and unnecessary interventions, enabling prompt and effective treatment.
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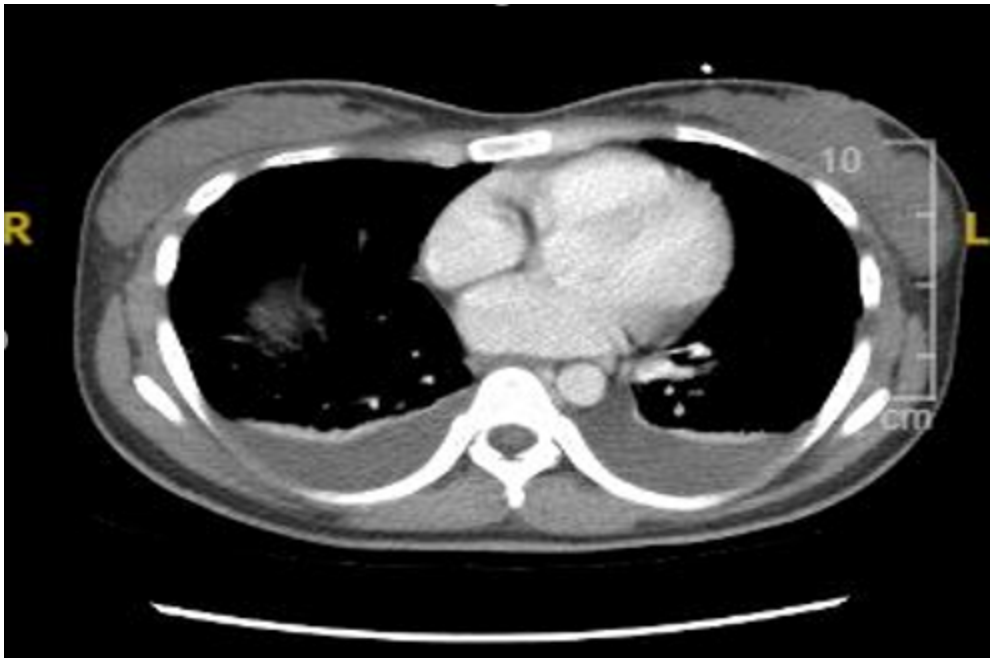
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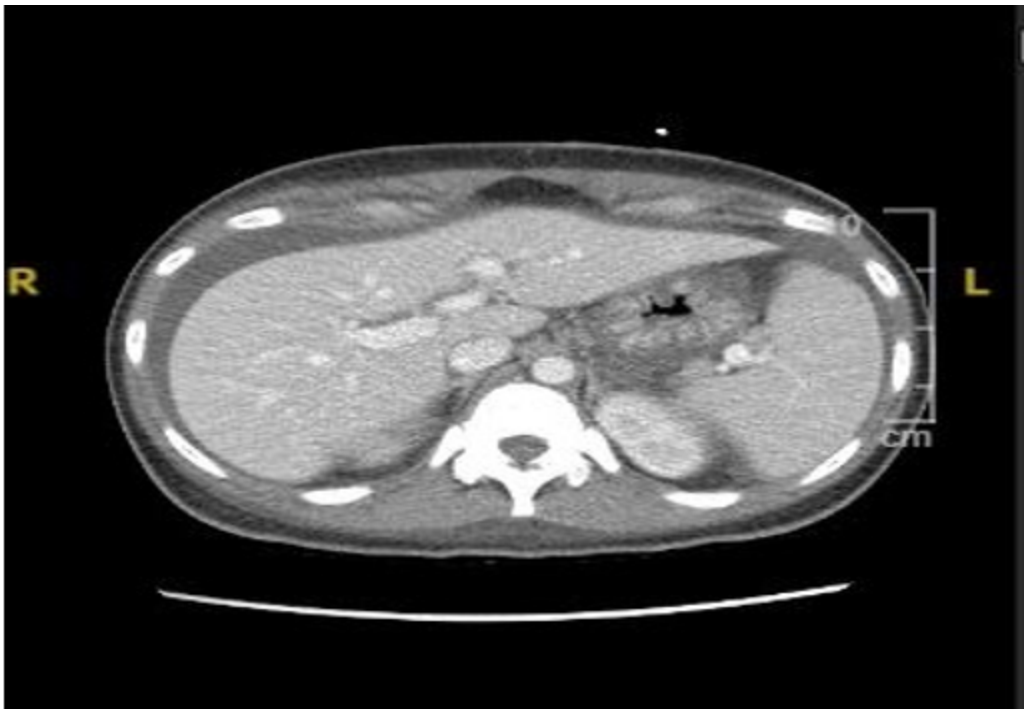
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Diagnostic Challenges in Hemophagocytic Lymphohistiocytosis: A Case of Overlapping Adult-Onset Still's Disease and HLH

Tarun Selvarajan MD, Lavanya Kannekanti MD, Samina Hayat MD
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Abstract

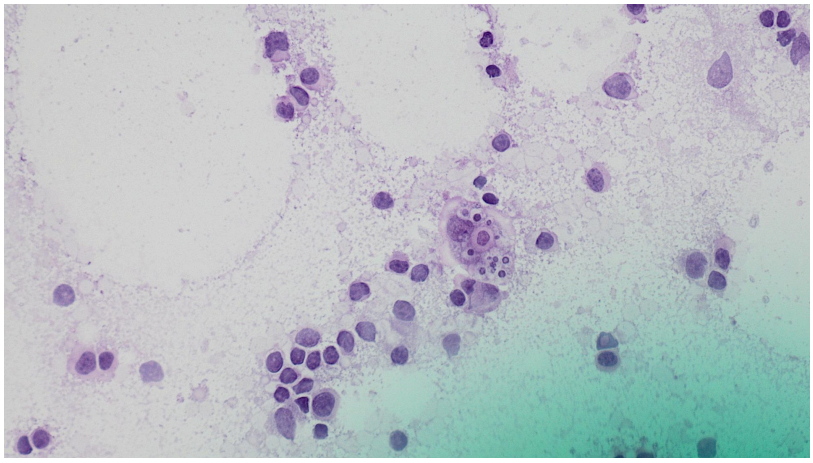
Introduction

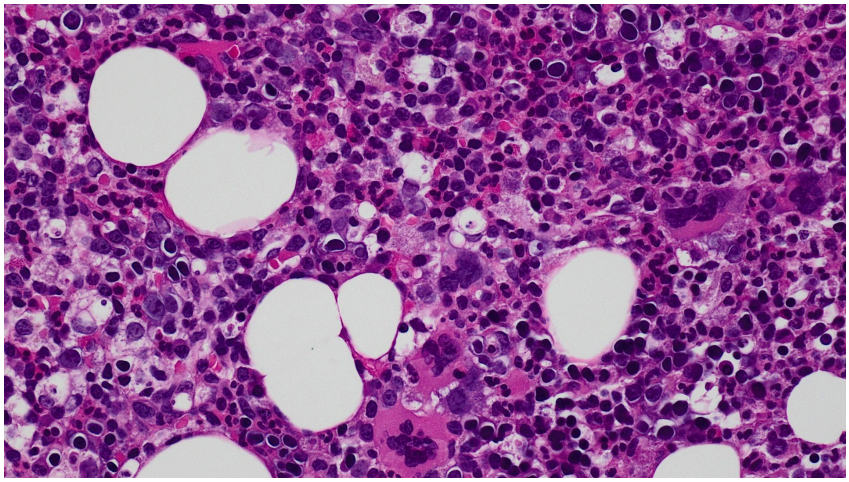
Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening hyperinflammatory syndrome characterized by immune dysregulation leading to multiorgan failure. HLH is classified as primary or secondary. The latter is more common in adults and is usually triggered by infections, malignancies, or autoimmune diseases and can mimic conditions like sepsis or hematologic malignancies, making early diagnosis challenging. Rheumatologists are key in identifying HLH, particularly in patients with features of macrophage activation syndrome (MAS-HLH) and persistent systemic inflammation.

Case

A 38-year-old female with a history of Still's disease and prior HLH presented with four days of fever, chills, nausea, vomiting, and non-bloody diarrhea.

- On initial evaluation, she was hypotensive with leukocytosis (13.95 $\times 10^9$/L), anemia (Hgb 9.9 g/dL), thrombocytopenia (34 $\times 10^9$/L), elevated procalcitonin (3.83 ng/mL), lactate (4.1 mmol/L), and hypofibrinogenemia (92 mg/dL). CT imaging revealed generalized lymphadenopathy, raising concern for inflammatory or lymphoproliferative pathology. She was started on broad-spectrum antibiotics.
- Her condition continued to worsen with increased leukocytosis (23.85 $\times 10^9$/L), thrombocytopenia (25 $\times 10^9$/L), hyperferritinemia (33,511 ng/mL), markedly elevated soluble IL-2 receptor levels (33,549 pg/mL), and coagulopathy. Differential diagnosis included HLH flare, sepsis, thrombotic thrombocytopenic purpura (TTP), or lymphoma. She was transferred to the ICU, and hematology and rheumatology were consulted. High-dose IV methylprednisolone (1 g/day) was initiated. Given her history of HLH, tocilizumab (8 mg/kg) was also started.
- Biopsy of lymph node and bone marrow were done; Lymph node biopsy confirmed HLH, while bone marrow biopsy showed no evidence of hemophagocytosis or malignancy. With clinical improvement, steroids were tapered, and she was discharged on oral prednisone with plans for outpatient tocilizumab and rheumatology follow-up.





Discussion

- This case illustrates the diagnostic complexity of HLH in the context of Still's disease. The patient met HLH-2004 criteria, and a high H-score supported the diagnosis. HLH should be considered in patients with unexplained systemic inflammation, especially with overlapping features of lymphoma. Early recognition through systematic laboratory evaluation and clinical suspicion is essential to initiate life-saving therapy.

No disclosures.

The Calcium Curveball

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Abstract

Background: Hypercalcemia, defined as serum calcium >10.5 mg/dL, is most commonly caused by primary hyperparathyroidism and malignancy. Granulomatous diseases can also induce hypercalcemia via extrarenal 1,25-dihydroxyvitamin D (calcitriol) production by activated macrophages. Rarely, foreign body reactions such as those triggered by silicone injections can mimic this mechanism.

Case Presentation: A 68-year-old female with a history of hypertension, stage III chronic kidney disease, and recurrent nephrolithiasis since 2021 presented after a mechanical fall and was found to have severe hypercalcemia (Ca 14.4 mg/dL; ionized Ca 6.9 mg/dL). Past evaluations revealed low PTH, low PTHrP, elevated and fluctuating calcitriol levels, with normal 25(OH)D, ACE levels, chest imaging, and serum protein electrophoresis. Kidney stones were predominantly composed of calcium phosphate. She denied typical hypercalcemia symptoms but reported a chronic buttock rash and remote history of silicone injections 25 years ago. CT imaging revealed diffuse gluteal and perineal granulomas with calcifications, consistent with silicone-induced granulomatous inflammation. She was treated with IV fluids, calcitonin, and zoledronic acid, with calcium normalization within one week. Surgical evaluation for silicone removal was arranged upon discharge.

Discussion: Silicone-induced granulomas trigger a chronic inflammatory response, wherein macrophages express 1α-hydroxylase, increasing calcitriol and resulting in enhanced calcium absorption and bone resorption. This pathophysiology resembles that of sarcoidosis. Clinical manifestations may occur decades post-exposure and include localized nodules, skin changes, and systemic complications such as hypercalcemia. In this patient, hypercalcemia contributed to nephrolithiasis and obstructive uropathy requiring multiple urological interventions.

Conclusion: Silicone-induced hypercalcemia is a rare but serious complication of cosmetic procedures. Clinicians should consider this etiology in unexplained hypercalcemia, especially in patients with a history of cosmetic silicone use. Early diagnosis and multidisciplinary management are essential to prevent long-term complications.

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Medication-Induced Systemic Lupus Erythematosus Following Dulaglutide Initiation: A

Case Report

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Abstract

Objective: To report a case of medication-induced lupus erythematosus (MILE) following dulaglutide initiation for type 2 diabetes mellitus, highlighting an unusual overlap syndrome with multisystem involvement.

Methods: Case report and analysis of clinical presentation, laboratory findings, diagnostic workup, treatment approach, and outcome in a 62-year-old female who developed symptoms of systemic autoimmunity three months after starting dulaglutide.

Results: A 62-year-old female with family history of systemic lupus erythematosus (SLE) presented with progressive weakness, polyarthralgia, intermittent fevers, leukocytoclastic vasculitis, and urinary symptoms three months after dulaglutide initiation. Laboratory evaluation revealed high-titer antinuclear antibodies (ANA >1:1280), anti-double-stranded DNA (1:2560), strongly positive anti-histone antibodies (3.1 Units), hypocomplementemia, and elevated inflammatory markers (ESR 130 mm/hr, CRP 238 mg/L). Liver biopsy confirmed autoimmune hepatitis with bridging fibrosis. The diagnosis of MILE was established based on temporal relationship with dulaglutide initiation, strongly positive anti-histone antibodies, and symptom resolution following drug discontinuation. Treatment included discontinuation of dulaglutide, high-dose corticosteroids, hydroxychloroquine, and azathioprine for hepatic manifestations, with subsequent clinical improvement.

Conclusion: This case describes an atypical presentation of MILE with features more commonly associated with idiopathic SLE, including high-titer anti-dsDNA antibodies and significant organ involvement. It contributes to growing evidence of GLP-1 receptor agonist-associated autoimmunity and underscores the importance of vigilance when prescribing these increasingly popular medications to genetically predisposed individuals.

Testicular Sarcoidosis as the initial manifestation of Sarcoidosis

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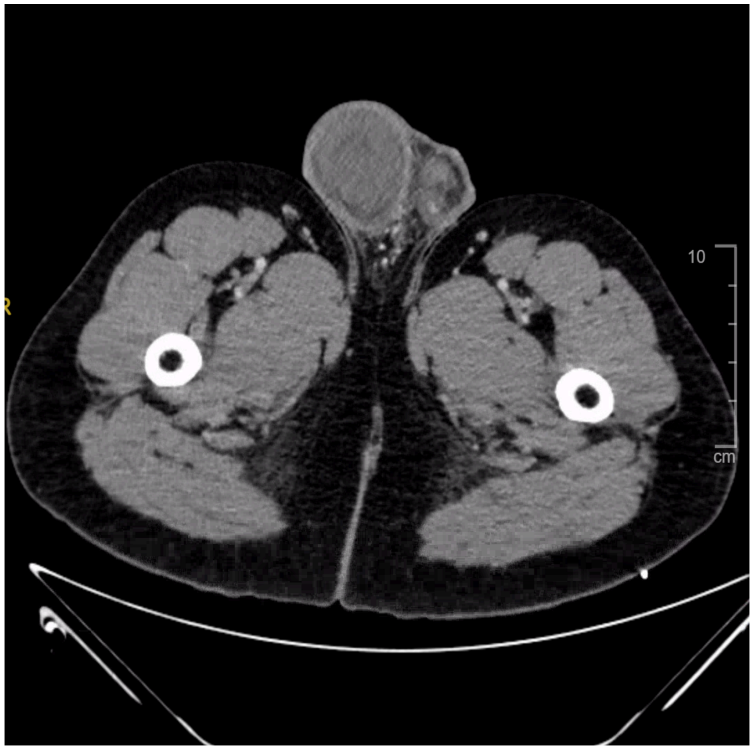
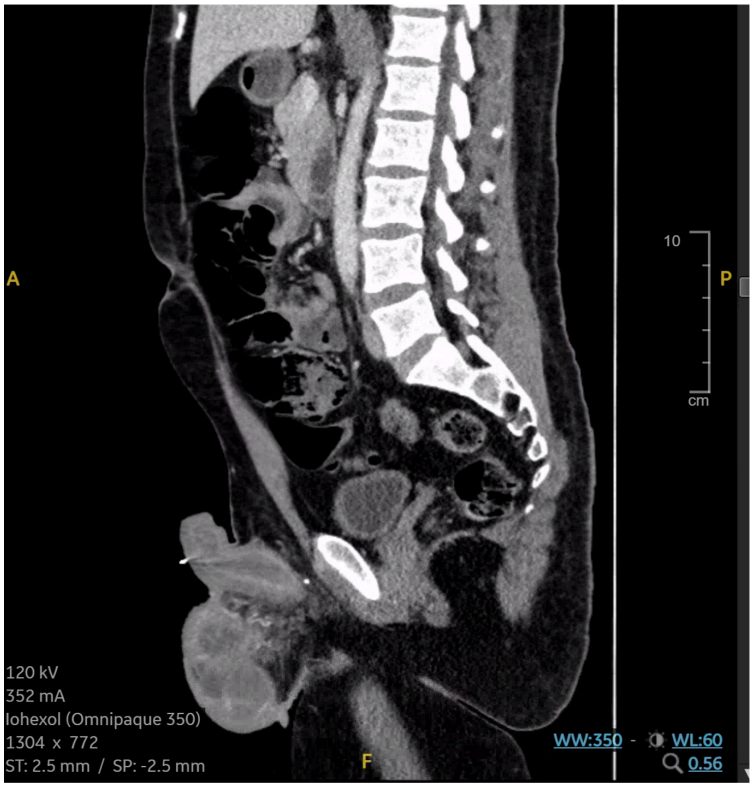
Abstract

Sarcoidosis is a systemic granulomatous disease, with genitourinary tract involvement accounting for only 0.2% of all sarcoidosis cases (1). Genitourinary sarcoidosis may present with a scrotal mass and testicular pain, often mimicking epididymo-orchitis or malignancy. Only 8 cases of genitourinary sarcoidosis have been reported in the literature in the last 14 years up to 2019 (1). The aim of this report is to highlight a rare and unusual presentation of sarcoidosis. A 29-year-old African American male presented with worsening bilateral testicular pain and swelling that had been ongoing for one year, along with shortness of breath and palpitations. Scrotal ultrasound showed bilateral cystic scrotal masses and 1 cm left testicular mass with vascularity, concerning for malignancy. CT of the chest, abdomen, and pelvis showed a large mediastinal lymph node, moderate-sized pericardial effusion, a heterogenous right testicular mass and a left-sided hydrocele. Tumor markers, beta-hCG and alpha-fetoprotein were normal, LDH mildly elevated. Urology was consulted. The patient underwent right testicular biopsy with pathology showing non-caseating granuloma concerning for Sarcoidosis. Flow cytometry analysis and staining from the biopsy were negative for CD30 and PAX-5, which would be positive in Hodgkin Lymphoma. These stains also ruled against a metastatic germ cell tumor. Additionally, the sample was negative for acid-fast bacilli and fungal organisms. He also underwent pericardiocentesis for pericardial effusion during the same hospitalization, and the pericardial fluid was negative for malignant cells. The patient was diagnosed with Sarcoidosis and high dose oral prednisone was prescribed. He was evaluated by Rheumatology outpatient with a plan to taper Prednisone and start Methotrexate. Labs obtained as outpatient were significant for elevated Angiotensin Converting Enzyme, Lysozyme and IL-2 Receptor Alpha, supporting the diagnosis further.

Testicular sarcoidosis is a rare initial presentation of sarcoidosis that can be mistaken for malignancy. Accurate differentiation is critical, as misdiagnosis may lead to unnecessary aggressive treatments like orchiectomy, chemotherapy, or radiation (3). Unlike malignancy, sarcoidosis is a benign granulomatous condition that often resolves on its own or with corticosteroids. While tumor markers and elevated serum ACE levels can aid in diagnosis, biopsy remains the definitive method for distinguishing between the two.





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Implementing high value fragility fracture care at a safety net facility in Massachusetts

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Abstract

Background
Fragility fractures are defined as pathological fractures that occur after minimal trauma, such as a fall from a standing height.[1,2] In the elderly, fragility fractures are associated with significant morbidity and mortality within year following initial injury.[1,3,4] Through surgical and medical co-management, patient outcomes improve drastically, leading to decreased mortality at one year.[3,5] In high resource and volume centers, dedicated fracture liaison services have helped coordinate care for these patients effectively. However, a dedicated service may not be possible in all community centers. In this work, we showcase an improvement plan for fragility fracture care at a safety-net facility, Cambridge Health Alliance, in Massachusetts that began in 2024. Our approach highlights opportunities to improve fragility fracture care in the community setting.

Objectives
To improve quality of care for admissions for surgical management of fragility fractures.
To improve the percentage of patients receiving mortality reducing medical care for fragility fractures (e.g., inpatient administration of bisphosphonates) during their inpatient stay.
To improve outpatient follow-up with orthopedic and rheumatology/endocrinology specialists.

Methods/Outcomes
This quality improvement initiative began with the identification of the current care pathway for patients with fragility fractures (Figure 1) through multidisciplinary meetings that included internal medicine, endocrinology, rheumatology, and orthopedics. This led to several initial interventions including an order set to standardize initial laboratory monitoring of vitamin D deficiency and hypocalcemia, a streamlined referral process to either rheumatology or endocrinology, and templates for evaluation for inpatient bisphosphonate administration. Additionally, a planned future process map (Figure 2) was created and a monitoring program with key performance indicators was initiated.

Patients included in prospective monitoring in this program included all individuals age >65 with one or more admission ICD codes for a surgically managed femoral or hip fracture. Key performance indicators include the percentage of patients for whom the fragility fracture admission order set was completed, the percentage of patients who received zoledronic acid while inpatient, percentage of patients who received vitamin D or calcium supplementation while inpatient, percentage of patients with scheduled orthopedic follow-up and either rheumatology or endocrinology visits. At the time of submission, these data are pending.
Figures

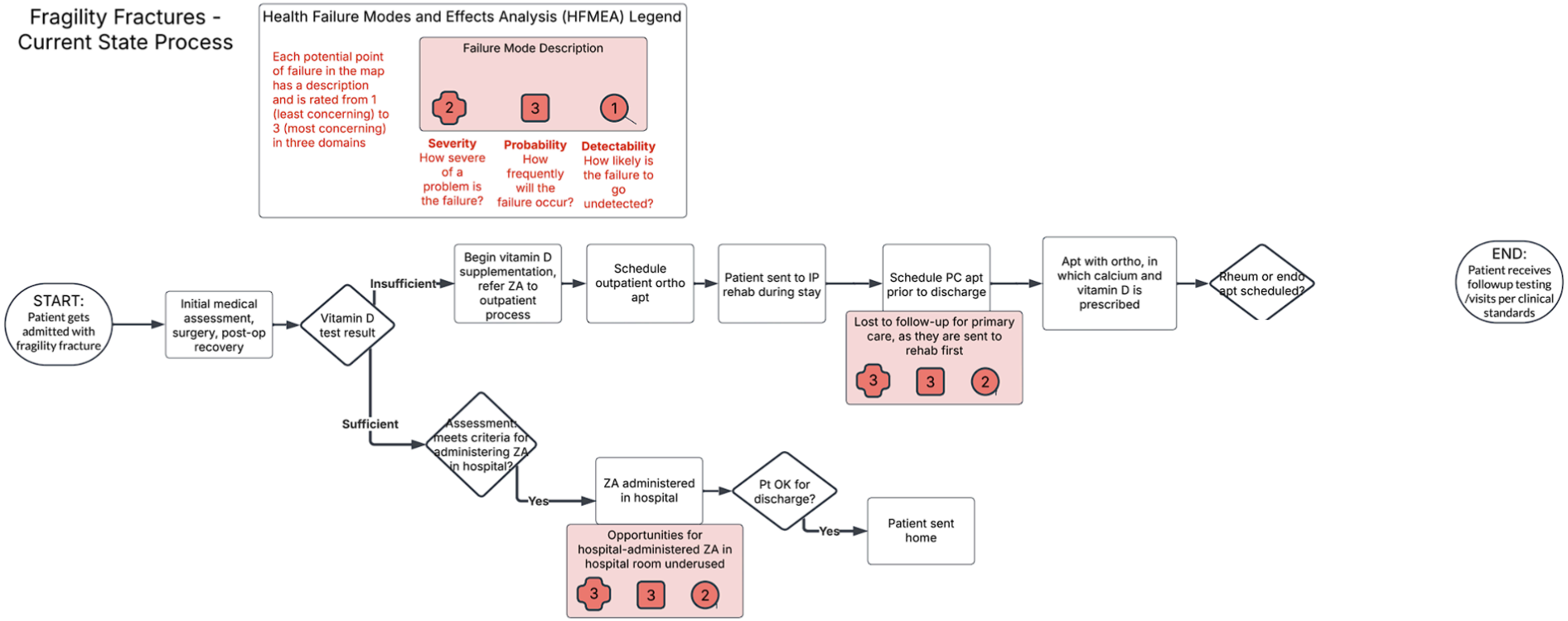

Figure 1. Current state process map

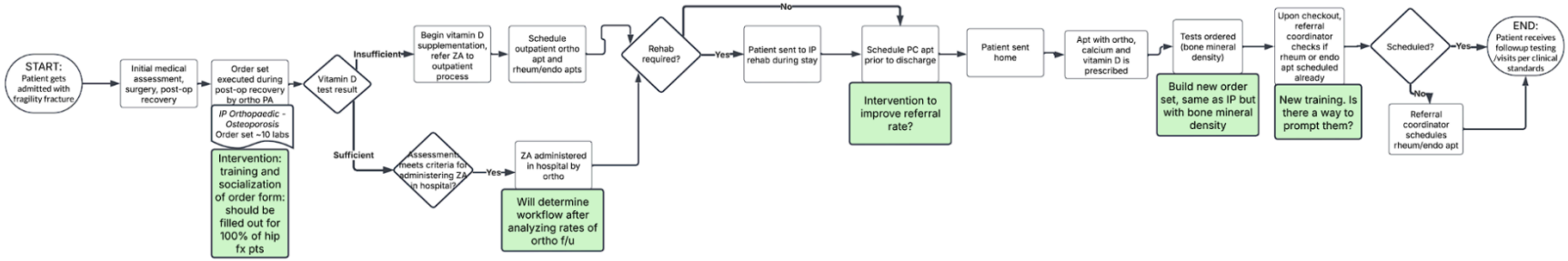

Figure 2. Planned improvement process map

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Unmasking IgG4 Related Disease – A Case of Illusive Diagnosis

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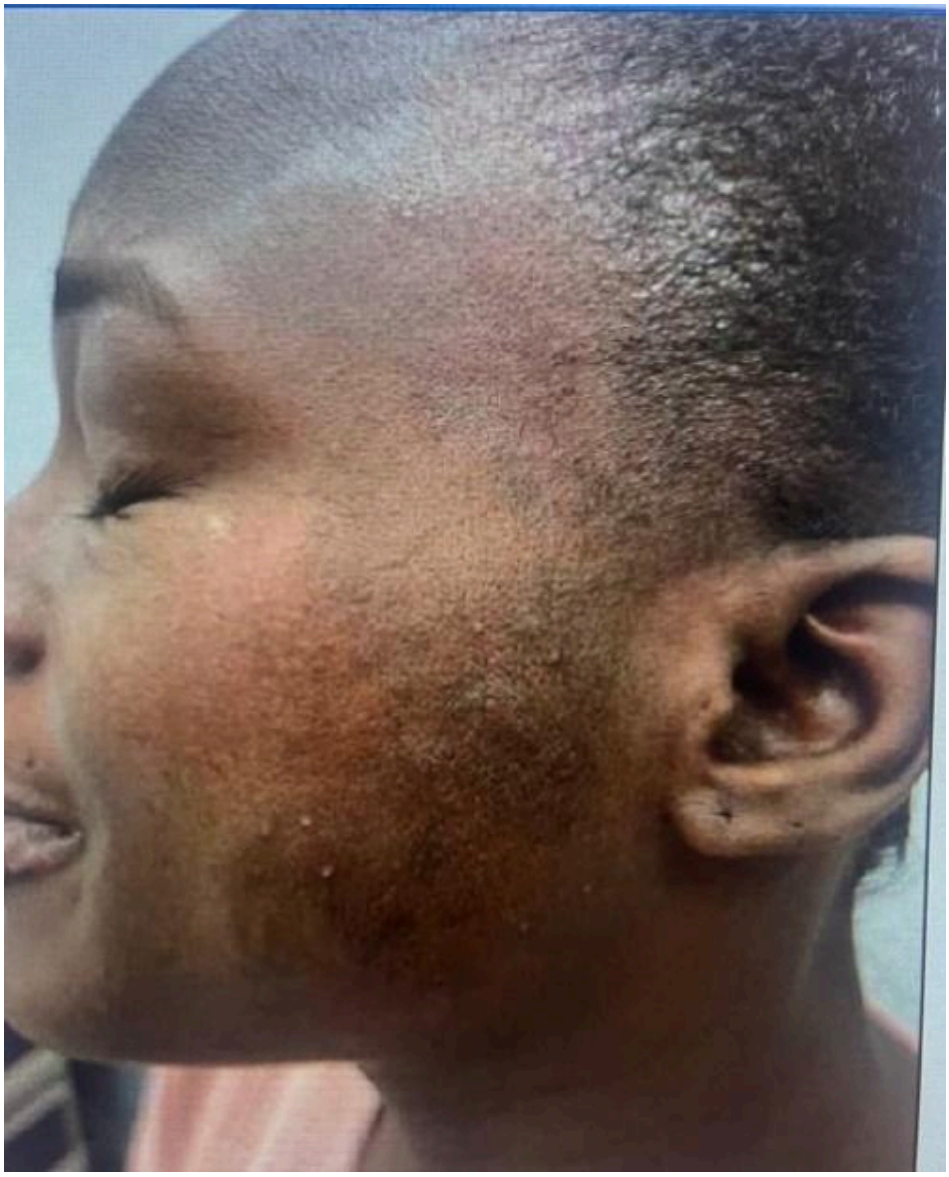
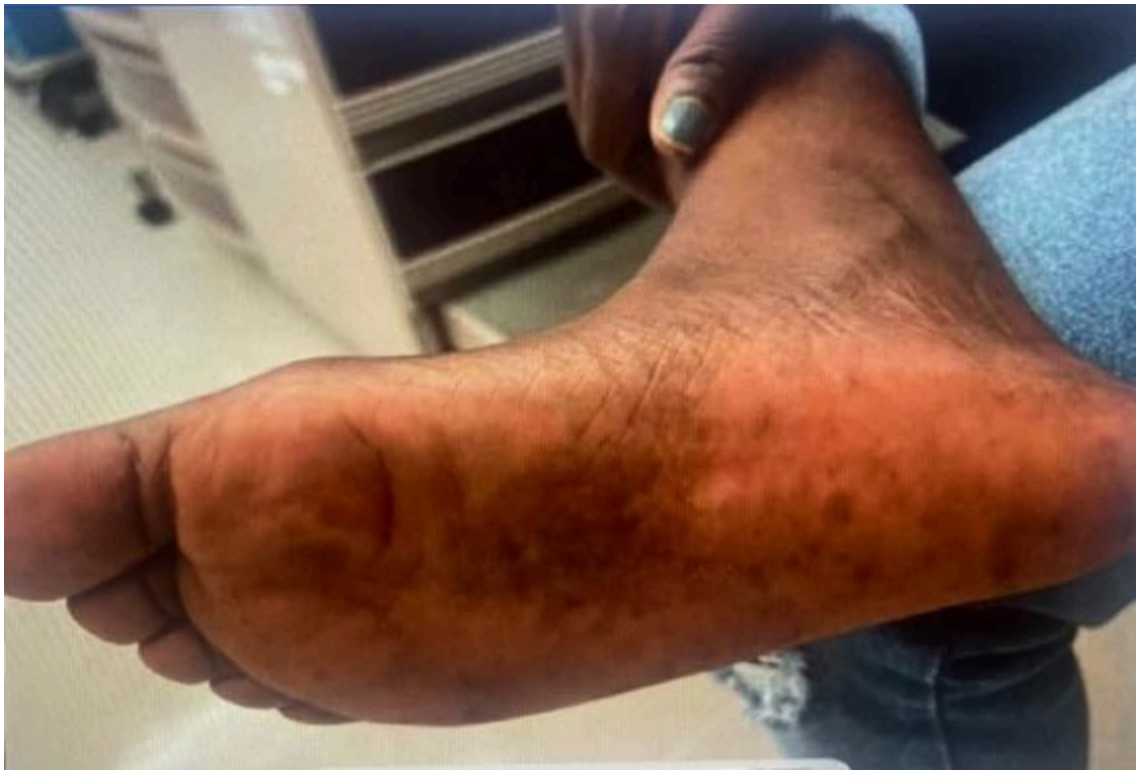
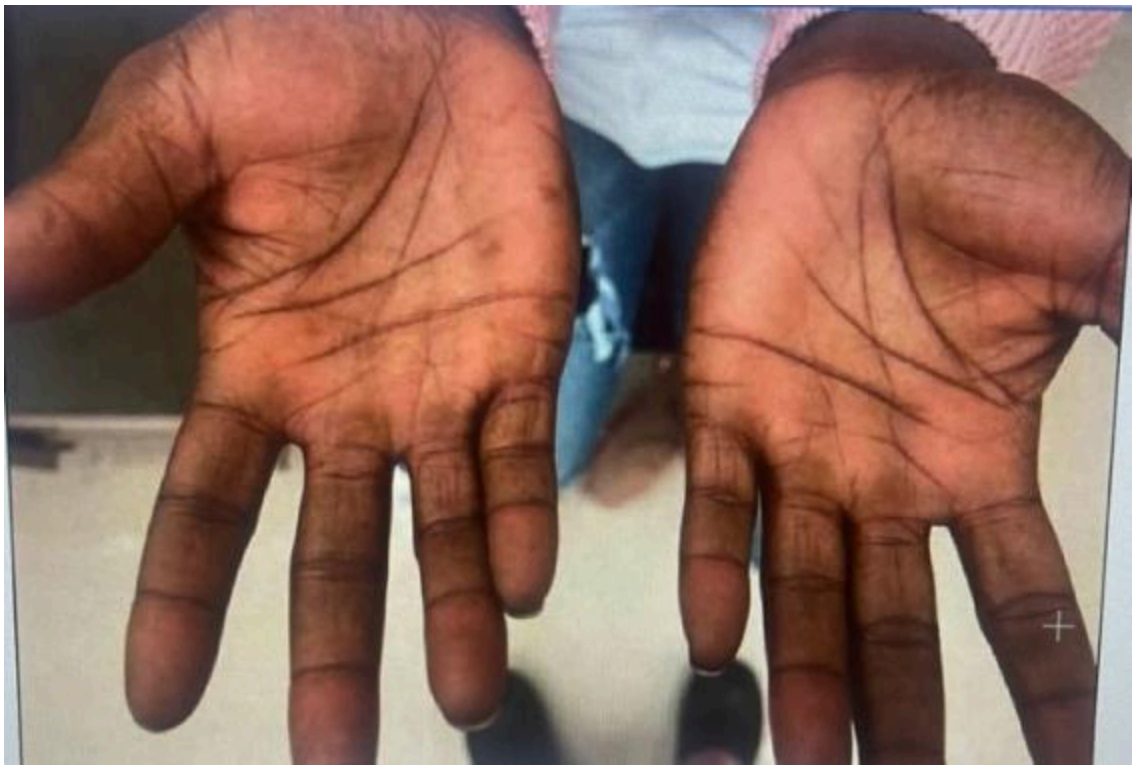
Abstract
Introduction:
IgG4-related disease (IgG4-RD), a systemic fibroinflammatory condition, was initially identified in patients with autoimmune pancreatitis, is now known to involve multiple organ systems, including salivary glands, pancreas, lymph nodes and kidneys.

Case presentation:

- 





- A 33-year-old African American female with a 10-year-long prior history of recurrent sinusitis presented with multiple joint pains, hyperpigmented skin rash on her face, palms and soles, pre-orbital swelling, cervical lymphadenopathy and dry mouth.
- Lymph node enlargement was noted. Biopsy suggested reactive adenopathy.

- Laboratory tests: ANA (1:320), other autoimmune workup including complement levels, Myo-marker panel and a vasculitis panel - negative. X-rays of the hands and chest - unremarkable.
- Started on hydroxychloroquine - minimal symptom improvement.
- Later developed acute pancreatitis. IgG4 levels were elevated at 209 mg/dL. Underwent follow-up MRCP and EUS, both showed persistent diffuse enlargement of the pancreas. Suspecting IgG4 related pancreatitis, the patient was started on 6 6-week steroid taper.
- Lymph node and nasal cavity mucosa biopsy was negative for IgG4 related disease, but the patient reported significant improvement in symptoms with steroid therapy.
- Repeat biopsy of minor salivary glands from the lower lip was positive for lymphoplasmacytic infiltrates, with 95% of plasma cells positive for IgG4.
- Rituxan was initiated.

Discussion:

- IgG4 RD often involves lymphoid and glandular tissue, presenting as swelling/ persistent mass with symptoms mimicking malignancies, infections, or other inflammatory/autoimmune disorders..
- The affected patients have lymphadenopathy without fever and have excellent responses to steroids, which can serve as a clue in narrowing the differential diagnosis.
- Elevated IgG4 levels are highly sensitive, but are insufficient to diagnose IgG4-RD alone, given low specificity.
- Biopsy with Histopathology and immunohistochemical staining is confirmatory in the right clinical context.
- Negative biopsy should not rule out the diagnosis where clinical suspicion remains strong. To ensure biopsy sample adequacy, core or excisional biopsy should be preferred over FNAC and repeat biopsy should be considered if a false negative is suspected.

Bullous lupus: A Rare Dermatologic manifestation in Systemic lupus erythematosus

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Abstract

Introduction:
Bullous lupus erythematosus (BLE) is a rare cutaneous manifestation of systemic lupus erythematosus (SLE), affecting 1-5% of patients. Early recognition and treatment are vital to preventing complications. This case highlights the presentation of BLE, and its management.

Case Description:
A 38-year-old African-American female with systemic lupus erythematosus (SLE), seronegative rheumatoid arthritis, and antiphospholipid syndrome presented with right eye swelling/itching, and swelling of the right elbow and left fourth digit.

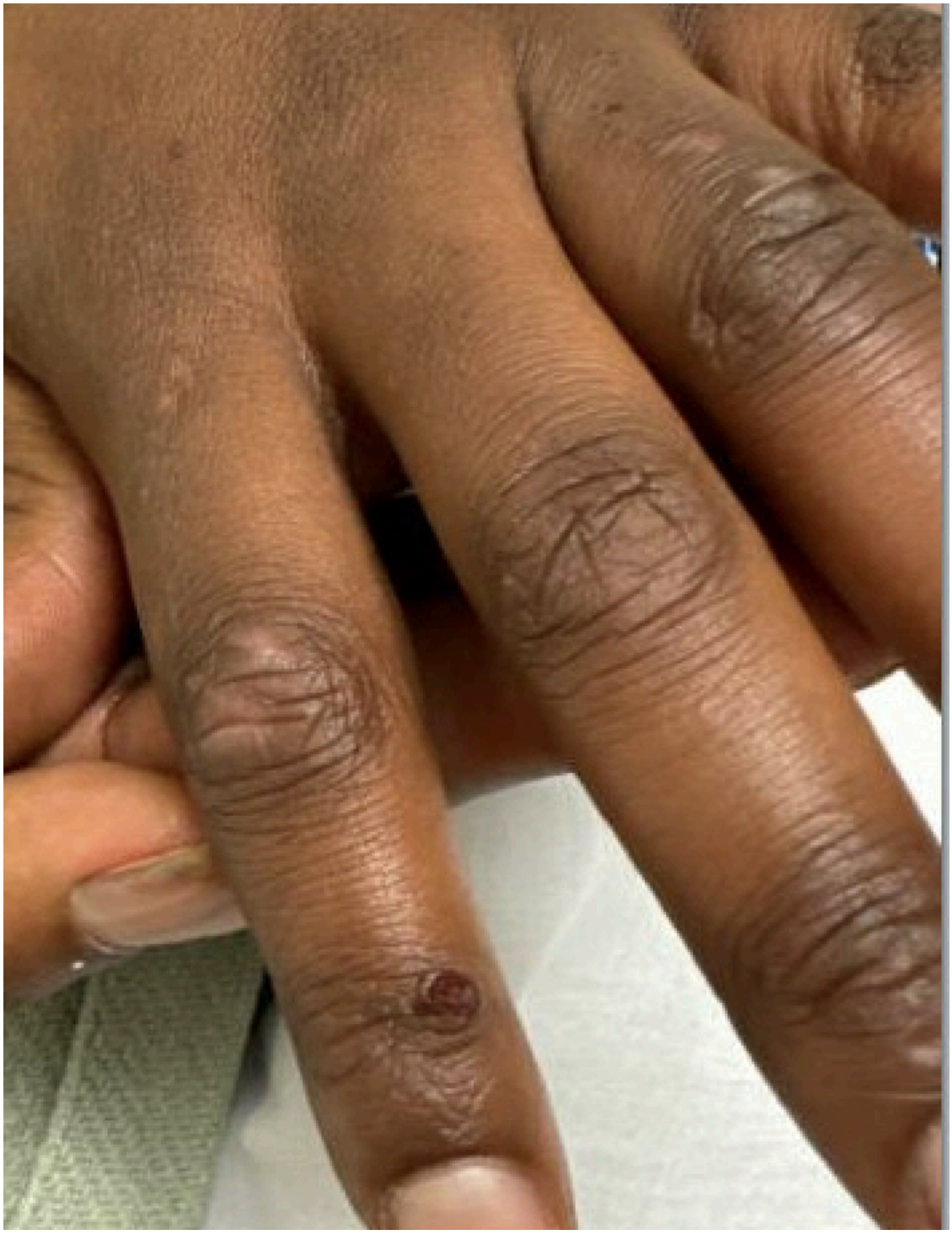
She had a history of significant inflammatory arthropathy and had limited success to treatment with multiple DMARDs and biological therapies. She is currently on Methotrexate(MTX) 20 mg PO weekly, Folate 1 mg daily, Hydroxychloroquine(HCQ) 300 mg daily, and Belimumab 200 mg SC weekly. She reported noticing recurrent blisters on all extremities.

No fever or leukocytosis were noted in this admission. CT Maxillary sinus/Face demonstrated concerns for periorbital cellulitis. Differentials included angioedema vs cellulitis, hence was briefly treated with antibiotics, but later improved with antihistamines alone. Tryptase level, C1 esterase inhibitor level and function were normal. Skin biopsy showed subepidermal blisters with abundant neutrophils, but IgG was negative. In the given clinical setting it was regarded as bullous lupus.

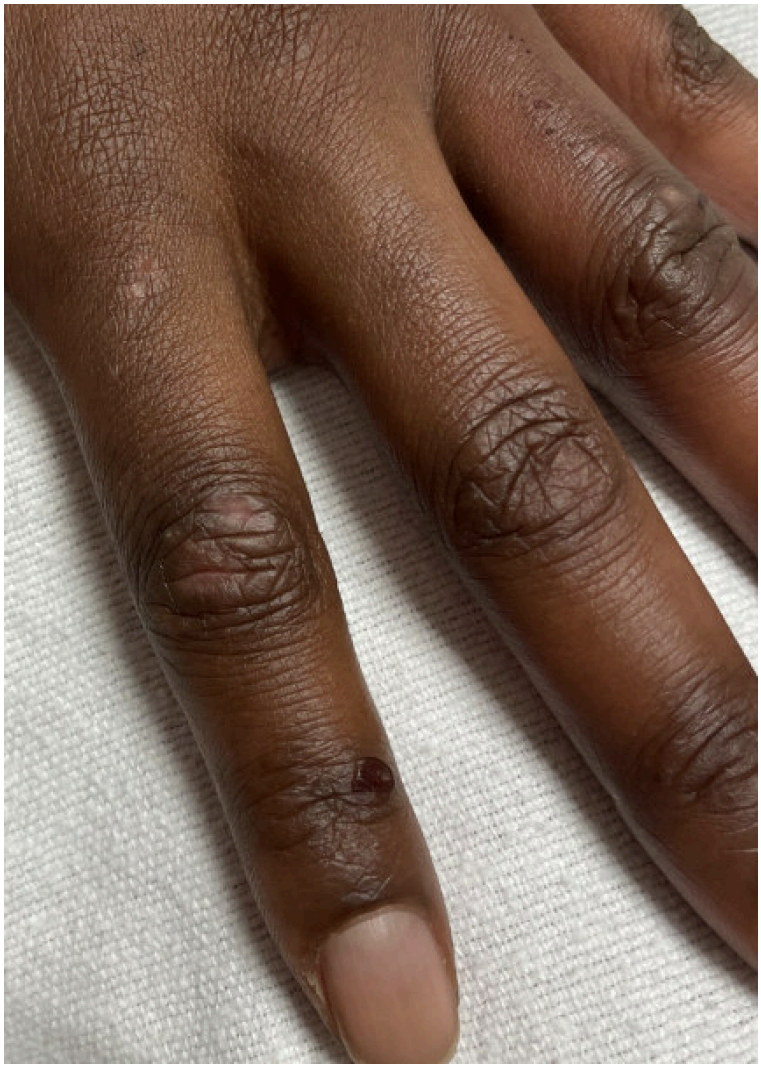
However, Dapsone was avoided due to G6PD deficiency. MTX, HCQ and Belimumab were continued. Given no significant improvement,she was later switched to Anifrolumab, which resulted in the complete resolution of skin lesions and improvement in joint pain.

Discussion:
BLE occurs in approximately 1-5% of lupus patients and is characterized by subepidermal blistering due to autoantibodies targeting type VII collagen. BLE commonly presents with widespread vesiculobullous lesions, predominantly on sun-exposed areas. Diagnosis is confirmed via histopathology and direct immunofluorescence demonstrating IgG and complement deposition at the dermal-epidermal junction. Management typically involves systemic corticosteroids, dapsone (if tolerated), immunosuppressants, and biologic therapies such as rituximab or Anifrolumab in refractory cases.

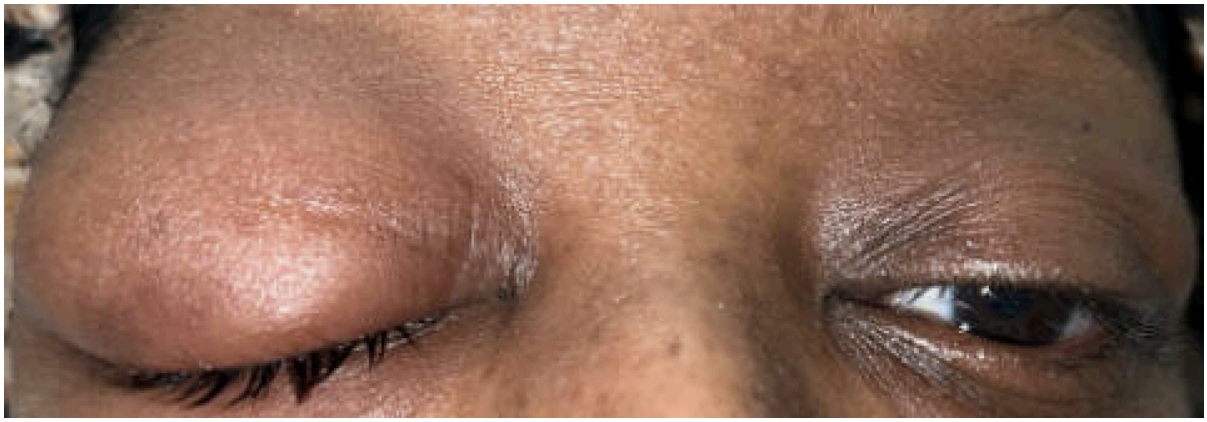
Conclusion:
Although rare, BLE should be considered as a differential diagnosis in patients with bullous lesions associated with SLE.



Finger showing bullous lesion



Bullous lesions and scars on fingers from old lesions



Swollen right eye on presentation

Unveiling Kikuchi-Fujimoto Disease: A 34-Year-Old Spanish Woman's Journey with Cervical Lymphadenopathy and Cutaneous Symptoms

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Abstract

INTRODUCTION:
Kikuchi-Fujimoto Disease (KFD), first identified in 1972, is a benign, self-limiting condition characterized by cervical lymphadenopathy and nonspecific symptoms, with rash being the main extranodal manifestation.

CASE PRESENTATION :
A 34-year-old Spanish-speaking female presented with left neck swelling, erythema, fever and pain since December. Initial IV steroids and amoxicillin were ineffective. Symptoms worsened with new nonpruritic leg rash, bilateral neck swelling and leukopenia, prompting hospital visit. Exam showed swollen left parotid and bilateral submandibular glands, along with maculopapular purpulish rash. Labs revealed leukopenia ($2.9\times10^3/\mu L$), anemia (10 g/dL), low albumin (2.9 g/dL), and elevated LDH (707 U/L). Ultrasound revealed bilateral cervical lymphadenopathy, prominent on left, up to 2.7 cm. CT neck showed similar findings. Excisional biopsy of the left cervical lymph node was done. While awaiting results, she received broad-spectrum antibiotics and an infectious workup, which was negative except for EBV. Biopsy showed scant viable lymphoid tissue with extensive necrosis,and apoptosis with CD20/CD79a/PAX5 and CD3/CD5 staining, suggesting Kikuchi syndrome. Autoimmune workup for mimickers was negative. Treatment with prednisone 60mg and IVIG therapy improved WBC, hemoglobin, reduced LDH to 260, and improved rash. She was discharged with rheumatology follow-up on prednisone taper.

DISCUSSION:
KFD predominantly affects young Asian females but is reported worldwide. It presents with unilateral cervical lymphadenopathy, with bilateral cases in 13.6%. Cutaneous symptoms occur in 16.6-40% of patients, including vasculitis, malar erythema, oral apthous ulcer, alopecia, and lupus-like papules on the upper body. Our patient's skin eruptions appeared on lower extremities. KFD's cause remains unclear, with viral or autoimmune origins suspected. Epstein-Barr virus and herpesvirus are implicated, alongside T-cell-mediated autoimmune responses. Diagnosis requires a lymph node biopsy showing a necrotic nodule and cellular apoptosis. Minimum criteria include aggregated histiocytic and scattered karyorrhexis. KFD mimics lymphoma, tuberculosis, lupus, and metastatic adenocarcinoma, requiring definitive workup. Labs typically show normal counts, although leukopenia (43%) and anemia (23%) may occur, with negative results for antinuclear antibodies and rheumatoid factor. Our patient mirrored these results. Treatment involves supportive care, though corticosteroids and immunoglobulins may accelerate recovery.
CONCLUSION:

Physicians should consider KFD in patients with neck lymphadenopathy and skin lesions to ensure timely diagnosis, prevent unnecessary tests, and reduce misdiagnosis.

Isolated Superior Mesenteric Vasculitis due to Coronavirus OC43

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Abstract

Introduction:
Vasculitis is a condition defined by inflammation of blood vessels. Often, vasculitis occurs as part of a systemic inflammatory condition, however it can occur in isolation. We present a case of isolated mesenteric vasculitis affecting the superior mesenteric artery (SMA), which occurred without signs of systemic inflammation, after coronavirus OC43 infection.

Case presentation:

Patient is a 40 yo AA F who presented to the ER for 3 days of worsening diffuse abdominal pain and two weeks of anorexia. She had presented to the ER two weeks prior due to a URI. Viral panel positive for coronavirus OC43. MRA abdomen showed thickening of the SMA and surrounding haziness, again consistent with vasculitis. Labs on presentation: ESR 71 mm/hr and CRP 36 mg/L, otherwise, unremarkable. She was given Methylprednisolone 125 mg IV with improvement within 12 hours transitioned to Prednisone 80mg daily. On discharge, prednisone was tapered by 20mg every two weeks. ESR and CRP trended down to 30 and 1.6mg/L; she was off prednisone. Repeat MRA showed no evidence of SMA wall thickening or fat-stranding, indicating resolution.

Figure 1A-B:

A B

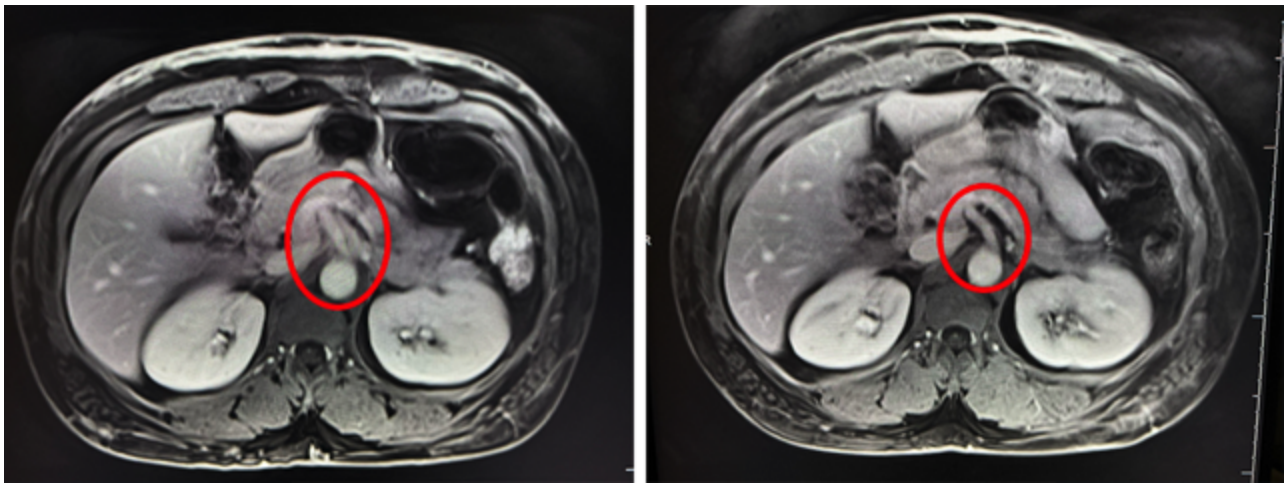

Figure 1A: Circled is the SMA as it leaves the abdominal aorta. SMA shows thickening and surrounding haziness which is consistent with vasculitis. Figure 1B: SMA shows resolution of thickening and surrounding haziness.

Discussion:

Coronavirus OC43 is most well known to cause mild URI symptoms. Diagnosis of isolated mesenteric vasculitis requires imaging studies or pathology, and angiography is now used to diagnose in most reported cases. MR angiography can effectively evaluate for vasculitis and may show arterial wall thickening, luminal narrowing and aneurysms. Medically, glucocorticoids remain the cornerstone of treatment. Management may require additional immunosuppressants to control the disease. As nearly 25% of patients will progress over a 5 year period, a minimum follow up of 6 months should be done to confirm the isolated nature of the condition.

Conclusion:

In conclusion, SMA vasculitis, while uncommon, should be considered when a patient presents with unexplained abdominal pain. This case exhibits a rare case of mesenteric vasculitis secondary to a common viral pathogen, coronavirus OC43.

Paraneoplastic Lupus Nephritis in Non-Small Cell Lung Cancer

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Abstract

INTRODUCTION: Paraneoplastic Systemic Lupus Erythematosus (SLE) is a rare autoimmune phenomenon triggered by malignancy. Even more uncommon is lupus nephritis presenting in this context and associated with non-small cell lung cancer (NSCLC).

CASE PRESENTATION: A 77-year-old man with a history of smoking presented with four weeks of progressive dyspnea, weakness and intermittent hemoptysis. He had experienced a 30-pound unintentional weight loss, arthralgias, and gait imbalance over the preceding months. Initial outpatient evaluations by neurology and rheumatology were inconclusive and he was treated empirically for polymyalgia rheumatica.

On admission, the patient was tachycardic and in respiratory distress. Lab results showed severe anemia (hemoglobin 6.0), thrombocytopenia (platelets 87), elevated BUN (102), creatinine (6), and proteinuria. Imaging revealed a right upper lobe lung mass. He deteriorated clinically with respiratory failure and sepsis requiring ICU admission and mechanical ventilation. Lung biopsy confirmed poorly differentiated NSCLC. Autoimmune serologies were notable for high-titer ANA (>1:2560), anti-dsDNA (>1:1280), p-ANCA (1:1280), anti-Smith/RNP (23), PR3 (136), MPO (73), anti-cardiolipin antibodies (IgG-34, IgM-150) and hypocomplementemia (C3-43, C4-3.6). He had a kidney biopsy which showed focal proliferative glomerulonephritis with hyaline thrombi and full house staining with focal segmental glomerulosclerosis, consistent with lupus nephritis. Given the absence of classic SLE symptoms and the newly diagnosed NSCLC, a diagnosis of paraneoplastic SLE with lupus nephritis was made. The patient was not a candidate for chemotherapy and passed away from multidrug resistant *Pseudomonas aeruginosa* bacteremia.

DISCUSSION: While paraneoplastic syndromes are well-documented in NSCLC, renal involvement mimicking lupus nephritis is rare and diagnostically challenging. The pathogenesis is hypothesized to involve tumor antigens or toxins produced by tumor cells that activate an auto-immune reaction. Immunologic markers may be markedly elevated despite the absence of traditional SLE features. This case emphasizes the significance of considering malignancy in atypical or serologically intense autoimmune presentations. Timely recognition of paraneoplastic autoimmunity is essential, as treating the underlying malignancy may lead to resolution of symptoms and prevent unnecessary immunosuppression.

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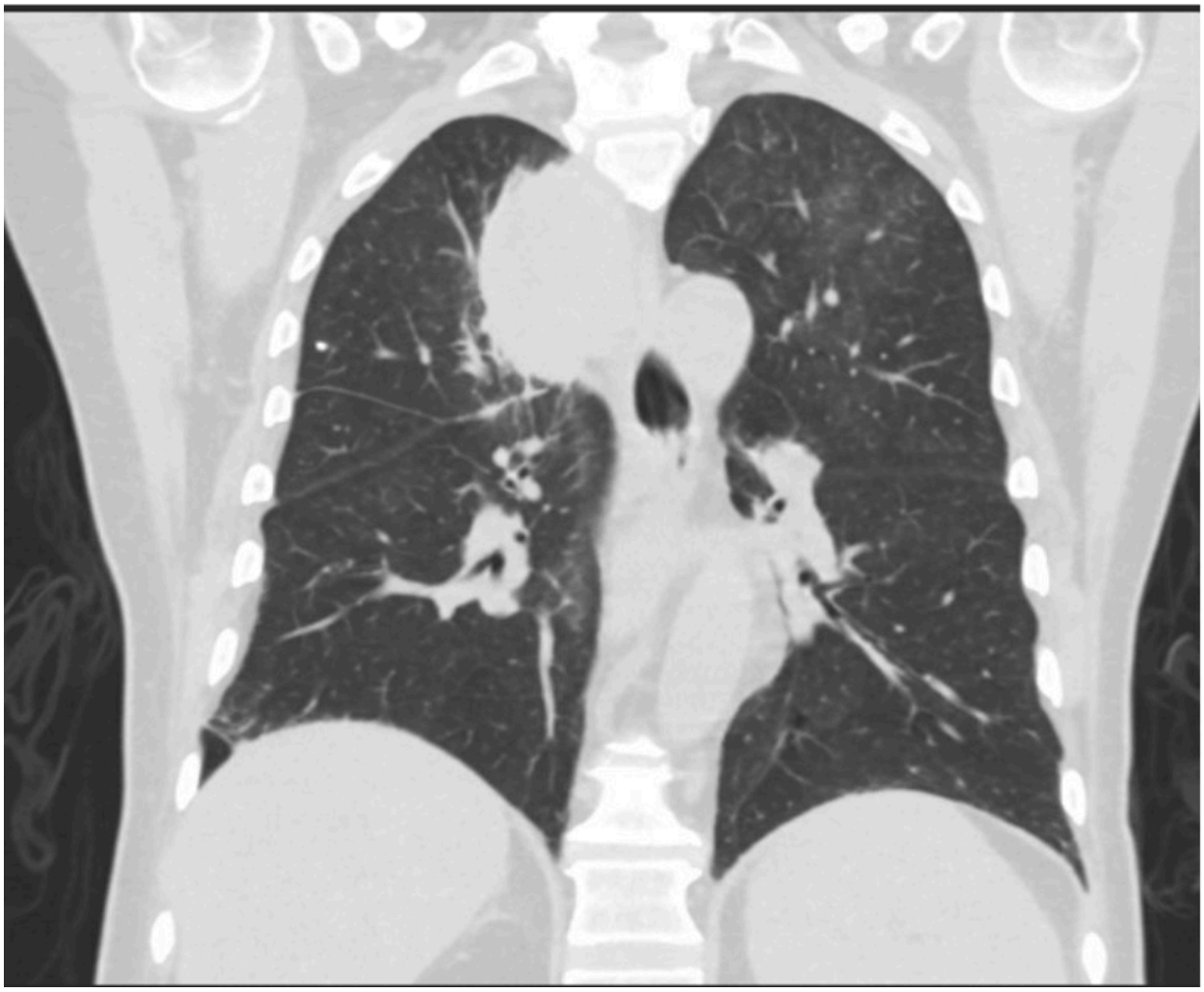

CT chest showing right medial upper lobe mass

Anti-SRP Immune-mediated Necrotizing Myopathy Associated With Coxsackie B Virus Infection

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Abstract

INTRODUCTION: Rhabdomyolysis (RM) is a condition with diverse etiologies, including trauma, infections, toxins, autoimmune disorders, and genetic factors. One of its rare causes is immune-mediated necrotizing myopathy (IMNM), a subtype of idiopathic inflammatory myopathy (IIM), which can lead to severe muscle damage. IMNM has a challenging and poorly understood pathogenesis. Here, we present a case of IMNM with a potential novel trigger that may provide insights into its pathophysiology.

CASE PRESENTATION: A 43-year-old previously healthy woman presented with bilateral lower limb weakness and pain. Four weeks prior, she experienced flu-like symptoms, followed by progressive muscle weakness and pain in the lower limbs a week later. On examination, she had bilateral lower limb tenderness and motor strength of 4/5. Initial laboratory results showed a significantly elevated creatine kinase (CK) of 22,207 U/L, elevated troponin of 579 ng/L, and elevated liver function tests (AST- 706 U/L, ALT- 589 U/L), while her creatinine levels were normal. There was no history of trauma, strenuous exercise, substance use, or medication exposure. MRI of the lower limbs revealed multifocal myositis, and empiric treatment with intravenous (IV) steroids and IV immunoglobulin (IVIG) was started.

On day 11, anti- Signal Recognition Particle (SRP) antibodies tested positive, followed by a positive result for Coxsackievirus B4 (CVB4) antibody. The patient received rituximab, and her steroids were tapered. By day 15, her CK had decreased to 7,400 U/L, liver enzymes improved, and she regained partial muscle strength. A muscle biopsy obtained 10 days later confirmed IMNM, showing perivascular and endomysial lymphocytic infiltration, myocyte necrosis, and regeneration.

DISCUSSION: This case presents a rare association between IMNM and CVB4 infection. While previous studies have implicated CVB4 in triggering autoimmune conditions like dermatomyositis and myocarditis, this is the first report of CVB4 being linked to anti-SRP positive IMNM. Our findings highlight the importance of considering virus-triggered IMNM in patients presenting with unexplained RM, particularly when standard risk factors are absent. This case may pave the way for future research on the role of viral infections in the pathogenesis of IMNM.

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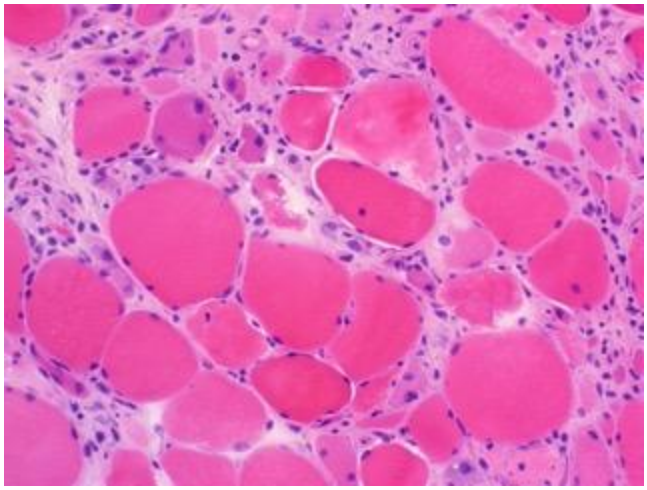
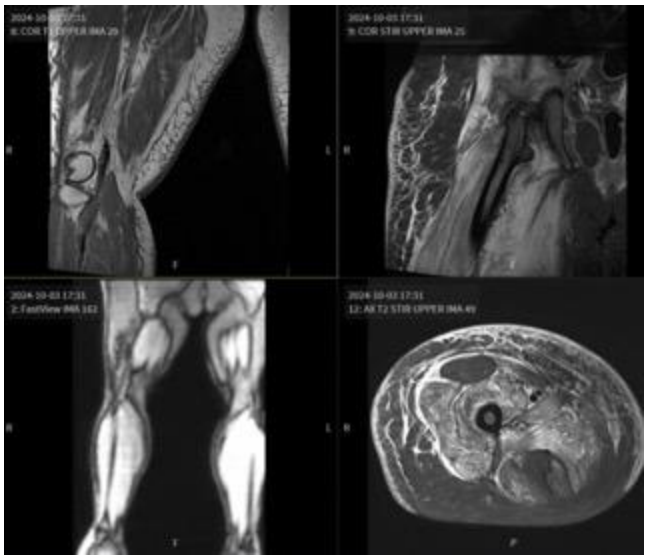
IMAGES



H&E stain of skeletal muscle biopsy (20X) showing myofibers of variable diameters with degeneration/ regeneration basophilia and endomysial areas with fibrosis and mixed lymphocyte and macrophage infiltrates



MRI of the right lower limb showing relatively diffuse heterogenous edema related to multifocal myositis

When Negative Isn't Always Positive: A Rare Case of ANA-Negative Lupus Nephritis

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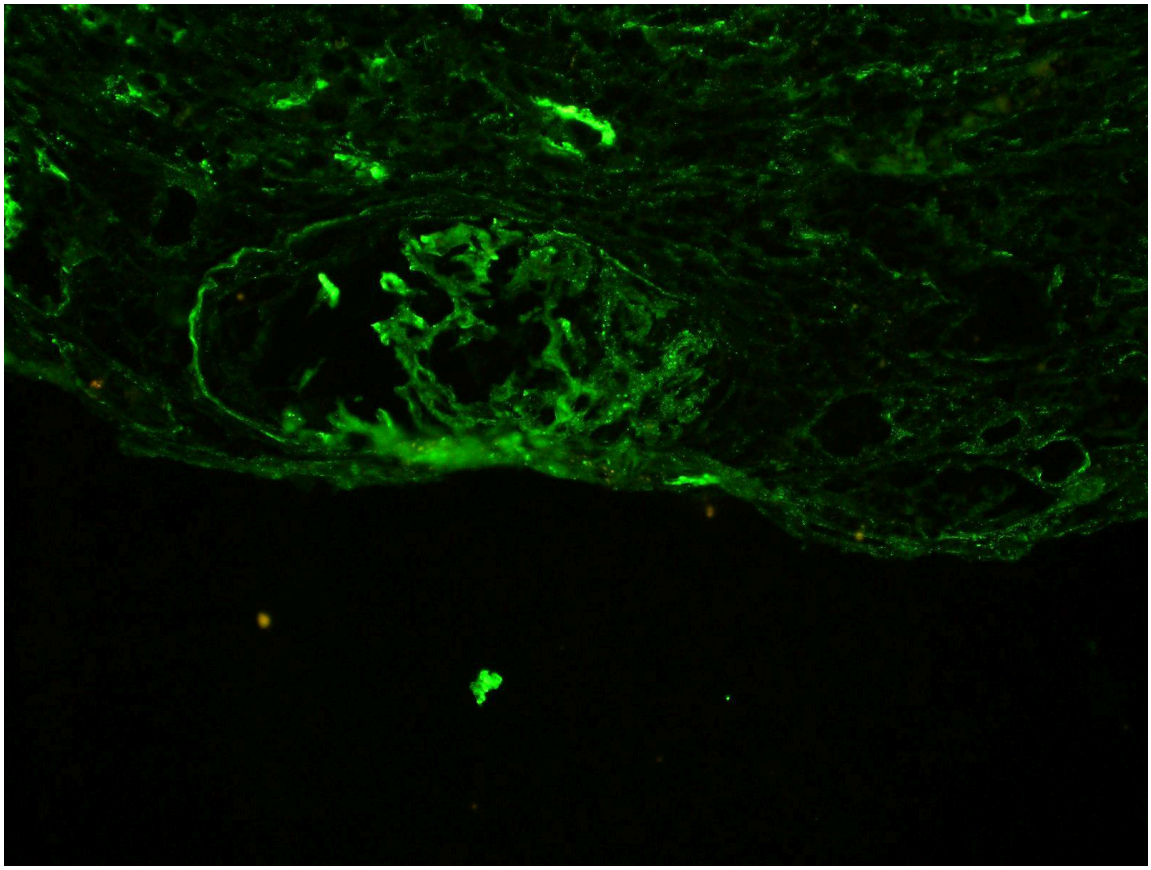
Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by multisystem involvement and autoantibody production, most notably antinuclear antibodies (ANA). However, approximately 2–5% of patients with SLE are ANA-negative, presenting a diagnostic challenge, particularly in cases of lupus nephritis. ANA-negative lupus nephritis is rare and may require a greater reliance on clinical presentation and biopsy findings due to non-classic or absent serologic markers.

We present the case of a 42-year-old male with no prior medical history who presented with unilateral leg swelling and was found to have an unprovoked deep vein thrombosis (DVT) and acute kidney injury. Initial workup revealed a creatinine level of 1.9 mg/dL and a negative ANA. At outpatient follow-up, creatinine rose to 4.1 mg/dL, prompting readmission. Urinalysis showed hematuria and 9 g/24h proteinuria. Serologic testing, including ANA and ANCA, remained negative. He was discharged on a steroid taper and antihypertensives. The patient was re-admitted a week later for worsening renal function (Cr >6 mg/dL) and symptoms including foamy urine, chronic nocturia, and fatigue. Anti-dsDNA was found to be positive, but ANA, ANCA, and complement levels remained unremarkable. A kidney biopsy revealed diffuse proliferative (Class IV) lupus nephritis. Treatment included pulse steroids, mycophenolate mofetil, and antihypertensive therapy. An AVISE test showed pan-negative autoantibodies. The patient responded well to treatment, with improving renal function and symptoms on follow-up. This case underscores the diagnostic complexity of ANA-negative lupus nephritis. Potential mechanisms for ANA-negativity include technical issues, the prozone effect, immune complex binding of ANA, or urinary loss of antibodies due to significant proteinuria.

According to the 2019 ACR/EULAR SLE classification criteria, ANA positivity is required, which would exclude this patient despite clinical and histological evidence of lupus nephritis. This case highlights the importance of considering lupus nephritis in the appropriate clinical context, even in the absence of ANA, and the need for a nuanced, clinical-pathologic approach to diagnosis and management.



"Unmasking Dermatomyositis: Respiratory Muscle Weakness Mimicking Interstitial Lung Disease"

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Abstract

Introduction: Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by proximal muscle weakness and distinctive skin manifestations. A significant complication of DM is interstitial lung disease (ILD), observed in approximately 20-40% of patients. Respiratory muscle weakness, though recognized, is less frequently reported and often overlooked. This case describes a 48-year-old woman with dermatomyositis presenting with respiratory failure secondary to respiratory muscle weakness, which can be misdiagnosed as ILD.

Case presentation: A 48-year-old female with a history of hypertension, asthma, and dermatomyositis presented with worsening shortness of breath and chest tightness ongoing for two weeks. Her symptoms worsened over two weeks, unrelieved by asthma treatment. She also experienced muscle weakness, dysphagia, and difficulty performing daily activities. Three years prior, she developed DM with mechanic's hands, facial erythema, heliotrope rash, and Gottron's papules post-COVID-19 infection. Despite treatment with solu-medrol, prednisone, mycophenolate, and rituximab, she had persistent muscle weakness. Examination revealed shallow breaths, minimal chest wall movement, elevated pCO2, and low pO2, which improved with HFNC. She had erythematous skin patches, MCP and PIP joint tenderness, and proximal muscle weakness, with a strength of 4-/5 in the left lower extremity and 4+/5 in all other extremities. Despite normal chest CT findings, pulmonary function tests indicated a restrictive pattern with a total lung capacity of 750 ml and low MIF and MEF levels. Anti-Jo-1 antibodies were elevated, while other muscle enzymes were normal. The patient was discharged following symptom improvement with methotrexate 15mg weekly, pulse solu-medrol followed by prednisone 40mg, and mycophenolate 1500mg twice daily, and was asked to follow up with her rheumatologist in the outpatient setting.

Summary: This case underscores the challenges in diagnosing and managing DM with respiratory muscle weakness, often misinterpreted as ILD due to the possibility of overlapping symptoms and imaging findings. Such cases are considered less common and complicate the diagnosis due to overlapping symptoms and imaging findings. Differentiating primary lung involvement from respiratory muscle weakness necessitates a comprehensive approach, including electromyography, muscle biopsy, and pulmonary function tests. A multidisciplinary approach is vital for accurate diagnosis and effective management of patients with dermatomyositis presenting with respiratory symptoms.

Post-COVID Autoimmunity: Unraveling the Risk of New-Onset Autoimmune Diseases After SARS-CoV-2 Infection

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Abstract

Background and objectives: Emerging evidence suggests a potential link between COVID-19 infection and the development of autoimmune diseases. This meta-analysis aims to assess the pooled risk of new-onset autoimmune conditions following SARS-CoV-2 infection.

Methods: A systematic literature search was conducted across PUBMED, Cochrane Library, and Google Scholar, with five rounds of data exploration. Only full-text, english-language cohort studies published between 2019 and 2023 were included. The key autoimmune outcomes analyzed included rheumatoid arthritis, Sjögren's syndrome, systemic lupus erythematosus, cutaneous and ANCA-associated vasculitis, psoriasis, mixed connective tissue disease (MCTD), inflammatory bowel disease, polymyalgia rheumatica, sarcoidosis, spondyloarthropathies, inflammatory myopathies, systemic sclerosis, adult-onset Still's disease, celiac disease, and type 1 diabetes mellitus. Standard meta-analysis methods were employed using the random-effects model, and heterogeneity was assessed using the I2% statistics.

Results: Five studies met the inclusion criteria, analyzing a total of 11,276,180 patients. Study cohorts included: Chang et al.(5,913,210 patients; COVID-19: 888,463, control: 2,926,016), Hielman et al. (3,908,592 patients, equally divided), Lim et al. (COVID-19: 354,527, control: 6,134,940), Syed et al. (COVID-19: 458,147, control: 1,818,929), and Tesch et al.(COVID-19: 641,704, control: 1,907,992).

The analysed data revealed the following hazard ratios, CLI and I2 heterogenicity for autoimmune diseases; SLE 1.12 (CI [0.53- 2.35, I2 0.99]); RA 1.35 (CI [0.86- 2.10, I2 0.99]); Psoriasis 1.39 (CI [0.98- 1.97, I2 0.99]); Sjogrens 1.24 (CI [0.79 -1.96, I2 0.97]); PMR 1.33 (CI [0.80- 2.22, I2 0.96]); Spondyloarthropathies 1.41 (CI [0.90-2.20, I2 0.96]); ANCA Vasculitis 1.93 (CI [1.46-2.55, I2 0.66]); Cutaneous Vasculitis 1.6 (CI [0.97-2.69, I2 0.86]); MCTD 2.04 (CI [0.90-4.92, I2 0.96]); Sarcoidosis 1.50 (CI [0.85-2.64, I2 0.95]); Inflammatory Myopathies 0.91 (CI [0.65-1.28, I2 0.23]); SS 1.15 (CI [0.61-2.18, I2 0.94]); Adult-onset Stills 1.00 (CI [0.63-1.59, I2 0]); IBD 1.34 (CI [0.94-1.90, I2 0.98]); Celiac Disease 1.57 (CI [0.90-2.73, I2 0.99]); Type 1 DM 1.66 (CI [1.08-2.56, I2 0.99]).

A significant association was found between COVID-19 and increased risk of ANCA-associated vasculitis and type 1 diabetes mellitus. Cutaneous vasculitis showed an elevated risk but did not reach statistical significance. MCTD showed a near-significant increase, warranting further investigation.

Conclusion: This meta-analysis identifies an increased risk of ANCA-associated vasculitis and type 1 diabetes mellitus following COVID-19 infection. While some conditions, such as cutaneous vasculitis and MCTD, show trends toward increased risk, additional studies are necessary to confirm these associations. Further research is crucial to understand the underlying mechanisms and long-term implications of COVID-19-induced autoimmunity.

A Diagnostic Odyssey: Differentiating IgG4-Related Disease from Lupus Lymphadenitis in a Young Adult with Persistent Fever and Lymphadenopathy

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Abstract

We report a case of a 20-year-old man presenting with fever, left cervical lymphadenopathy, and facial swelling. Initial infectious workup was inconclusive. He was given antibiotics but there was worsening of symptoms. Repeat Imaging showed extensive lymphadenopathy throughout the neck, left greater than right, asymmetric enlargement of left submandibular gland and obstruction of left IJ vein.

Histopathological analysis of deep cervical lymph nodes demonstrated markedly expanded lymphocytic population, storiform fibrosis, focal necrosis, obliterative phlebitis of some of the vessels. IgG4/IgG ratio>40. The histological findings raise the possibility of IG4-related disease. However, the presence of necrosis, unilateral enlargement of submandibular gland and persistent fever posed diagnostic challenges, as these are atypical for IgG4-RD.
As the initial biopsy was inconclusive, patient underwent a second biopsy which showed histiocytic necrotizing lymphadenitis with differential of Kikuchi disease and lupus lymphadenitis. These findings contrasted with the findings of initial biopsy. Further workup showed positive ANA 1:1280, ENA with positive dsDNA, RNP and chromatin antibodies.
In the setting of positive histology and serologies, patient was started on hydroxychloroquine and prednisone for possible lupus lymphadenitis, though patient had no typical features of lupus.

On subsequent clinic visits, he had improvement of symptoms. He was tapered off prednisone and was continued on hydroxychloroquine.

A few months later, he was evaluated again in the ED for fevers. Complement levels were normal with negative dsDNA. Infectious workup was largely negative except for a possible finding of left pyelonephritis with phlegmon development. Patient underwent kidney biopsy which showed highly fibrotic cortical tissue with mild inflammation, with 15 cells/HPF of IgG4 plasma cells and the IgG4/IgG ratio of 50%, consistent with IgG4 related disease.
Patient was started on prednisone 40 mg daily for a working diagnosis of IgG4- related disease and was discharged with plan for further outpatient management.

This case underscores the complexities in diagnosing IgG4-RD, particularly when clinical and histological features overlap with other autoimmune conditions. It highlights the importance of integrating clinical, serological, radiological, and histopathological data in establishing a diagnosis. Furthermore, it emphasizes the need for heightened awareness of IgG4-RD's diverse presentations and the potential for atypical manifestations.

From voice to limbs: unmasking the clinical clues of anti-NXP2 Dermatomyositis

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Abstract

Introduction: Anti-NXP-2 dermatomyositis (DM) is a rare subtype of idiopathic inflammatory myopathies. It accounts for about 3% of myositis cases and presents with severe generalized muscle weakness, edema, dysphagia, and an increased risk of malignancy.

Case Description: 55-year-old woman with hypertension presented with progressive generalized limb swelling, persistent hoarseness and dysphagia over several months. Her symptoms began with bilateral upper extremity pain and swelling, progressing to her lower limbs. She visited the emergency department multiple times, where initial workups revealed mildly elevated creatine kinase (CK) and ESR, with imaging studies (CTA of the neck, arms, and chest, venous duplex ultrasound) showing no acute pathology. Upon admission, she had profound diffuse limb swelling with associated weakness, more pronounced proximally than distally, and markedly elevated CK (5,888 U/L) level. Hyperpigmentation was noted on her elbows and hands. MRI confirmed diffuse myositis, and EMG findings supported an inflammatory myopathy. Bedside laryngoscopy showed extreme swelling of the base of her tongue limiting visualization of the larynx. Muscle biopsy, done after receiving immunosuppressive therapy, was inconclusive. Autoimmune workup showed a high ANA titer (1:640), elevated rheumatoid factor, and positive NXP-2 antibodies. IVIG and mycophenolate were initiated. A malignancy screen, including CT scans of the chest, abdomen, and pelvis, was unremarkable. She was discharged on a prednisone taper and mycophenolate, showing initial improvement. However, within weeks, she experienced a relapse with worsening dysphagia, necessitating PEG tube placement and multiple readmissions. Her condition deteriorated, and she passed away 15 months after diagnosis due to a major aspiration event.

Discussion: Anti-NXP2 dermatomyositis is a rare but aggressive form of inflammatory myopathy, often presenting with unusual features that delay prompt diagnosis. This patient exhibited severe edema and dysphonia—both emerging indicators of poor prognosis. Edema reflects severe inflammation and muscle damage, while dysphonia suggests oropharyngeal involvement, and higher aspiration risk, a major cause of mortality. Early recognition of these symptoms is crucial to initiate timely interventions such as prophylactic feeding tube placement. Anti-NXP2 positivity typically signals a more severe, treatment-resistant course often necessitating multiple immunosuppressive agents. Despite aggressive management, our patient's disease progressed, highlighting its refractory nature.

Conclusion: Our patient displayed atypical symptoms of DM, causing delays in diagnosis and treatment. Dysphonia and edema indicate a worse prognosis. Anti-NXP2 antibodies are linked to a severe form of DM, requiring multiple treatment modalities. Early recognition of these atypical features is essential for prompt diagnosis and effective treatment

When Crystals Target the Spine: An Uncommon Manifestation of Pseudogout

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Abstract

Introduction
Calcium pyrophosphate dihydrate deposition (CPPD), also known as pseudogout, results from calcium pyrophosphate dihydrate crystals depositions in various joints, most commonly found in the knees and wrists. Under the microscope, positively birefringent rhomboid shaped crystals are examined with polarized light. Although CPPD is common, affecting 4 to 7% of adults over 60 and mostly males, CPPD in the spine is uncommon..

We present a case of a patient presenting with low back pain and radiculopathy with positive inflammatory markers, initially concerning for infection or malignancy, but noted to have CPPD in a lumbar biopsy pathology.

Case Presentation
A 60 year old female with a past medical history of motor vehicle accident presented to the emergency department for unresolvable low back pain. Approximately a month ago, she began exercising due to elevated hemoglobin A1c levels; the following day, she had low back pain radiating to her bilateral lower flanks, along with paresthesia in bilateral lower extremities worse with rest. She endorsed recurrent visits to the emergency room and her primary care provider and had minimal improvement with Toradol and Valium leading to her hospitalization. At hospitalization, her CRP was elevated (56), and lumbar spine MRI showed spondylolisthesis, spinal stenosis at L4/5 and inflammatory changes concerning for possible infection. Infectious disease was consulted and lumbar biopsy with culture pursued to rule out infection or neoplasm. Biopsy of the L4/5 facet joint was significant for basophilic crystalline deposits seen in CPPD. Patient was discharged to an inpatient rehab facility and was started on prednisone.

Discussion
Spinal CPPD, though uncommon, is increasingly recognized in elderly patients presenting with back pain and neurologic symptoms. A systematic review reported lumbar involvement in up to 24% of patients with peripheral CPPD, though it remains underdiagnosed due to nonspecific imaging findings and clinical overlap with infection or degenerative spine disease. In this case, elevated inflammatory markers and MRI findings prompted a biopsy, which confirmed CPPD. Advanced imaging modalities may show inflammatory changes; however, histologic confirmation remains the diagnostic gold standard.

Conclusion
Low back pain is a common presentation in elderly patients. Although uncommon, it is recommended to consider CPPD in the lumbar spine that is not improving with standard treatment, as it can often be misdiagnosed or underdiagnosed. References

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Unmasking the Skeletal Impostor: Hypophosphatasia Masquerading as Fibromyalgia and Osteoporosis in Rheumatoid Arthritis

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Abstract

Hypophosphatasia (HPP) is a rare inherited metabolic bone disorder caused by mutations in the ALPL gene, resulting in low alkaline phosphatase (ALP) activity and impaired bone mineralization. With an estimated prevalence of 1:100,000 for severe forms, HPP often presents with a wide spectrum of nonspecific symptoms that can mimic fibromyalgia (FMS) or osteoporosis, leading to underdiagnosis, delayed diagnosis, or inappropriate treatment. Clinical features of HPP vary by severity and age of onset and include recurrent fractures, osteomalacia, chronic musculoskeletal pain, premature loss of teeth, chondrocalcinosis, pseudogout attacks, respiratory complications in severe forms, and ectopic calcification of ligaments.

We report a 67-year-old female with seropositive rheumatoid arthritis (RA), osteoporosis with multiple fractures, fibromyalgia, and poor dentition. Despite RA remission with golimumab and methotrexate and prior osteoporosis treatment with bisphosphonates, she experienced persistent bone pain and recurrent pathological fractures of the bilateral feet requiring multiple surgical interventions. Laboratory evaluations revealed consistently low ALP levels (24–39 U/L) and elevated fasting vitamin B6 (59.6 µg/L). Genetic testing confirmed a heterozygous ALPL mutation, consistent with the diagnosis of HPP. Enzyme replacement therapy with asfotase alfa was initiated.

This case underscores the diagnostic complexity of HPP, which can present with chronic pain resembling FMS or pathological fractures attributed to osteoporosis due to overlapping clinical features. Diagnostic criteria include persistently low ALP levels, elevated PLP, and urinary PEA levels, which are sensitive biomarkers for HPP diagnosis and treatment monitoring. Genetic testing plays a pivotal role in confirming the diagnosis by identifying pathogenic mutations in the ALPL gene. Importantly, bisphosphonates should be avoided in HPP due to their potential to worsen bone fragility. Enzyme replacement therapy with asfotase alfa has been shown to significantly improve skeletal mineralization, reduce fracture risk, alleviate musculoskeletal pain, enhance functional mobility, and improve survival rates in severe forms of HPP.

This case highlights the importance of considering HPP in the differential diagnosis of patients with unexplained fractures, low ALP levels, or musculoskeletal pain misattributed to FMS or osteoporosis. Early recognition through laboratory markers and genetic testing followed by treatment with enzyme replacement therapy can significantly improve patient outcomes while preventing further skeletal complications.

Improved Outcomes of Acute Myocardial Infarction Among Patients with Rheumatoid Arthritis: A Propensity Matched National Study

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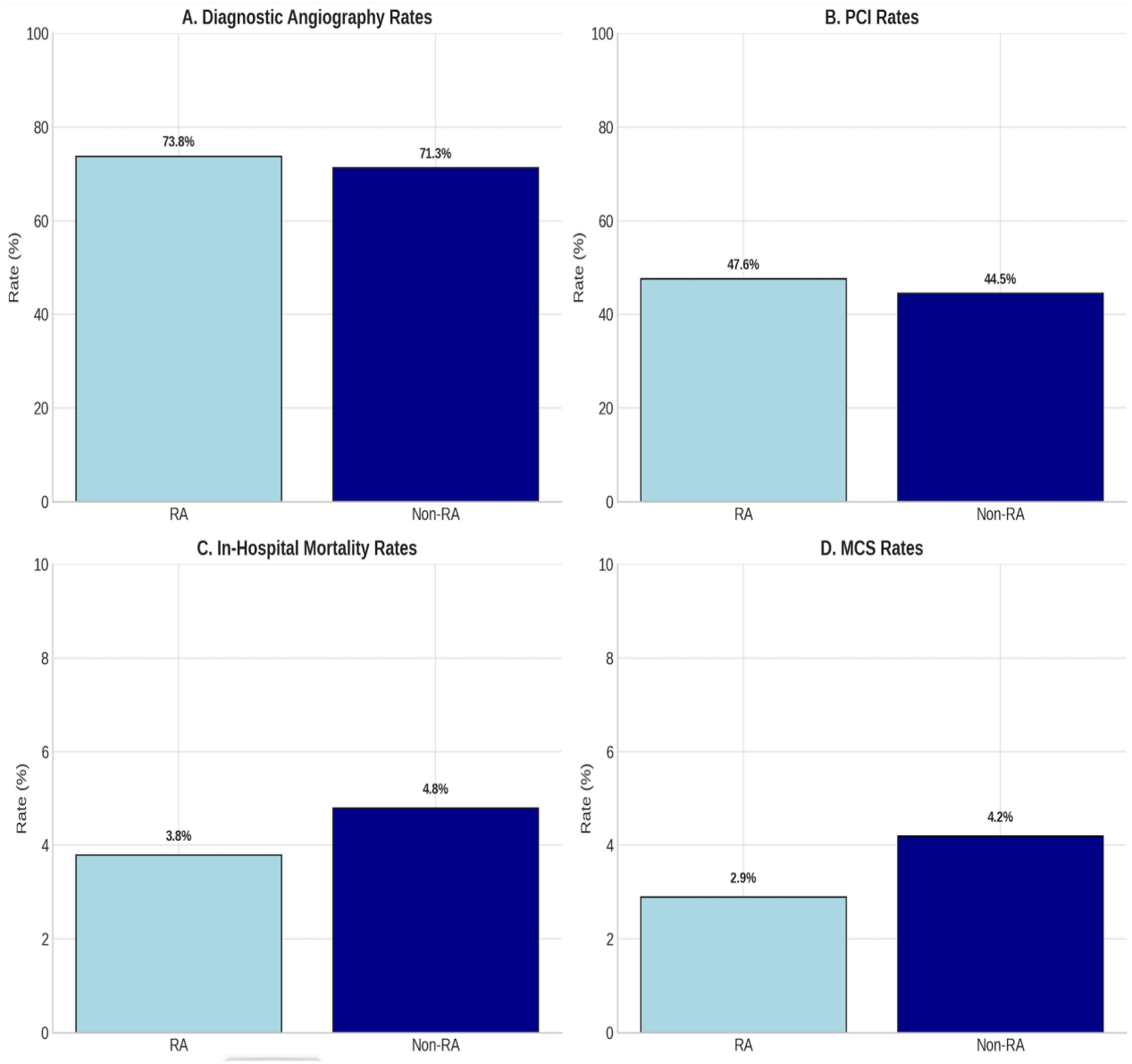
Abstract

Methods:
This retrospective population-based study investigated in-hospital outcomes among adult patients admitted with acute myocardial infarction (AMI) between January 1, 2018 and December 31, 2021, with a specific focus on the impact of rheumatoid arthritis (RA) on these outcomes. Data were derived from the National Inpatient Sample (NIS) database. The primary endpoint of the study was in-hospital mortality, with secondary outcomes including the utilization of diagnostic angiography, rates of PCI, and the incidence of complications such as AKI, cardiogenic shock, cardiac arrest, ventricular tachycardia, ventricular fibrillation, as well as requirements for mechanical ventilation, tracheal intubation, and mechanical circulatory support (MCS). Additional parameters examined were length of stay and total hospitalization charges.

The study population comprised of 2,275,150 patients, predominantly Caucasian (71%), followed by African American patients (11%), with females representing 36.4% of the cohort. The mean age across the population was 66.5 years (±2.8 years). Among these patients, 39,270 (1.7%) had a documented history of rheumatoid arthritis. Owing to the significant difference in sample size, a propensity score matching (PSM) algorithm was employed to generate comparable groups. Post-matching, 15,703 patients from each cohort were selected, ensuring a similar distribution of age, sex, race, and Charlson Comorbidity Index score.

Results:
Comparative analysis of the matched cohorts revealed notable differences in outcomes. Patients with RA experienced a lower rate of in-hospital mortality (3.8% vs. 4.8%, $P < 0.001$) compared to those without RA. Additionally, the utilization of mechanical circulatory support was lower in the RA group (2.9% vs. 4.2%). RA cohort demonstrated higher rates of diagnostic angiography (74% vs. 71%, $P < 0.001$) and PCI (48% vs. 44%), suggesting a more aggressive diagnostic and interventional approach.

Conclusion:
The study highlights that despite previous association of rheumatoid arthritis with increased cardiovascular mortality, advancements in therapeutic interventions may have contributed to improved in-hospital outcomes in this subgroup. The observed higher utilization of diagnostic and interventional procedures in the RA group may reflect heightened clinical vigilance and a lower threshold for invasive testing in this high-risk population. Overall, these findings underscore the importance of continued research as well as patient and physician education to optimize outcomes.


A. Diagnostic Angiography Rates
Rate (%)
100
80
60
40
20
0
73.8%
71.3%
RA
Non-RA
B. PCI Rates
Rate (%)
100
80
60
40
20
0
47.6%
44.5%
RA
Non-RA
C. In-Hospital Mortality Rates
Rate (%)
10
8
6
4
2
0
3.8%
4.8%
RA
Non-RA
D. MCS Rates
Rate (%)
10
8
6
4
2
0
2.9%
4.2%
RA
Non-RA

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When the Eye Deceives: Unmasking a unique case of Orbital Inflammation

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Abstract

Introduction:

IgG4 related disease (IgG4-RD) is a multi-organ fibro-inflammatory condition of unknown etiology and distinct histopathology (1) It most commonly involves the pancreas, kidneys, orbital adnexa and retroperitoneum. (2) The exact prevalence is unknown, estimates suggest 0.78–1.39 per 100,000 person-years in the US using claims-based data (3-4). Elevated serum IgG4 levels are often observed, along with elevated IgE levels in 40% of cases (5) Primary goal of management is to reduce disease activity and prevent irreversible damage. If left untreated, it may spread to other organs over time. Globally, glucocorticoids are used for induction followed by DMARDS for maintenance therapy (6)
Case history

54-year-old male presented with orbital swelling for 5 years. Initially evaluated in an allergy clinic, did not have improvement with antihistamines. Interestingly, family history was positive for similar findings in father which later caused blindness. CT scan showed proptosis and orbital edema, raising concern for thyroid related orbitopathy, however thyroid function tests were negative. Given the CT findings suggestive of myositis and pseudotumor, ophthalmology started prednisone 60 mg and referred the patient to rheumatology. Lab tests showed elevated IgG4 (363), with normal ANA, TSH, and TSI. He was started on Cellcept 500 mg twice daily and a slow taper of steroids. The patient showed improvement, and after receiving a Rituxan infusion, the swelling significantly reduced. Steroids were successfully tapered off while increasing the dose of Cellcept.
Discussion:

Described in 2001, IgG4-RD has diverse manifestations, making its true prevalence difficult to estimate. [7] Orbital involvement usually presents with proptosis, pain, and vision changes (8) Atopic conditions may be associated with IgG4-RD in head and neck areas, which could delay diagnosis, as seen in our case.

Diagnosis requires correlation of clinical, lab, and imaging findings. Biopsy is confirmatory, however not always feasible. IgG4 levels are helpful for both diagnosis and disease monitoring. While glucocorticoids are first line however the side effects limit long-term use and disease flares are common after discontinuation (6) The clinical response to rituximab in IgG4-RD patients is often rapid, resulting in significant improvement in disease activity.

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An enlarging tender skin depression in a young adult female

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Abstract

Introduction:
Idiopathic localized involutional lipoatrophy (ILIL) is a rare condition characterized by a focal fat loss without significant inflammation or an identifiable etiology. Due to limited reported cases, its clinical course and optimal management remain poorly defined. We present a case of ILIL in a young woman with rheumatoid arthritis (RA).

Case:
A 22-year-old female with RA, treated with etanercept and hydroxychloroquine, presented with a tender, enlarging depression on her right lower back. She denied trauma or injections to the area. A previous biopsy, initially read as morphea, prompted ineffective treatment with tacrolimus and topical steroids. Examination showed a 5 cm x 5 cm violaceous, atrophic plaque. ANA, ENA10, and dsDNA were negative. Re-evaluation of the biopsy showed peri-eccrine fat atrophy without collagen hyalinization. A second biopsy confirmed lipoatrophy without panniculitis or morphea; CD68-positive macrophages were present.

Three months later, the lesion expanded with central tenderness. She also reported a 30-pound weight loss and worsening joint pain. A new area of tenderness on the left lower back was noted. A third biopsy was similar. Though mycophenolate mofetil and tacrolimus therapy was planned, the lesion spontaneously regressed within a month.

Discussion:
Lipodystrophies are classified as generalized, partial, or localized, and are typically caused by trauma, injections, or autoimmune inflammation. ILIL, however, lacks identifiable triggers. It most often affects the buttocks and proximal limbs, with histology showing fat cell loss and minimal inflammation. CD68-positive macrophages may play a role in adipocyte involution. While spontaneous improvement is common, treatments like calcineurin inhibitors, antimalarials, or steroids may be used in persistent cases.

This case aligns with literature suggesting a female predominance in localized lipoatrophies, potentially due to hormonal or genetic factors. Females may also face higher metabolic risks in some lipodystrophies.

Similarly, it is unclear whether our patient's history of RA was contributory. Autoimmune conditions have been linked to other acquired generalized and localized lipodystrophies. It is plausible that an immunological response to adipocyte antigens leads to macrophage infiltration, cytokine release, and adipocyte destruction.To our knowledge, this is the first case to describe a patient with RA and ILIL.

ILIL is rare, however likely under-reported given its frequently self-limited nature. Additional studies are required to better understand the risk factors, clinical spectrum, and management

strategies for ILIL. Earlier recognition may prevent unnecessary invasive tests and therapies.

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Figures:

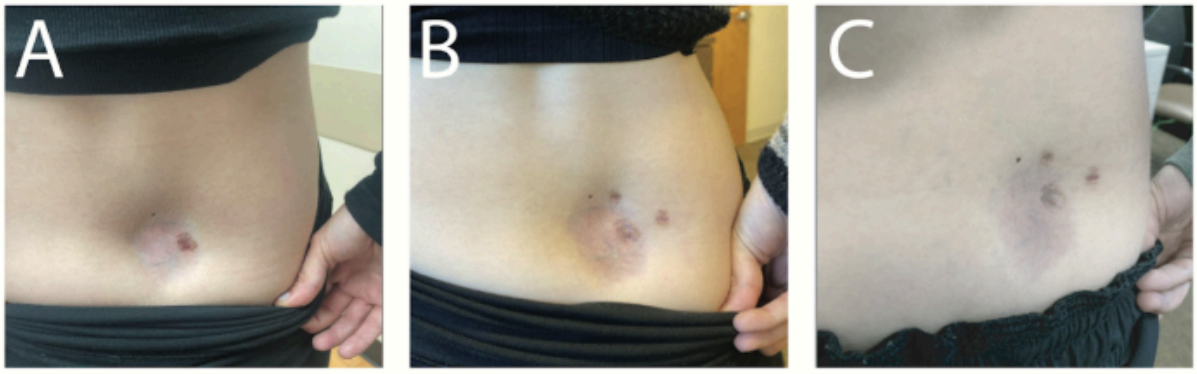

Figure 1. Well-circumscribed, atrophic depression measuring 5cm x 5cm at her initial visit (A) and 5.5cm x 6cm at subsequent visit 3 months later (B). (C) demonstrates improvement in cutaneous depression 4 months after initial visit.

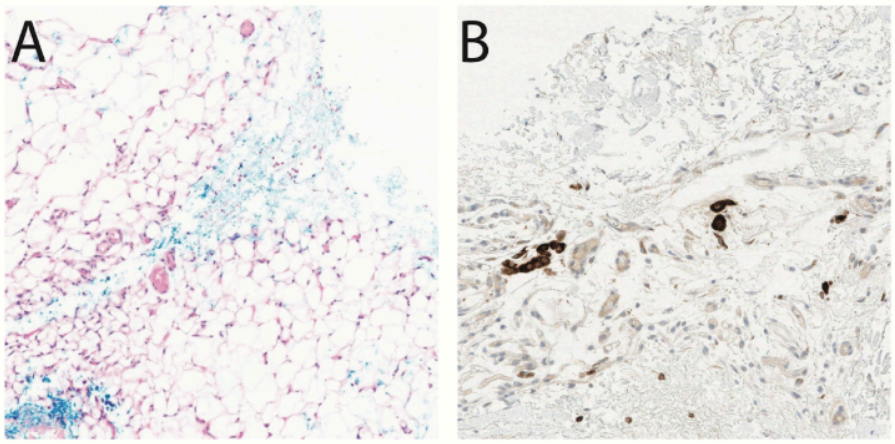

Figure 2. A representative biopsy revealing lipoatrophy without active panniculitis or morphea (A). Immunohistochemistry revealed CD-68 positive cells (brown stain, B).

A Rare Case of Linear Atrophoderma: Clinical Presentation

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Abstract

Introduction:
Linear atrophoderma of Moulin (LAM) is a rare disease defined by hyperpigmented, atrophic lesions on the skin that typically follow Blaschko's lines, often found in children or adolescents. [1] There is a lack of evidence to determine the etiology and treatment for LAM.[2] We present a rare case of a 19-year-old male with both unilateral and bilateral linear atrophoderma lesions.

Case Summary:
This 19-year-old white male had skin changes at the age of 16. These manifested as patches of pigmented lesions on various parts of the body, including both posterior shoulders, torso, abdomen, right arm, back, and extending down to the hips.
There was no history of other diseases (besides growth hormone deficiency), or similar family history. They lesions were described as: "hyperpigmented atrophic plaques to the trunks seemingly blaschkoid in some regions."
The biopsy showed: "dermal sclerosis with dermal thickening and perieccrine entrapment and mild epidermal papillomatosis with basilar hyperpigmentation. The microscopic features are not entirely specific. However, in the context of linear atrophic hyperpigmented patches, these changes could potentially represent linear atrophoderma (of Moulin), or related entities."
The patient was diagnosed with LAM and treated with methotrexate (MTX). The lesions have regressed.

Discussion:
The precise mechanism underlying the development of LAM remains uncertain, however this could be from genetic mosaicism or positive serological testing. [3] The disease often stops progressing and occasionally regresses with time.[5]
There are approximately 30 cases of LAM[8] that have been described in the literature.[9] [2] In this case, the patient presents with a unique pattern, exhibiting areas of both unilateral and bilateral involvement and dermal sclerosis with dermal thickening.[5]
Unfortunately, there is no standard treatment for LAM. Some cases reported had been given MTX, penicillin, potassium aminobenzoate, or topical steroids. [11]
We believe that there is more to be determined about LAM.[12] We now see patients present with features that vary from what was originally thought to define the disease. The etiology and causality have yet to be determined. Rheumatologists should be aware of this rare condition as this could be a great mimicker of other cutaneous rheumatic diseases.

Takayasu Arteritis and Pulmonary TB Reactivation: A Clinical Crossroads

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Abstract
Background:

Takayasu arteritis (TAK) is a rare granulomatous vasculitis of the aorta and its major branches, often affecting women under 40, particularly of Asian descent. Immunosuppressive therapy that is required to manage TAK raises the risk of latent tuberculosis (TB) reactivation, especially in patients from endemic regions. Recent research also suggests a potential immunopathogenic link between *Mycobacterium tuberculosis* (MTB) and TAK, with implications for diagnosis, treatment, and disease pathogenesis.
Case Presentation:

A 50-year-old Indian-born woman presented with fever, productive cough, dyspnea, and chills after failing multiple outpatient antibiotic courses. She had a history of TAK diagnosed a year earlier, treated with prednisone, methotrexate, and infliximab. Past medical history included diabetes, hypothyroidism, thalassemia minor, and severe aortic regurgitation under evaluation for TAVR. She had recently returned from volunteer work in India with underserved populations. Physical examination revealed exophthalmos, thyromegaly, a decrescendo diastolic murmur, and diminished peripheral pulses. Lab findings included microcytic anemia and elevated CRP. Imaging showed bilateral pulmonary nodules and CT angiography demonstrated occlusion of bilateral subclavian arteries and signs consistent with chronic large-vessel vasculitis and arterial inflammation.

Despite a negative Quantiferon test, which was likely secondary to immunosuppression, bronchial AFB smears confirmed reactivation of pulmonary TB. Her hospital course was complicated by septic shock, respiratory failure requiring intubation and tracheostomy, and acute renal failure requiring temporary dialysis. After two months of inpatient care and RIPE therapy, she stabilized and was discharged with plans for TAVR.

Infectious Workup:

Procalcitonin	1.37*
Coccidioides AB IGG	Positive
Cryptococcus Antigen	Negative
Coccidioides immitis AB Titer	negative
Mycoplasma pneumoniae AGM	525 WNL
Streptococcus pneumoniae AG, Urine	Negative
Legionella Pneumophila Ag	Negative
SARS-COV-2 Nucleocapsid ANTIBODY	Positive
TB Gamma Interferon	Negative
TB Ag stimulated	M. Tuberculosis infection NOT likely
MRSA Nares	Negative
Parainfluenza Virus RNA PCR	Detected!

CMV IGG	>8.0*H
CMV DNA Viral Load, PCR	1,540.00*H
CMV DNA, PCR LOG	3.19*H
EBV PCR	<200 copies
Nocardia culture	Neg
Actinomyces	Neg
Fungal Culture	Neg
Blood culture	NGTD

Discussion:

This case illustrates the critical importance of aggressive TB screening in immunosuppressed TAK patients, particularly those from endemic areas. Latent TB tests may yield false negatives in immunocompromised individuals, supporting the case for empiric treatment in high-risk scenarios. Notably, MTB-specific proteins such as HSP65 and gene sequences (IS6110, HupB) have been identified in arterial tissue of TAK patients, suggesting a possible role for MTB in TAK pathogenesis.

Conclusion:

Managing TAK requires a delicate balance between immunosuppression and infection prevention. Clinicians should adopt a globally informed, multidisciplinary approach to care, including proactive TB screening and consideration of empiric treatment in high-risk patients.

Abstracts Published

Compounding: a Glimpse inside the Black Box

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Abstract

Objectives

Drug compounding is the combining, mixing, or alteration of active pharmaceutical ingredients (API) into doses or dosage forms for individual patients. While often prescribed by rheumatologists, the methodology behind compound pricing by pharmacies remains unclear to most physicians. In this study, we reviewed the pricing for one of the most prescribed compounds: transdermal diclofenac 10% in PLO.

Methods

The constituent costs of a drug compound were determined by interviewing members of the Alberta Pharmacist's Association.

API and excipient ingredient costs were determined by reviewing listed wholesaler prices. Published insurance plan information was used for dispensing fees and insurer permitted mark-ups.[1] These mark-ups are at the pharmacy's discretion if a patient pays cash. Price quotes were attained by calling pharmacies across Alberta. Independent, chain, and compounding pharmacies were called to ensure representation from the different pharmacy types. Only one pharmacy from each brand/banner was called.

Results

Table 1A. Quoted cost of 100g of Diclofenac 10%

	Pharmacy type	Cost for 100g	Made on site
Benchmark Lowest Pharmacy Quote	-	$33.55	Yes
Pharmacy 1	Chain	$55.82	No
Pharmacy 2	Chain	$45.20	No
Pharmacy 3	Chain	$70.00	No
Pharmacy 4	Chain	$43.28	No
Pharmacy 5	Chain	$38.42	No
Pharmacy 6	Compounding (Independent)	$205.00	Yes
Pharmacy 7	Compounding (Independent)	$87.35	Yes
Pharmacy 8	Compounding (Independent)	$37.83	Yes
Pharmacy 9	Compounding (Chain)	$58.93	Yes
Pharmacy 10	Compounding (chain)	$52.67	Yes
Pharmacy 11	Independent	$170.00	No
Pharmacy 12	Independent	$55.75	No
Pharmacy 13	Independent	$80.71	No
Pharmacy 14	Independent	$56.69	No
Pharmacy 15	Independent	$110.00	No

Table 1B. Costs associated with this compound. API and excipient costs

	Wholesaler	Pack size	Pack Size Cost	Cost for amount in 100g of Diclo % in PLO
Diclofenac1	Wholesaler 1	100g	$49	$4.90
Diclofenac	Wholesaler 1	1kg	$330	$3.30
Diclofenac	Wholesaler 1	25kg	$7300	$2.92
PLO2	Wholesaler 1	500g	$77	$13.86
PLO	Wholesaler 1	20kg	$2300	$10.35
Diclofenac sodium USP	Wholesaler 2	100g	$172.90	$17.29
Diclofenac sodium USP	Wholesaler 2	25kg	$7628.50	$3.05
PLO	Wholesaler 2	500g	$96	$17.28
PLO	Wholesaler 2	100kg	$9628	$8.67

1. 10g is the quantity of diclofenac in 100g of Diclofenac 10% in PLO
2. 90g is the quantity of PLO in 100g of Diclofenac 10% in PLO

Container cost

	Unit Cost
White container	$0.12-$2.19
Tube	$0.78-$5.66
Dispensing device	$0.90-$8.50

Staff Cost

	Wage/hr (ALIS)
Pharmacist	$53.18
Pharmacy Technician	$33.12
Time to Make 100g cream/lotion/ointment	
Insurer Compensated Time	6 minutes[2]
-Cost to compound if made by pharmacist	$5.32
-Cost to compound if made by pharmacy tech	$3.31

Fees

Dispensing Fee (compound made in store)	$18.45
Mark-up (compound made in store)	Insurance: ((Base cost * 1.075)*1.07) Cash: At discretion of pharmacy
Dispensing Fee (repackaged compound)	$12.15
Mark-up (repackaged compound)	Insurance: No mark-up Cash: At discretion of pharmacy

A comprehensive overview of price constituents for diclofenac 10% in PLO is seen in Figure 1. Using the information available, we determined the theoretical lowest cost for this compound was $31.78. Contextually, the cost per 10g of diclofenac (amount needed to make compound) ranged from $2.92-$17.92 The cost per 90g of PLO (amount needed to make compound) ranged from $8.67-$13.86. Mark-up and dispensing was the largest contributor to final price. The real price varied considerably by pharmacy. The average price for 100g of 10% diclofenac was $50.54 (range: 38.42-55.82) at chains, $88.36 (range: 37.83-205.00) at compounding pharmacies, and $94.63 (range: 55.75-170.00) at independent pharmacies.

Conclusions

A wide range of prices for the same compound were determined.

Several factors contribute to the large price variations. They most important be summarized as follows:

1. Differing acquisition costs from wholesalers for the active pharmaceutical ingredient and excipients (diclofenac, PLO),

2. Mark-ups, which can be changed at the pharmacy's discretion for patients without insurance

3. Dispensing fees

4. Hidden repackaging costs (charges associated with a non-compounding pharmacy acquiring compounds from another pharmacy)

It is important to note that items 1-3 provide profit to the pharmacy after covering labour and material costs.

These findings have important implications for prescribing rheumatologists and patients without adequate insurance who may be exposed to elevated prices without their knowledge. A range of price from $33.55 to $205.00 a product costing $31.78 to make highlights the need for greater price transparency. Rheumatologists are well-situated as prescribers to advocate on behalf of patients.

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Immune-Mediated Necrotizing Myopathy Triggered by Semaglutide on a Background of Statin Use

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Abstract

Introduction: Immune-mediated necrotizing myopathy (IMNM) is a subgroup of the idiopathic inflammatory myopathies, commonly resulting from statin use. There have been no reported cases of semaglutide-triggered IMNM.

Case: 77-year-old Caucasian male with a history of HTN, DM, and CAD presented to the hospital for progressively worsening proximal muscle weakness involving bilateral upper and lower extremities. He also reported dysphagia, dyspnea, and weight loss of 25 lbs in the past 3 months. He was previously seen at his PCPs office for myalgias 4 weeks after starting semaglutide and was found to have new elevation of liver enzymes >10x upper limit of normal (AST 682, ALT 735) with a CK of 9363. Further workup ruled out primary liver disease. His other medications were aspirin, metoprolol, metformin, and atorvastatin, which he had been taking for 4 years without adverse effects. Physical exam revealed atrophy of triceps muscles, symmetric weakness in proximal muscles (strength 3/5 on shoulder abduction, 4/5 on shoulder adduction, 2/5 on hip flexion, 4/5 on hip abduction), and inability to stand up unassisted. There were no rashes, periungual erythema, mucosal lesions, or synovitis and nailfold capillaroscopy was normal. Labs were significant for elevated CK of 3762, transaminases (AST 354, ALT 135), LDH 565, and normal renal function. EMG showed findings suggestive of inflammatory myopathy. Muscle biopsy confirmed necrotizing myopathy consistent with anti-HMGCR associated IMNM, with elevated serum anti-HMGCR antibody (124, normal value <20). After a 3-day course of IV steroids in the hospital, he was discharged with an oral prednisone taper and started on IVIG infusions as an outpatient. He was advised to stay off of semaglutide and statins. He has had a remarkable clinical recovery and is able to perform his daily tasks independently.

Discussion: While statins are a well-known trigger for IMNM, our patient was on atorvastatin without issue for years. His myalgias occurred after initiation of semaglutide. Rhabdomyolysis, but not IMNM, associated with semaglutide has been reported in only one case report (1). The pathogenesis remains unclear, but it is possible that semaglutide potentiated muscular injury, priming the muscles against autoantibody attack.

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Influence of Modifiable and Non-Modifiable Risk Factors on In-Hospital Mortality Among Rheumatoid Arthritis Patients in the United States of America: A National Inpatient Sample Study.

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Abstract

Introduction: Rheumatoid arthritis (RA) is the most common autoimmune arthritis, affecting 17.6 million people globally in 2020, with projections of 31.7 million by 2050[1]. The mortality rate for RA patients has improved since the 1990s due to better treatment strategies [2].
Methods: This study uses data from the 2018-2020 National Inpatient Sample (NIS) database to examine the link between mortality in hospitalized RA patients. We analyzed adults aged 18 and older diagnosed with RA through ICD-10 codes, with mortality as the primary outcome assessed via logistic regression models.

Results: Mortality rates among patients with rheumatoid arthritis (RA) increase with age (odds ratio [OR] 1.04, 95% confidence interval [CI] 1.03-1.04, $p < 0.001$) and are higher for Native Americans (OR 1.39, 95% CI 1.08-1.80, $p = 0.01$). A Charlson comorbidity index score of ≥3 significantly increases mortality (OR 2.36, 95% CI 2.15-2.60, $p < 0.001$), as does having self-pay insurance (OR 1.37, 95% CI 1.08-1.74, $p = 0.01$). In contrast, individuals in middle-income (OR 0.93, 95% CI 0.86-0.99, $p = 0.04$) and high-income tiers (OR 0.92, 95% CI 0.84-0.99, $p = 0.03$) experience lower mortality. The low-income tier ($50,000-$64,999) shows increased mortality but is not statistically significant (OR 1.02, 95% CI 0.95-1.09, $p = 0.57$). Patients facing hospital costs over $100,000 had a significantly higher mortality risk (OR 4.16, 95% CI 3.88-4.47, $p < 0.001$) compared to those with costs between $50,000 and $100,000 (OR 1.67, 95% CI 1.50-1.85, $p < 0.001$). Additionally, RA patients admitted to larger hospitals had a higher risk of mortality (OR 1.20, 95% CI 1.17-1.29, $p < 0.001$) compared to those in smaller hospitals (OR 1.11, 95% CI 1.03-1.20, $p = 0.01$).

Conclusion: Our study's strength lies in the nationwide database, which reflects the U.S. population. However, it does not account for the timing of RA diagnosis or treatment methods that may affect mortality rates. We found significant disparities in RA mortality related to socioeconomic status, race, and healthcare access. Addressing these issues requires a multi-level approach focusing on clinical risk stratification, enhanced healthcare access, and socioeconomic support to improve outcomes for vulnerable RA patients.

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Pericarditis Induced by Adalimumab in a Patient with Rheumatoid Arthritis: A Case Report

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Abstract

Introduction:
Adalimumab, a tumor necrosis factor (TNF) inhibitor, is widely used in managing autoimmune conditions like rheumatoid arthritis (RA). While generally well-tolerated, rare adverse effects, including pericarditis, may occur. This case highlights pericarditis as a potential complication of adalimumab therapy, initially unrecognized as medication-induced, and its resolution upon discontinuation.

Case Presentation:
A 44-year-old female with seropositive rheumatoid arthritis on Adalimumab with irregular compliance for 10 years presented with recurrent chest pain and was diagnosed with pericarditis. Initial evaluation by cardiology included echocardiography and CT imaging, which revealed a large pericardial effusion. Despite treatment with colchicine and ibuprofen, the fluid persisted, and cardiology considered surgical intervention with pericardial stripping. The patient's history of intermittent adalimumab use and symptom onset during therapy led to the suspicion of medication-induced pericarditis. Adalimumab was discontinued, resulting in symptom resolution and normalization of follow-up imaging. RA management was subsequently transitioned to abatacept, with effective disease control and no recurrence of pericarditis.
Discussion and Conclusion:
This case highlights a rare but clinically significant adverse effect of adalimumab-induced pericarditis. While pericarditis is a known manifestation of systemic autoimmune diseases like RA, it is crucial to distinguish disease-related pericarditis from drug-induced causes, particularly in patients on long term biologic therapies. In this case, cardiology initially attributed the recurrent pericarditis to non-specific factors like smoking, hx of breast mass or viral etiology and even considered invasive surgical intervention (pericardial stripping), but symptom resolution after discontinuing adalimumab confirmed it as the cause.
The pathophysiology of TNF inhibitor-induced pericarditis remains unclear. Paradoxically, while TNF-α inhibitors suppress inflammation, they can also dysregulate immune responses in some individuals. Previous reports of adalimumab-related pericarditis emphasize the need for clinician awareness.

This case also demonstrates the importance of considering alternative therapies when adverse drug reactions occur. Abatacept, a T-cell co-stimulation inhibitor, provided effective disease control without recurrence of pericarditis. This aligns with evidence suggesting abatacept's favorable safety profile, particularly in patients with a history of cardiovascular or inflammatory complications.

Prompt recognition of medication-induced pericarditis is critical to prevent unnecessary interventions, such as invasive procedures, and to minimize patient morbidity. Comprehensive evaluation of the patient's medication history and temporal correlation between drug use and symptom onset are essential diagnostic steps. This case reinforces the need for

interdisciplinary collaboration between rheumatology and cardiology to optimize outcomes in complex cases like these.

Breaking the Heart: The Role of SLE Myocarditis in the Evolution of Dilated Cardiomyopathy

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease affecting multiple organ systems, with cardiac involvement occurring in approximately half of cases. Lupus myopericarditis (LM) though rare, is a potentially life-threatening condition with a low prevalence. It often presents late in the disease course with symptoms of congestive heart failure including dyspnea, palpitations, and arrhythmias.

A 62-year-old African American female with hypertension and hyperlipidemia initially presented with generalized weakness, weight loss, and leukopenia. Workup revealed positive ANA, anti-Sm, anti-RNP, anti-dsDNA, and a small pericardial effusion, leading to a diagnosis of systemic lupus erythematosus (SLE). Two months later, she developed worsening orthopnea, volume overload, and acute kidney injury. Echocardiography showed a reduced EF of 10-15% with severe left ventricular dilation. Cardiac catheterization ruled out ischemic cardiomyopathy, guiding us to identify dilated cardiomyopathy as a result of newly diagnosed SLE. She was transferred for advanced heart failure evaluation and started on inotropic support. Further workup revealed class III lupus nephritis, requiring mycophenolate mofetil initiation. With optimized guideline-directed medical therapy, her EF improved to 30-35%, allowing weaning off inotropes. This case highlights SLE as a rare but severe etiology of acute dilated cardiomyopathy, highlighting the importance of early recognition and multidisciplinary management.

This patient met multiple EULAR/ACR criteria for systemic lupus erythematosus (SLE), including pericardial effusion, positive dsDNA, anti-Smith, ANA, leukopenia, low C3, and biopsy-proven lupus nephritis. Cardiac involvement occurs in approximately 50% of SLE patients, but lupus myocarditis (LM) is a rare complication, seen in a small number of cases. Despite its rarity, LM can cause acute heart failure with significant prognostic implications. Risk factors include hypertension, pericarditis, low complement levels, and lupus nephritis. Diagnosing LM is challenging, with endomyocardial biopsy as the gold standard, but it is rarely performed due to its invasiveness. Cardiac MRI is the preferred noninvasive diagnostic tool, revealing myocardial necrosis or fibrosis via late gadolinium enhancement. Treatment includes guideline-directed medical therapy (GDMT) for heart failure, high-dose corticosteroids, and immunosuppressive therapy, such as hydroxychloroquine, cyclophosphamide, or mycophenolate mofetil. Severe cases may require inotropes, ventricular assist devices, or extracorporeal membrane oxygenation (ECMO). The prognosis of LM-related dilated cardiomyopathy varies, but early intervention often leads to EF recovery. Maintaining a high level of clinical suspicion is crucial for timely treatment and improved patient outcomes. While commonly seen in juvenile SLE, LM is rare in adults. It is also a rare cause of acute heart failure. Patients typically present in fulminant dilated cardiomyopathy with no other precipitating factors. Many diagnostic modalities exist; however, the gold standard includes TTE, cardiac MRI, and endomyocardial biopsy. Given the high mortality risk of SLE-associated dilated cardiomyopathy, early recognition and aggressive immunosuppressive therapy are crucial to improving outcomes. Management includes immunosuppressants, steroids, and GDMT. In

severe cases, inotropes, ventricular assist devices, or ECMO may be required. This case highlights the importance of early suspicion and multidisciplinary management in preventing irreversible cardiac damage and improving survival in lupus myopericarditis.

Rapidly progressive scleroderma with cardiac, lung, skin involvement misdiagnosed as vitiligo

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Abstract

I.B is a 21-year-old gentleman with a past medical history of newly diagnosed heart failure with reduced ejection fraction (HFrEF) and myocarditis who was admitted to an academic medical center for advanced heart failure management. One month prior, the patient was hospitalized for palpitations and was found to have HFrEF EF 25% and myocarditis. Cardiac MRI showed diffuse gadolinium enhancement and endomyocardial biopsy showed no giant cells. He was started on therapies for heart failure discharged with a wearable defibrillator. He was seen by outpatient cardiology one month later and found to be in ambulatory cardiogenic shock and was re-admitted. He was noted to have skin thickening for which rheumatology was consulted. He first noticed thickening of his skin and skin hypo/hyperpigmentation about 6-12 months prior and was diagnosed with vitiligo (limited medical records are available but show vitiligo listed in his chart four months prior to admission). Progress notes from the hospitalization one-month prior also document evidence of skin thickening on exam. He also endorsed new dysphagia, Raynaud's, digital ulcers, weight loss. Physical exam was significant for heart murmur, contractures of elbows, wrists and hands, sclerodactyly, healing digital ulcers, diffuse skin thickening across face, chest, abdomen, extremities and classic “salt and pepper” skin pigmentations across chest, extremities. Laboratory studies revealed positive ANA >1:1280 nuclear, homogenous, elevated Scl-70 (155), RNA polymerase III (30). CT chest showed bilateral emphysematous bullous disease with faint ground glass. He was diagnosed with rapidly progressive scleroderma and he was given cyclophosphamide. He had been previously started on prednisone and efforts were made to taper. He was lost to follow up for about a month and received a second dose of cyclophosphamide. Unfortunately, 2 months after his initial diagnosis, he represented to the hospital with progressive shortness of breath. He was found to have hypotension, hypoxia, massive saddle PE and had a cardiac arrest where ROSC was achieved but unable to be sustained. This case highlights the importance of recognizing and treating rapidly progressive scleroderma. It also highlights diagnostic momentum and how this patient’s skin changes were incorrectly diagnosed as vitiligo.

WHEN HISTORY REVEALS ITSELF- UNMASKING THE VASCULITIS MASQUERADER

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Abstract

We present the case of a 36-year-old with a history of untreated hypertension, type B aortic dissection, and recently diagnosed carotid and abdominal artery aneurysms, who presented to the intensive care unit (ICU) with hemorrhagic shock in the setting of recurrent hematochezia and hematemesis. His hospital course was complicated by multiple episodes of GI bleeding, culminating in the diagnosis of an aorto-biliary fistula on an initial angiography, thought to be a consequence of his prior aneurysm repair. In the process of working up his GI bleeding, he underwent a mesenteric angiography, which demonstrated diffuse arterial beading suggestive of vasculitis, prompting a rheumatologic evaluation to rule out autoimmune vasculitis. While the patient had no systemic rheumatologic symptoms, he reported a strong maternal family history of aneurysms and unexplained premature deaths. On examination, he exhibited arachnodactyly, widespread joint laxity, arm span exceeding his height (arm span to height ratio 1.08:1), and mild concavity of his central chest wall. His autoimmune serologies were negative. A comprehensive review of his medical records thereafter revealed prior genetic testing from a previous hospitalization, identifying a pathogenic transforming growth factor β receptor 1 (TGFBR1) variant consistent with Loeys-Dietz Syndrome (LDS).

LDS is a rare autosomal dominant genetic connective tissue disorder caused by heterozygous mutations in the genes encoding TGFBR1, TGFBR2, SMAD3, or TGFB2. Individuals with LDS are predisposed to widespread and aggressive arterial aneurysms that can affect large, medium, and small vessels, with vessel tortuosity and beading commonly seen on imaging. LDS is also characterized by skeletal manifestations like pectus excavatum, joint laxity, arachnodactyly, craniofacial features like hypertelorism, craniosynostosis, and cutaneous findings like easy bruising and dystrophic scars. Many of these features were seen in our patient, further solidifying the diagnosis of LDS.

Our team recommended further imaging to assess for additional vascular complications, blood pressure optimization, and genetic counseling. Since discharge, the patient has remained stable and is recovering well from his hospitalization. This case highlights the importance of recognizing genetic connective tissue disorders in patients with unexplained vascular complications and, most importantly, emphasizes the art of mindful history-taking and physical examination in clinical practice.

Autoimmune Crossroads: An Unusual Case of Coexisting Psoriasis and Systemic Lupus Erythematosus

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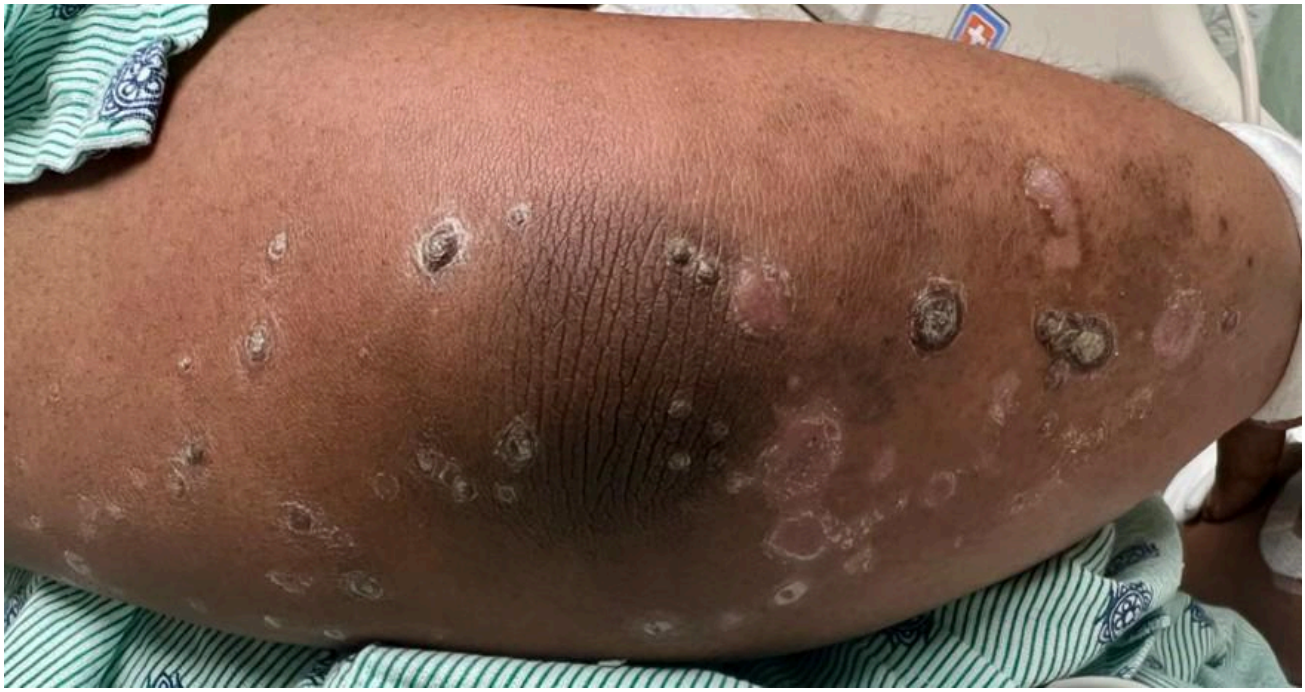
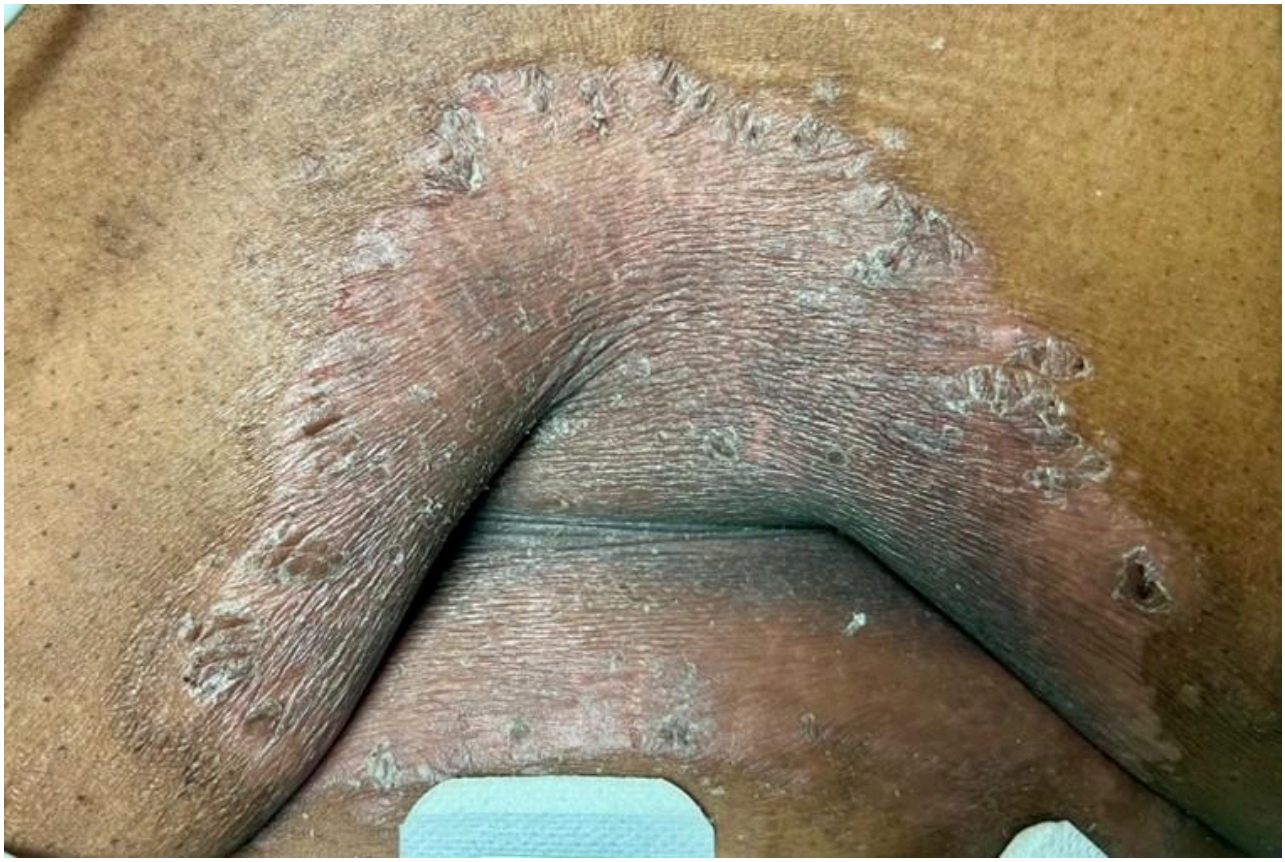
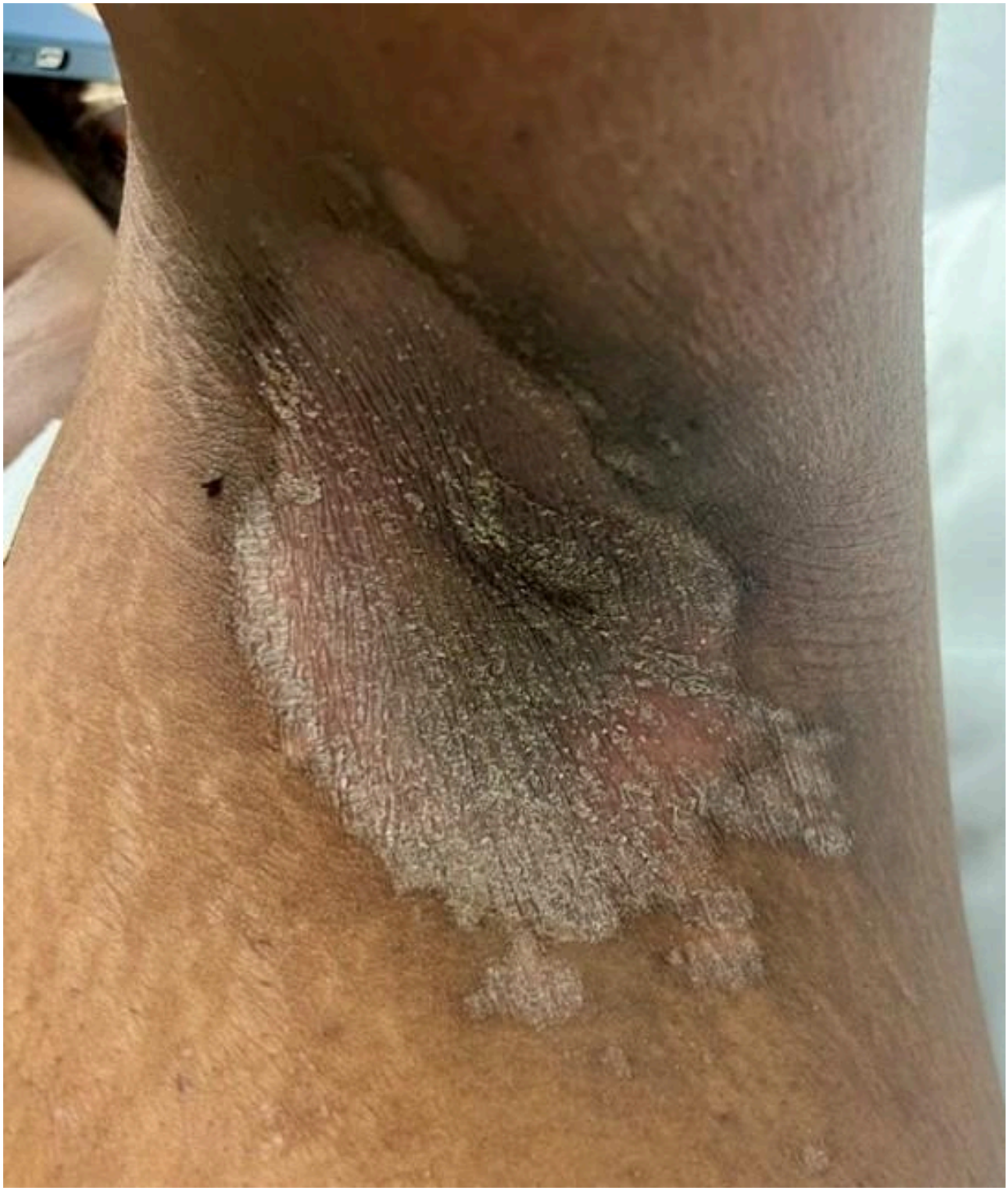
Abstract

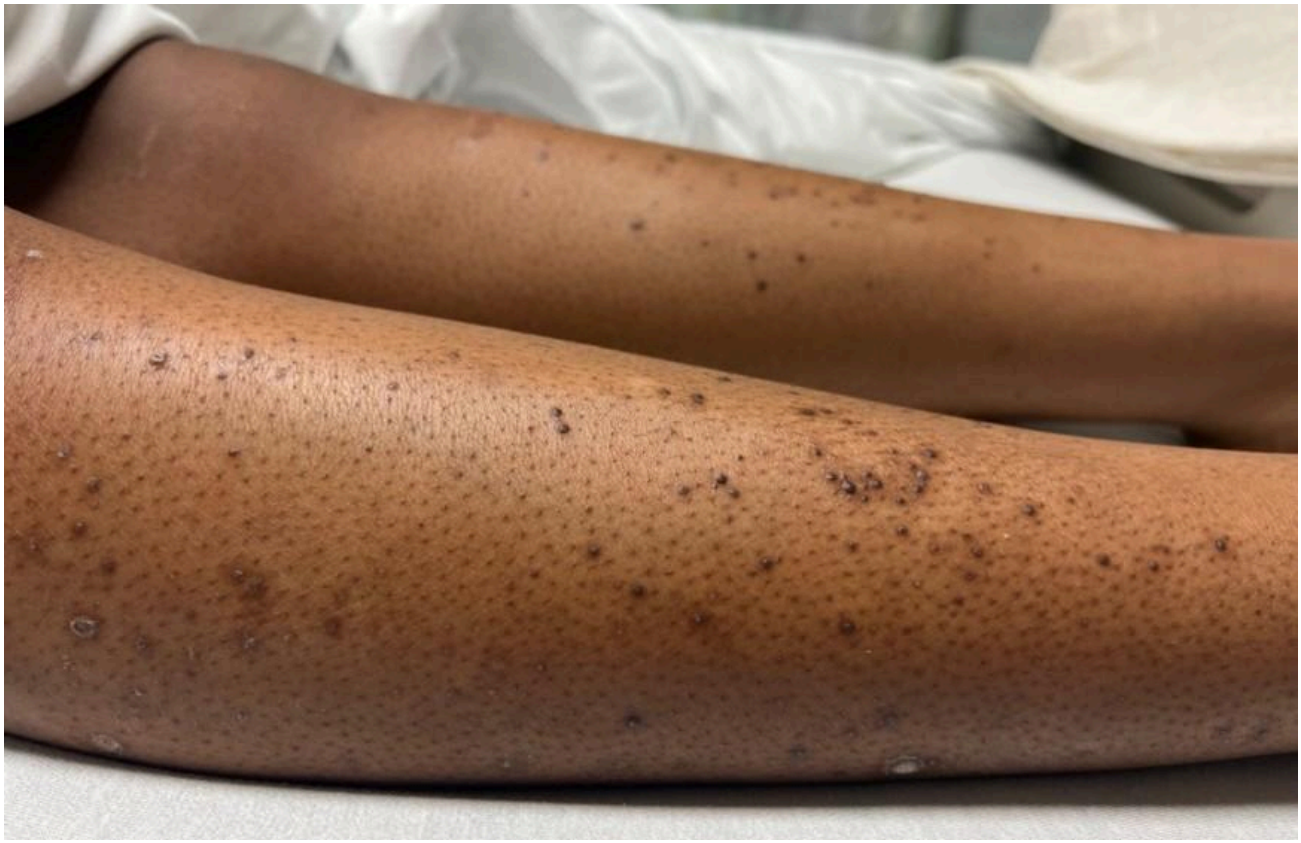
A 21-year-old female presented to the emergency department with symptoms of fever, diffuse rash, and polyarticular joint pain predominantly affecting her knees and hands. The rash initially appeared on her face and was worse with sun exposure. The rash subsequently spread to her chest, beneath her breasts, and progressed to involve her extremities, including her elbows, hands, and legs. She endorsed hair loss, but denied oral or nasal ulcers, weight loss, sicca symptoms, Raynaud's phenomenon, history of DVT/PE, or miscarriages.

Physical examination revealed tenderness upon palpation of the bilateral metacarpophalangeal joints, several proximal interphalangeal joints, and bilateral knees. There was hyperpigmentation on both cheeks, although no distinct malar rash was present. Multiple scaly, annular plaques were present on both elbows. A large hypopigmented scaly rash was observed beneath the breasts and in the bilateral axillary regions. A petechial rash was additionally identified on the lower extremities.



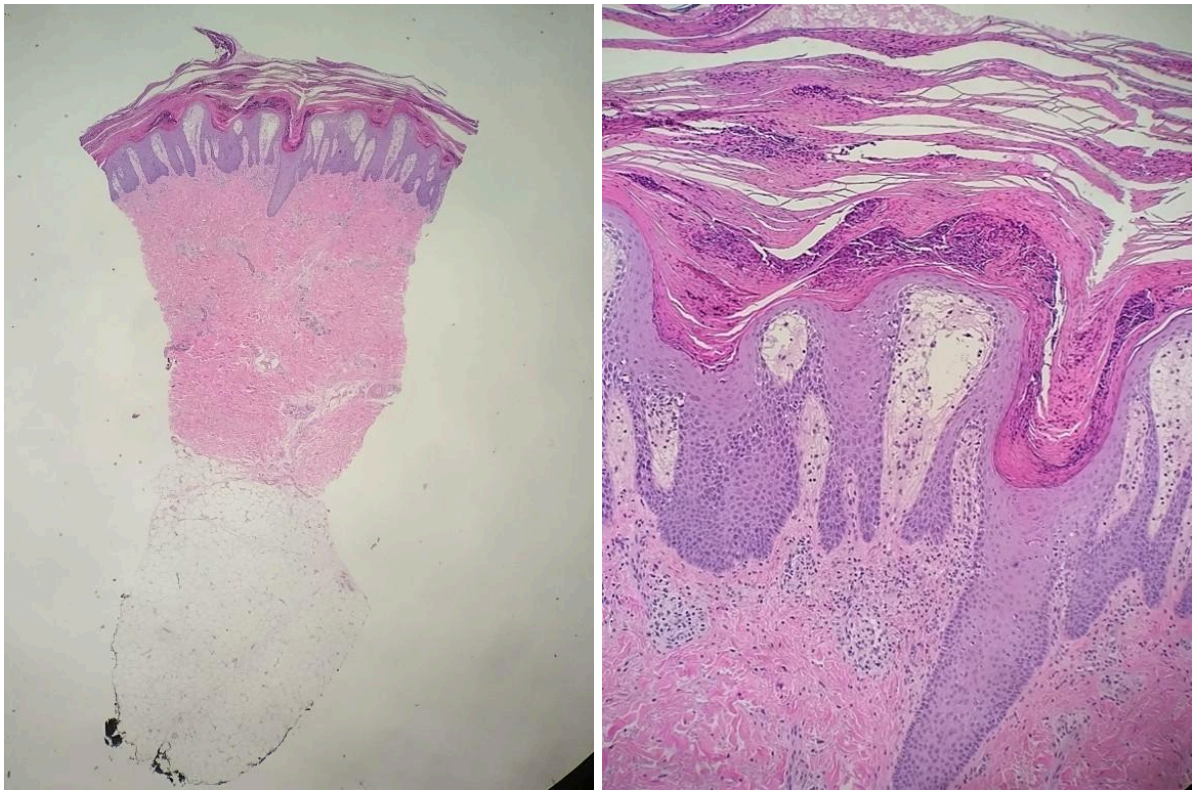






Laboratory Test	Level (Range)
ESR	100 (0-14 mm/hr)
CRP	22.3 (<5.0 mg/L)
ANA	1:2560 (<1:80)
Anti-dsDNA	5 (0-9 IU/mL)
Anti-SM	136.2 (0-24.9 AAU/mL)
Anti-SSA	124.3 (0-24.9 AAU/mL)
Complement C3	54 (87-200 mg/dL)
Complement C4	<8 (19-52 mg/dL)
RF	19 (<15 IU/mL)
CCP	6 (0-19 IU/mL)

Pathology punch biopsy of skin: Marked hyperkeratosis with parakeratosis and intracorneal pockets of neutrophils. Small subcorneal abscesses are seen. The rete ridges and dermal pegs show elongation and clubbing. These findings are definitive for plaque stage psoriasis in the appropriate clinical context.



X-ray of right hand: Erosive arthritis or old avulsion fracture of the base of the proximal phalanx of the little finger laterally.

Outcome

The patient was started on hydroxychloroquine 300 mg daily in conjunction with a topical triamcinolone 0.1% and hydrocortisone 2.5% ointment. Immunosuppressive therapy, including prednisone, was withheld due to the potential risk of worsening skin rash. Future therapeutic strategies include assessing her response to hydroxychloroquine, possible initiation of either azathioprine or belimumab following a thorough evaluation in the outpatient rheumatology clinic.

Discussion

The patient was diagnosed with concurrent plaque psoriasis and SLE. The simultaneous manifestation of both conditions is clinically rare. Therapeutic management of these coexisting autoimmune disorders also presents a significant challenge, as treatment of one condition may inadvertently exacerbate the other. For example, administration of hydroxychloroquine and discontinuation of glucocorticoids during treatment of SLE can potentially induce or worsen psoriasis. Additionally, phototherapy, commonly used to treat psoriasis, may trigger or aggravate SLE. The therapeutic approach should therefore be centered on maintaining a balance in the management of both conditions.

Synovial Osteochondromatosis: A Rare Cause of Joint Pain

Simranjit Kaur MBBS[1], Mercedes Quinones MD[2], David Nashel MD[2]
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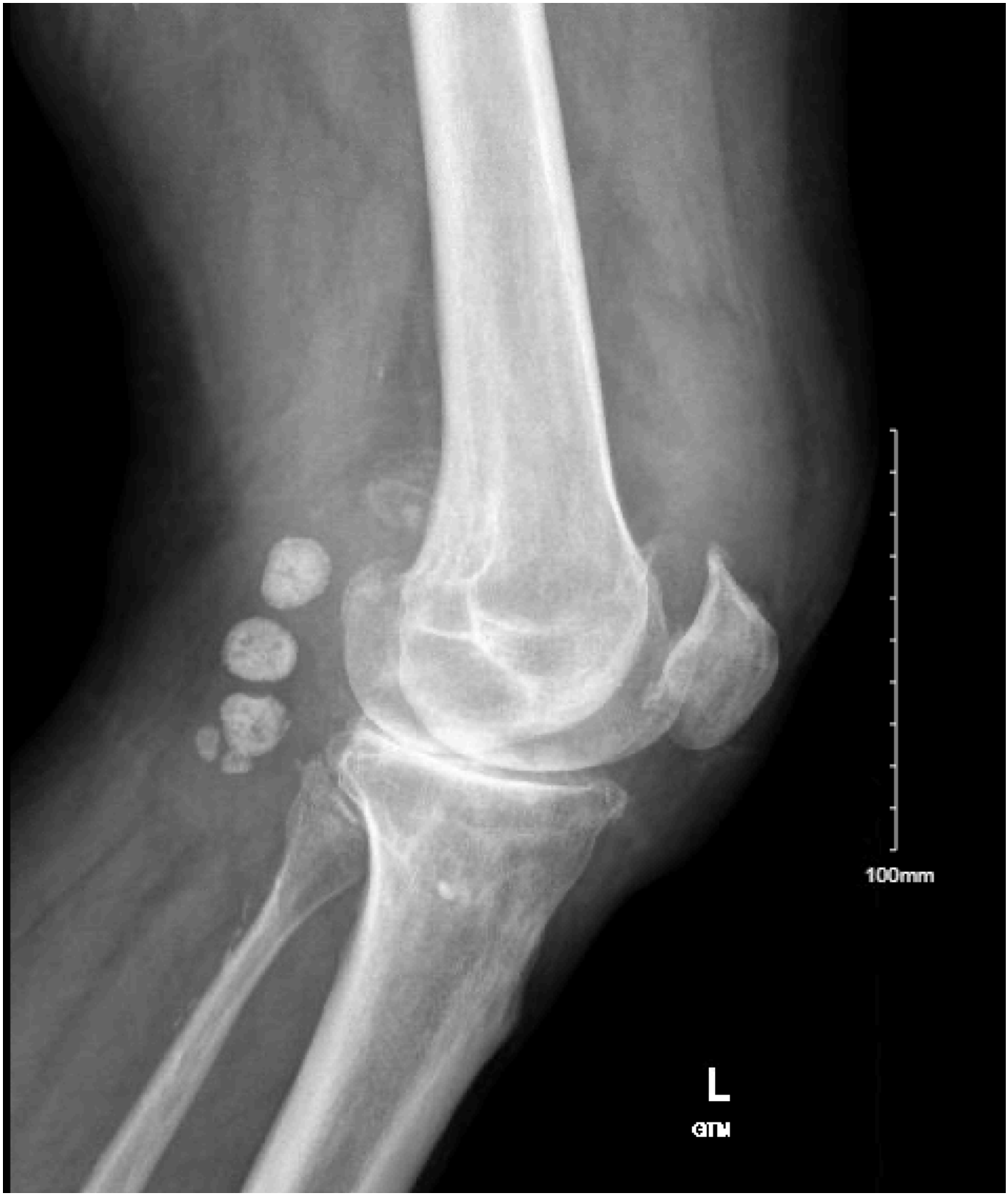
Abstract
Objectives/Methods:

Synovial osteochondromatosis (SO) is a rare, benign joint disorder characterized by the formation of cartilaginous nodules within the synovium, which may ossify and detach into the joint space. It manifests in two forms: primary (idiopathic) and secondary (associated with underlying joint pathology). This retrospective case series describes the clinical, radiographic, and demographic characteristics, comorbidities, management strategies, and outcomes of patients diagnosed with SO at the DC VA Medical Center. Medical records of nine patients diagnosed radiographically with SO were reviewed. Data collected included demographics, clinical presentation, comorbidities, imaging findings, laboratory parameters, and management approaches.
Results:

The cohort included nine patients—seven males and two females—with a mean age of 62.5 years. Eight were African American and one Caucasian. The knee was the most commonly affected joint (n=7), including one bilateral case; two patients had shoulder involvement. All patients presented with joint pain. Additional symptoms included swelling (n=4), decreased range of motion (n=3), joint locking or buckling (n=3), and tenderness (n=3). All patients had coexisting osteoarthritis in the SO-affected joint, indicating secondary SO. Other associated conditions included joint trauma (n=4), chondrocalcinosis (n=3), gout (n=2), rheumatoid arthritis (n=1), hypothyroidism (n=1), and hyperparathyroidism (n=1). Laboratory studies revealed vitamin D deficiency in five of seven tested patients, hyperuricemia in six, and abnormal TSH in three. Management of knee SO was primarily conservative, involving analgesics, physical therapy, and intra-articular injections. One patient underwent knee arthroplasty, and another had arthroscopic meniscectomy. Both patients with shoulder involvement ultimately required shoulder arthroplasty. No cases of malignant transformation were observed.
Conclusion:

All patients in this series had secondary SO, reinforcing its association with degenerative, inflammatory, and metabolic joint diseases—particularly osteoarthritis. The higher mean age at diagnosis may reflect delayed recognition or later disease onset. While knee SO was typically managed conservatively, shoulder involvement led to greater impairment and surgical intervention. The predominance of African American patients likely reflects the VA population rather than an ethnic predisposition. This series contributes to limited literature on SO and underscores the need for larger studies to clarify prognostic factors, guide treatment, and explore potential ethnic or metabolic risk associations.



X-ray of the left knee from one of patients included in the study showing osteochondromatosis secondary to knee osteoarthritis.

A Case of Distinct Finger Demarcation in a Patient with Eczema and Erosive Osteoarthritis: Expanding the Differential Diagnosis of Digital Discoloration

Nikita Shah DO
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Abstract

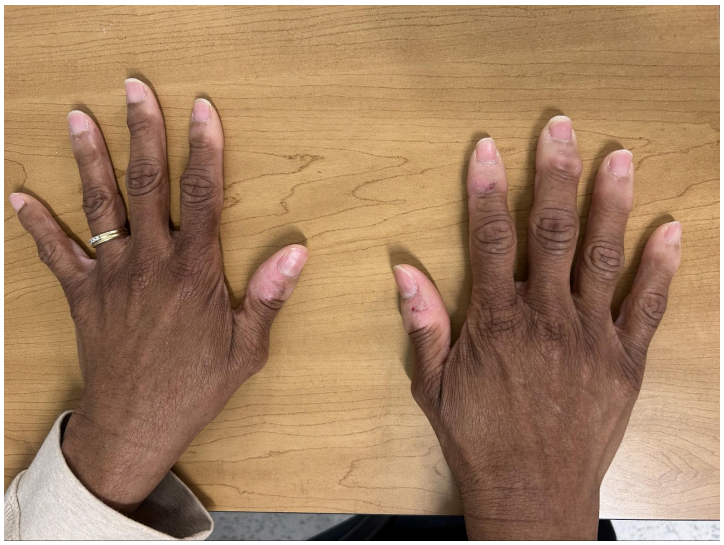
Introduction

Skin changes in older adults with autoimmune and inflammatory conditions can present diagnostic challenges, particularly when involving progressive discoloration with clear demarcation. This phenomenon is commonly seen in eczema, chronic irritant dermatitis, and pigmentary disorders, but may also raise suspicion for autoimmune conditions like systemic sclerosis, Raynaud's phenomenon, or inflammatory arthritis-related skin involvement. In patients with concomitant severe erosive osteoarthritis, distinguishing between mechanical joint disease and inflammatory rheumatologic conditions is crucial.

Case Presentation

A 68-year-old woman presented with progressive discoloration in her fingers and toes over four years, which began while living in Virginia. Her skin developed sharply defined discolored areas that thickened and cracked, leading to bleeding especially during cold weather. She also reported episodic facial and periocular rashes and white skin spots. Her history included adult-onset eczema and severe erosive osteoarthritis but no Raynaud's, lupus, or systemic sclerosis. Her treatment history included ongoing use of Ruxolitinib cream and Dupilumab as biologics while Betamethasone, Tacrolimus, various soaps and cosmetics had been ineffective.

On examination, her skin findings included sharply demarcated discoloration of her fingers (Figure 1) and toes, with associated thickening, fissuring, and bleeding. A facial rash with periorbital involvement and depigmented spots was also. Joint examination revealed severe joint deformities consistent with erosive osteoarthritis, without signs of synovitis or dactylitis. Vascular examination showed no overt Raynaud's phenomenon, though her color changes worsened with cold exposure. Labs revealed a low-positive RNA polymerase III antibody, with ANA, RF, and anti-CCP results pending, as well as no prior documentation of a scleroderma-specific autoantibody panel. Plans were made to repeat autoimmune panels and inflammatory markers for further assessment.



Discussion

The main diagnostic focus remains chronic eczema with pigmentary and vascular changes because of the distinct discoloration boundaries, skin thickening, and resistance to treatment. Her medical history coincides with eczema-related dyspigmentation along with chronic irritation and environmental sensitivity. Since she did not respond to treatment, her eczema appears severe or atypical. Systemic sclerosis should be another diagnostic consideration because of her slight positive RNA polymerase III antibodies. The likelihood of systemic sclerosis as a diagnosis decreases significantly when hallmark features such as sclerodactyly, digital ulcers, Raynaud's phenomenon, nailfold capillary changes, and fibrosis on imaging are absent. Her skin alterations cannot be attributed to EOA because distinct skin demarcation and discoloration are not common presentations of this condition. EOA leads to joint-related erythema and swelling but does not explain her four-year progressive skin discoloration, facial rash, and cold sensitivity. Raynaud's phenomenon emerges as a potential alternative diagnosis since her symptoms intensify during cold, although triphasic color changes or ulceration remain absent.

Conclusion

This case highlights the diagnostic challenge of distinct digital demarcation, particularly in a 68-year-old woman with eczema and erosive OA. While eczema remains the most likely cause, a low-positive RNA polymerase III antibody raises the question of early systemic sclerosis. The absence of Raynaud's, nailfold capillary changes, or systemic sclerosis features suggests that scleroderma is unlikely at this stage. However, given the chronicity and progression of symptoms, ongoing evaluation is warranted.

Purpuric Rash: A Case of IgA Vasculitis with Renal Involvement

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John D. Dingell VA Medical Center, Detroit, MI, USA

Abstract

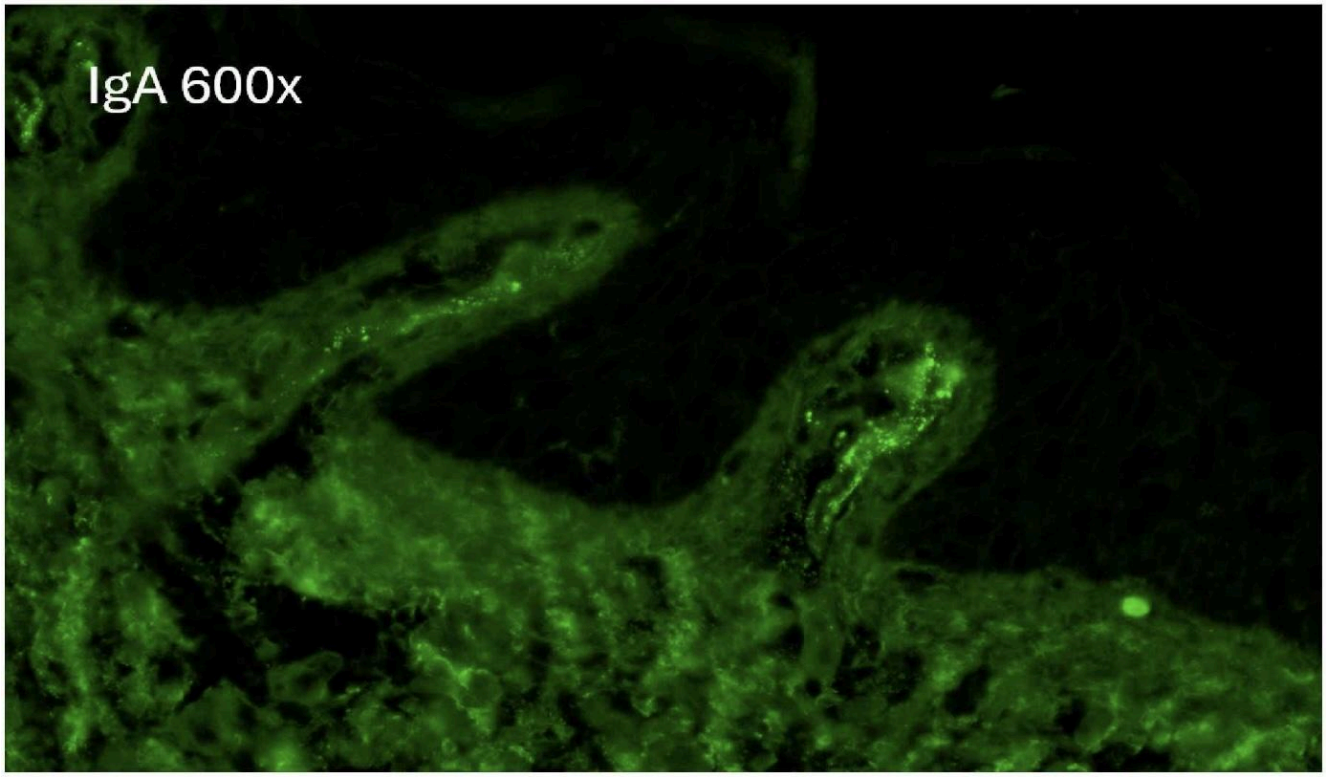
A 76-year-old male with past medical history of hypertension, chronic diastolic heart failure, paroxysmal atrial fibrillation, diabetes mellitus type 2, venous insufficiency, and obesity class III presented with new skin rash.

Patient reported upper respiratory infection (URI)-like symptoms a few weeks ago. Shortly after, he developed a skin rash followed by peripheral edema, fatigue and diarrhea. On examination, there was a palpable purpuric rash involving all extremities and abdomen. Initial labs showed Cr 1.2 mg/dL (at baseline) and urinalysis with microscopic hematuria, but no proteinuria. Additional lab findings included elevated CRP 3.9 mg/dL, elevated ESR 42 mm, positive anti-histone antibody 2.7 units, and mildly positive MPL phospholipid antibody IgM 13 units. Additional serologies including ANA, anti-dsDNA, ANCA, RF, CPK, cryoglobulin, HIV screen and hepatitis panel were negative. He underwent punch biopsy of the skin lesion for suspected vasculitis. He was discharged on prednisone 60 mg daily for 4 days, then taper to 40 mg daily for 4 days and then 20 mg for 12 days.

During follow-up, skin biopsy revealed leukocytoclastic vasculitis with positive findings by direct immunofluorescence supporting IgA vasculitis. He had extensive ulcerating skin rash, and labs showed acute renal injury (Cr 2.1 mg/dL and GFR 32 mL/min) with evidence of hematuria and non-nephrotic range proteinuria. Given concern for IgA vasculitis with renal involvement, he was admitted and given IV Solu-Medrol 1 gm daily for 3 days and underwent kidney biopsy. Kidney biopsy revealed necrotizing glomerulonephritis with early crescent consistent with IgA nephropathy. Kidney function normalized to baseline, and skin involvement was improving. He was discharged on prednisone 60 mg daily with gradual taper and outpatient follow-up.

Outpatient follow-up revealed non-adherence to treatment plan, labs showed worsening renal function (Cr 2.0 mg/dL and eGFR 32 mL/min), elevated inflammatory markers, and urinalysis with trace proteinuria, mild hematuria and hyaline casts. Patient was started on Rituximab infusions while inpatient.

IgA vasculitis is a rare and potentially life-threatening small-vessel vasculitis in adults with increased morbidity associated with renal involvement. Aggressive treatments are warranted to induce remission in patients with a high burden of disease.


IgA 600x

The Great Mimicker

Jenna Bellafiore DO[1], Vaishnavi Gurumurthy MD[2], Eugenio Capitle MD[1]
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Abstract

Granulomatosis with polyangiitis (GPA) has various cutaneous manifestations including petechiae, painful skin lesions, or maculopapular rash. In patients with a prior history of GPA, new cutaneous symptoms will prompt evaluation for a flare. However, it is important to consider that these patients are typically on immunosuppressive medications, such as Rituximab, which predispose them to infection and make traditional laboratory results for infections less reliable.

This is a case of a 63 year old male with history of GPA (diagnosed in 2011 with positive serologies and history of three prior GPA flares) who is maintained on Rituximab, Avacopan and low dose prednisone therapy. Patient presented to the emergency department with a two week history of a raised, pruritic rash starting on the left antecubital fossa spreading to the arms, legs, scalp, neck, back, palms and soles. He also admitted to increasing cough and shortness of breath.

Workup was not suggestive of exacerbation of GPA. Laboratory testing was significant for low IgG and zero CD 19+ B cells. He reported unprotected sex 6 months prior, and rapid plasma reagin (RPR) came back positive at 1:1 though FTA-ABS was negative.

Due to the positive RPR titer, with no known history of syphilis and a consistent clinical presentation, the patient was treated for late latent secondary syphilis with IM Penicillin G (2.4 MU, three doses). His rash improved after the first dose.

Diagnosis of syphilis relies on serologic testing of both non-treponemal (e.g., RPR, VDRL) and treponemal (e.g., FTA-ABS) tests. Both tests, however, rely on antibody formation to produce a positive result.

Immunosuppressive therapies, such as Rituximab (anti-CD 20), reduces the humoral immune response causing delayed seroconversion, low titer or even negative serological tests. In this case, the immunosuppression likely contributed to the low RPR titer and negative FTA-ABS, necessitating clinical suspicion for diagnosis.

Role of Infliximab in IBD-related Orbital Myositis

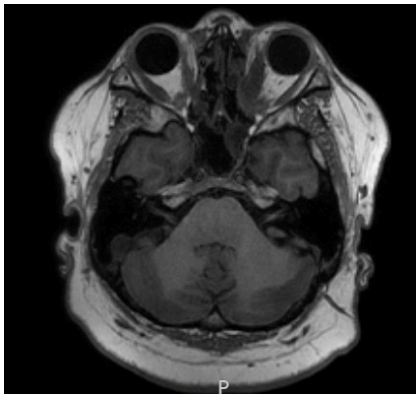
Tarun Selvarajan MD, Lavanya Kannekanti MD, Daniyal Nadeem MD, Kinza Muzaffar MD
LSU Health, Shreveport, LA, USA

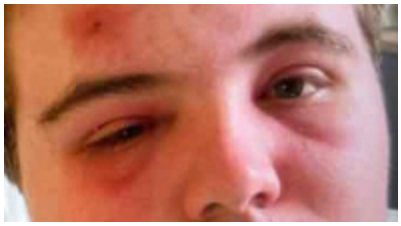
Abstract

Introduction

Orbital myositis is a rare extra-intestinal manifestation of Crohn's disease, typically presents with symptoms such as periorbital pain, proptosis, and diplopia. The case was initially concerning for thyroid eye disease (TED) but evolved into an unusual orbital inflammatory syndrome. This report highlights that TNF-alpha inhibitors works better for orbital inflammation compared to IL12/23 inhibitors.

Case Presentation





- A 26-year-old male with a history of Crohn’s disease with a family history of thyroid cancer, recently transitioned from Infliximab to Ustekinumab, presented with a subacute, progressive ophthalmoplegia and vision loss.
- Examination by an ophthalmologist revealed mild exophthalmos, conjunctival injection, decreased vision in the left eye. A workup for thyroid dysfunction, including TSH, T3/T4, TPO, anti-thyroglobulin, and anti-thyrotropin receptor antibodies, was negative.
- Brain MRI showed severely enlarged extraocular muscles (EOM) bilaterally, with tendon-sparing and left optic nerve stretching. Given concern for TED versus orbital inflammatory syndrome, the patient underwent left orbital decompression. Biopsy showed extensive fibrosis.
- Post-discharge, he experienced recurrent eye inflammation and headaches. A repeat MRI showed significant improvement in EOM enlargement, suggesting orbital pseudotumor over TED.
- Treatment included a 5 day course of IV methylprednisolone with partial improvement, followed by orbital decompression. A slow steroid taper was initiated with 80 mg prednisone daily, though side effects led to adjunctive methotrexate for steroid-sparing benefits.

Discussion

- This case highlights an atypical orbital inflammatory syndrome in a Crohn’s disease patient recently transitioned from Infliximab to Ustekinumab. Biologic therapies, including TNF-alpha and IL-12/23 inhibitors, may trigger ocular inflammation, which

can be an immune-mediated inflammatory response.
- The transition to IL-12/23 inhibition did not control inflammation, leading to relapse and disease progression. Clinicians should consider the potential risks of biologic switching and closely monitor patients with extraintestinal manifestations, particularly those with orbital involvement.

From Translation to Validation: Protocol for Assessing Measurement Equivalence in Cross-Cultural PROMS Development

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[1]International University of Rabat (UIR), Health Sciences Research Center (CReSS), Clinstat Unit, Faculty of Medicine, Rabat, Morocco. [2]Laboratory of biostatistics, Clinical Research and Epidemiology, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco. [3]Department of Rheumatology, El Ayachi Hospital, CHU Rabat-Salé, Rabat, Morocco. [4]UMR 1319 INSPIIRE, Lorraine University, Campus Brabois Santé, Vandœuvre-lès-Nancy, France

Abstract
Background: Patient-reported outcome measures (PROMS) are essential for assessing patients' perspectives on their health conditions and treatments. However, when adopting a PROM instrument for use across different cultural and linguistic contexts, it is necessary to ensure measurement equivalence to preserve the validity and reliability of the instrument in diverse populations. Protocol: The adaptation will follow a structured approach to ensure content validity and cultural relevance for the target population. The first phase will involve cross-cultural adaptation. The original questionnaire will be translated and adjusted to reflect the linguistic and cultural nuances of the intended audience. Cognitive Debriefing will be conducted with a sample from the target group to assess each item's clarity and cultural appropriateness. A Structural Analysis will then be performed using factor analysis to examine the underlying structure of the adapted instrument. Reliability will be assessed by measuring internal consistency among the items within the scale, while test-retest will ensure stability over time. Convergent and Divergent Validity will also be evaluated to ensure the instrument's accuracy. Finally, Scale Validity will be examined using Item Response Theory and Measurement Invariance, ensuring the instrument remains valid and reliable across diverse populations. Discussion: This rigorous methodological approach will ensure the adapted PROM retains its original psychometric properties and cultural appropriateness. Failure to adhere to COSMIN guidelines in Arabic PROM adaptations can compromise conceptual, semantic, and quantitative equivalence. The objective of this protocol will be to establish a methodologically sound framework for the cross-cultural adaptation and validation of PROMs, following COSMIN recommendations and contributing to developing reliable health outcome measures. From translation to psychometric validation, achieving measurement equivalence in cross-cultural PROM development will be a complex yet essential process. This methodological framework will guide researchers and clinicians to ensure the robustness and applicability of PROMs in multinational studies and clinical practice.
References:
1-Mokkink LB, Prinsen CA, Patrick DL, Alonso J, Bouter LM, de Vet HC, et al. COSMIN Study Design checklist for Patient-reported outcome measurement instruments.
2-García de Yébenes Prous MAJ, Rodríguez Salvanés F, Carmona Ortells L. [Validation of questionnaires]. Reumatol Clin. 2009;5(4):171–7.

Differential Effects of GLP-1 Receptor Agonists versus SGLT2 Inhibitors on Hypoglycemia and Infection Risk in Diabetic Patients with Inflammatory Arthritis

Sila L Mateo Faxas M.D.[1], Godbless Ajenaghughrure M.D.[1], Kim Nguyen M.D.[1], Gurjot Singh M.D.[1], Nirys Mateo Faxas M.D.[2]
[1]Trihealth Good Samaritan Hospital, Cincinnati, Ohio, USA. [2]Independent Author, Santo Domingo, Dominican Republic, Dominican Republic

Abstract

Background

While both SGLT2 inhibitors and GLP-1 receptor agonists improve glycemic control in type 2 diabetes (T2D), their safety profiles, particularly regarding hypoglycemia and infection risk, may differ in patients with inflammatory conditions. This study compared these outcomes in patients with T2D and inflammatory polyarthropathy treated with either drug class.

Methods

This retrospective cohort study analyzed data from the TriNetX global health research network, identifying adult patients with T2D and inflammatory polyarthropathy receiving either SGLT2 inhibitors (n=3,377) or GLP-1 receptor agonists (n=4,733). After propensity score matching (n=2,838 per group), we evaluated the incidence of hypoglycemia, respiratory infections, urinary tract infections, sepsis, and other infection-related outcomes over a 5-year follow-up period.

Results

Patients treated with SGLT2 inhibitors experienced higher rates of sepsis (8.1% vs 5.9%, HR 1.609, 95% CI 1.318-1.965, $p<0.001$) and acute kidney injury (16.9% vs 13.6%, HR 1.371, 95% CI 1.216-1.547, $p<0.001$) compared to those on GLP-1 receptor agonists. Hypoglycemic events were more common in the SGLT2 inhibitor group (1.8% vs 1.1%, HR 1.594, 95% CI 1.028-2.472, $p=0.036$). The SGLT2 inhibitor cohort also showed higher rates of endocarditis/valve disorders (3.8% vs 2.9%, HR 1.478, 95% CI 1.098-1.990, $p=0.043$) and bacteremia (3.4% vs 2.4%, HR 1.627, 95% CI 1.205-2.196, $p=0.001$). Pneumonia incidence was similar between groups. Acute respiratory distress was more frequent with SGLT2 inhibitors (11.0% vs 6.7%, HR 1.699, 95% CI 1.480-1.949, $p<0.001$).

Conclusion

In patients with T2D and inflammatory polyarthropathy, treatment with GLP-1 receptor agonists was associated with lower risks of serious infections, acute kidney injury, and hypoglycemia compared to SGLT2 inhibitors. These findings suggest that GLP-1 receptor agonists may offer a more favorable safety profile in this specific patient population. Clinicians should consider these differences when selecting glucose-lowering agents for patients with T2D and inflammatory arthritis.

Cardiovascular Event Risk in Psoriatic Arthritis Patients with Diabetes: Impact of BMI Status

Sila L Mateo Faxas M.D.[1], Godbless Ajenaghughrure M.D.[1], Kim Nguyen M.D.[1], Gurjot Singh M.D.[1], Nirys Mateo Faxas M.D.[2]
[1]Trihealth Good Samaritan Hospital, Cincinnati, Ohio, USA. [2]Independent Author, Santo Domingo, Dominican Republic, Dominican Republic

Abstract

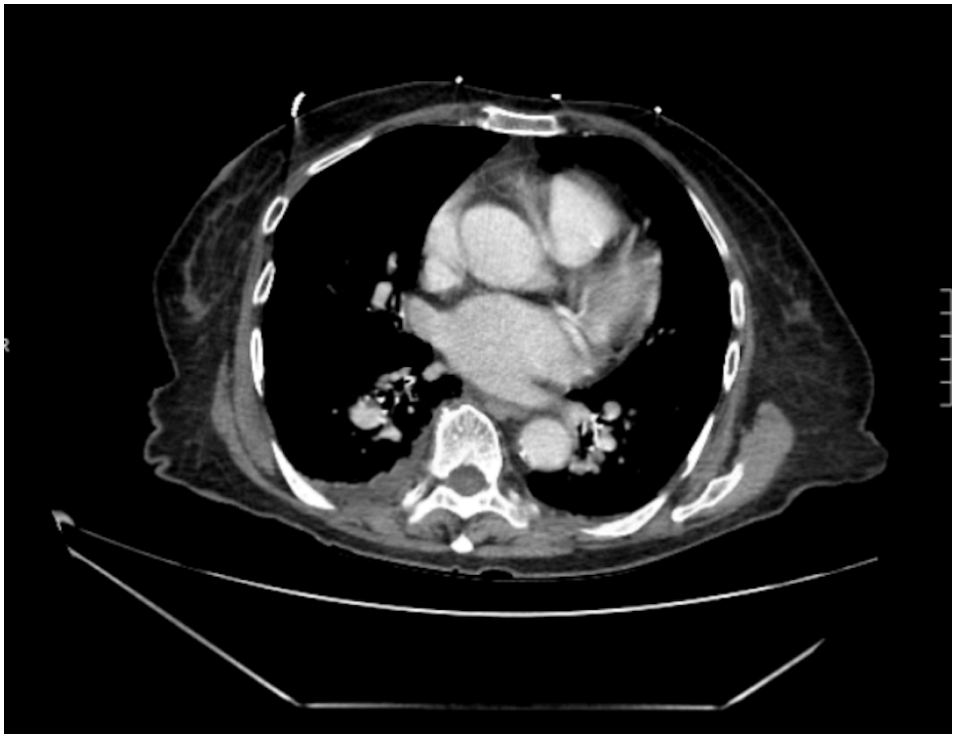
Background:
Psoriatic arthritis (PsA) and type 2 diabetes mellitus (T2DM) are independently associated with increased cardiovascular risk. However, the impact of obesity on cardiovascular outcomes in patients with both conditions remains poorly understood.
Methods:
Using the TriNetX global federated health research network, we conducted a retrospective cohort study comparing cardiovascular outcomes between patients with PsA and T2DM stratified by BMI. Cohort 1 included non-obese patients (BMI <30 kg/m^2; n=9,762) and Cohort 2 included obese patients (BMI 30-60 kg/m^2; n=9,762) matched by propensity score across demographic and clinical characteristics. Outcomes were assessed over a 5-year follow-up period.
Results:
After propensity score matching, the risk of all-cause mortality was significantly higher in non-obese patients compared to obese patients (9.1% vs 5.9%, risk difference 3.2%, 95% CI 2.5-4.0%; p<0.001). Non-obese patients demonstrated lower survival probability at the end of the 5-year follow-up (84.7% vs 90.2%, p<0.001) with a hazard ratio of 1.65 (95% CI 1.49-1.83). The non-obese cohort also exhibited higher incidence of cardiac arrest (2.5% vs 1.1%), cerebral infarction (6.6% vs 4.3%), heart failure (17.1% vs 9.6%), and pulmonary embolism (3.3% vs 2.0%) compared to the obese cohort (all p<0.001).
Conclusions:
In patients with concurrent PsA and T2DM, those with BMI <30 kg/m^2 demonstrated significantly higher cardiovascular risk compared to matched obese patients. This apparent "obesity paradox" warrants further investigation into underlying mechanisms, including potential disease severity, inflammatory burden, and metabolic differences between these populations.

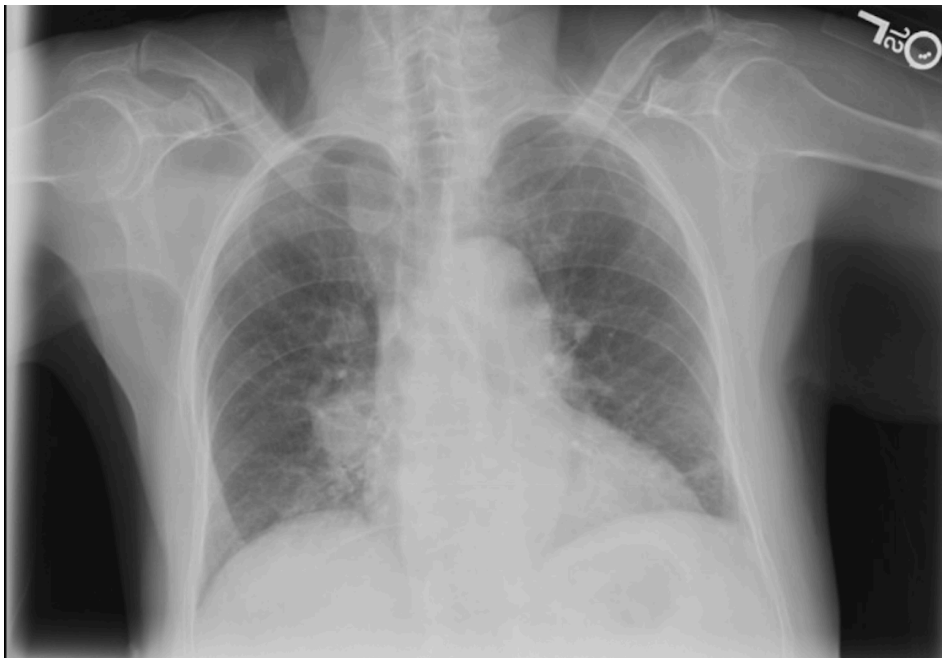
A Diagnostic Curveball: Granulomatous Disease Mimicking Metastatic Cancer

Greeshma Erasani MD[1], Punith Chirumamilla MD[2], Ashu Cingareddy Bachelor of Science[3], Lavanya Kannekanti MD[4], Julian Magadan MD[5,6]
[1]University of Missouri-Kansas City school of medicine, Kansas city, Missouri, USA. [2]Baptist Memorial Hospital- North Mississippi, Oxford, Mississippi, USA. [3]University of Texas-Dallas, Dallas, Texas, USA. [4]LSU Health Shreveport School of medicine, shreveport, Lousiana, USA. [5]Kansas City Physician Partners, Kansas city, Missouri, USA. [6]Saint Luke's Hospital of Kansas City, kansas city, Missouri, USA

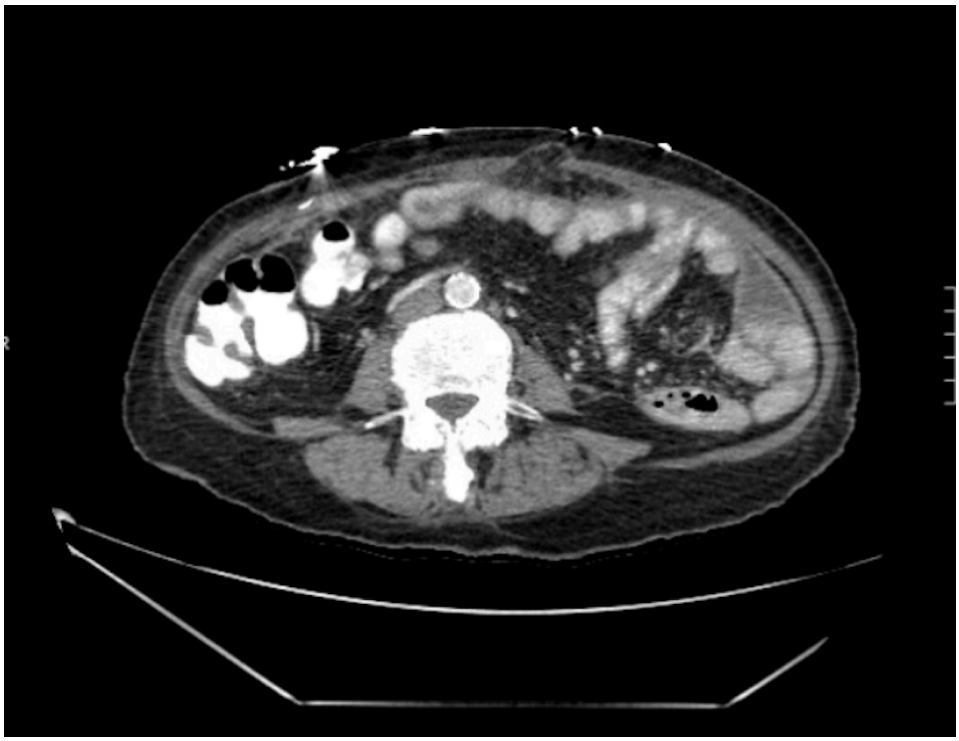
Abstract

CT chest and Chest X-Ray showing multiple pulmonary nodules





CT abdomen and pelvis showing multiple peritoneal lesions



Introduction:

Sarcoidosis is a multi-system granulomatous disorder most commonly affecting the lungs and lymph nodes. However, it can present with a wide range of atypical manifestations that mimic malignancy, infection, or other inflammatory conditions. We hereby present a case of atypical manifestation of Sarcoidosis in an elderly patient.

Case Presentation:

A 79-year-old female with a history of seropositive non-erosive rheumatoid arthritis (RA), MGUS, chronic kidney disease stage 3, gout, and Alzheimer's dementia presented for follow-up of inflammatory arthritis and recurrent hypercalcemia requiring multiple hospitalizations.

A CT chest was performed due to concerns for malignancy versus sarcoidosis, which demonstrated mediastinal lymphadenopathy with multiple new punctate pulmonary nodules and also raised concern for mesenteric carcinomatosis. Subsequent CT abdomen/pelvis revealed a mixed cystic-solid right lower quadrant mass with findings consistent with peritoneal carcinomatosis. PET imaging showed metabolically active peritoneal and mediastinal disease, raising strong suspicion for metastatic ovarian cancer.

A bone marrow biopsy was performed to evaluate for possible multiple myeloma in the context of hypercalcemia, abnormal Serum protein electrophoresis and associated anemia, but the results were negative. Tertiary hyperparathyroidism due to chronic kidney disease was also considered; however, PTH levels were found to be low. A biopsy of her peritoneal mass showed benign fibro-adipose tissue with non-caseating granulomatous inflammation consistent with sarcoidosis.

The patient was started on azathioprine 50 mg daily and a prednisone taper, along with infliximab 100 mg IV on week 0,2,6 followed by every 8 weeks for systemic sarcoidosis and RA. On follow-up, she remained clinically stable without synovitis, abdominal pain, or respiratory complaints. RA therapy included hydroxychloroquine, with plans for discontinuation if biologic therapy is well-tolerated. Hypercalcemia is being monitored, and follow-up imaging is planned in 6 months.

Conclusion:

This case illustrates the deceptive nature of systemic sarcoidosis, presenting with imaging findings indistinguishable from metastatic malignancy. Definitive diagnosis through biopsy was pivotal in guiding appropriate, non-oncologic management. Tailored immunosuppressive therapy led to clinical improvement. It reinforces the value of tissue confirmation in complex diagnostic scenarios.

CNS Lupus Vasculitis Complicated by Intracerebral Hemorrhage (ICH): A Case Report and Literature Review on Patterns of ICH in Systemic Lupus Erythematosus

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Geisinger Medical Center, Danville, PA, USA

Abstract

Introduction: Intra-cerebral hemorrhage (ICH) due to vasculitis is a rare manifestation of neuropsychiatric SLE (NPSLE). Vasculitis-related infarcts and sub-arachnoid hemorrhages (SAH) are more commonly seen. Thrombosis-related complications are often seen with co-existing antiphospholipid syndrome (APS). We report a case of ICH occurring in a patient with stable SLE in the absence of APS.

Case: A 19-year-old African American female with SLE complicated by class IV lupus nephritis, renal thrombotic microangiopathy, and HTN presented with acute onset of vomiting and seizures. She required intubation and nicardipine infusion for hypertensive emergency. At the time of presentation, she was on prednisone, Mycophenolate mofetil, Belimumab, and Plaquenil, and her SLE monitoring labs were within normal parameters two weeks before admission. Anti-phospholipid antibodies were negative. Imaging demonstrated an acute large intraparenchymal hemorrhage in the right parieto-occipital and left occipital lobes with surrounding edema and midline shift. CTA showed abnormal appearing bilateral MCAs and PCAs. MRI brain demonstrated several areas of T2 FLAIR hyperintensity and enhancement in both cerebral hemispheres, notably in right frontal corona radiata and right cingulate gyrus, most suggestive of an inflammatory process. In this patient with known SLE, the most likely explanation for these findings was CNS vasculitis complicated by hemorrhage, which is quite rare. We initiated pulse-dose glucocorticoids and escalated her regimen to IV cyclophosphamide. She gradually made clinical recovery and was discharged with little to no residual neurological deficits.

Discussion: Table 1 reviews case reports of patients with SLE presenting with ICH as a complication of SLE vasculitis, APS, or both. In contrast to hypertensive brain bleeds (basal ganglia most common location), we can appreciate how SLE-related cerebral bleeds are mainly in the lobar regions. This radiological distinction is important to identify from other etiologies of ICH, since vasculitis manifesting as hemorrhage requires escalation of immunosuppressive regimen and an aggressive approach to treatment. In some cases, this may even be the initial presentation of SLE, hence, familiarity with imaging findings can aid in early recognition of a life-threatening presentation.

	Age/Sex	Race/Country	Location of ICH	Co-existing APS/aPL antibodies	Treatment escalation after bleed
Our case	19 F	African American/USA	Right parieto-occipital and left occipital	No	Pulse-dose steroids, IV CYC, Belimumab
1.	33 F	Filipino/ Philippines	Right occipital and parietal lobes	No	Prednisone, MMF, anti-HTN, anti-seizure regimen
2.	52 F	Japanese/ Japan	Right frontal and left temporo-occipital area	No	Emergent surgical evacuation of hematomas

3.	18 F	Nepalese/Nepal	Left occipital lobe and hemorrhagic foci in right frontoparietal lobe	Yes	Emergent evacuation of hematoma, prednisolone, MMF, Plaquenil, warfarin
4.	31 F	Unknown/Greece	Not specified	Yes	Pulse dose steroids, IV CYC, IVIG
5.	28 F	Hispanic/USA	Right temporal and left occipital lobe	No	IV glucocorticoids, Intra-arterial injection of verapamil in cerebral vessels
6.	32 F	Taiwanese/Taiwan	Right frontal parasagittal region and SAH	No	Pulse-dose steroids
7.	35 F	South Asian/ India	Multiple small intraparenchymal bleeds	No	Mannitol, anticonvulsants, steroids
8.	56 F	Unknown/Pakistan	Right frontoparietal lobe	No	Pulse dose steroids, craniotomy for hematoma evacuation

Timely Escalation of Care in New-Onset SLE-Associated Hemophagocytic Lymphohistiocytosis: A Systematic Review of 24 Cases

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Abstract

Objectives: To describe clinical and laboratory features of hemophagocytic lymphohistiocytosis (HLH) as the initial presentation of systemic lupus erythematosus (SLE), assess the impact of timely inter-hospital transfers on outcomes, and recommend strategies for early recognition in emergency or community settings.

Methods: We systematically reviewed cases of new-onset SLE-associated HLH and searched the University of Maryland Health Sciences Human Services Library databases using the keywords "SLE," "HLH," and "Macrophage Activation Syndrome (MAS)." We included peer-reviewed, English-written abstracts, case reports, and case series from 2001 to 2025 that met the HLH diagnostic or H-Score criteria. 23 articles were included from 186 initially screened (Figure 1). Data on demographics, symptoms, labs, treatment, and outcomes were extracted and compared to our case.

Results: Among 24 SLE-associated HLH cases, 70.8% were female, ranging from 4 to 73 years (mean age of 27 and median age of 25). Common features include fever, cytopenias, hyperferritemia, hypertriglyceridemia, and a positive ANA, along with other SLE-related manifestations such as rash, arthralgia, or renal involvement. While all patients received corticosteroids, additional immunosuppressive therapies varied, including IVIG (16.6%), rituximab (12.5%), etoposide (20.8%), cyclophosphamide (29.2%), cyclosporine (24%), mycophenolate (20.8%), and plasmapheresis (12.5%). The crude mortality rate was 8.3%. Our patient experienced rapid clinical deterioration, marked by severe hyperferritinemia (17,280 ng/mL), transaminitis (AST 5,059 U/L; ALT 1,557 U/L), hypertriglyceridemia (1,399 mg/dL), hemolytic anemia (LDH 9,418 U/L), renal failure (creatinine 5.69 mg/dL; GFR 10 mL/min/1.73m^2), and encephalopathy. Transfer to an affiliated tertiary care center was initiated within 24 hours of admission. Among the other cases, there was another patient who also required transfer for specialized treatment.[1]

Conclusion: Our case highlighted the importance of early recognition of rapid deterioration and timely escalation of care, which was facilitated through a streamlined transfer process. Early recognition in emergency and community settings can be facilitated by maintaining a high index of suspicion for HLH and considering early ferritin testing. Integrated healthcare networks are necessary to connect community-based care and high-level tertiary services, thus enabling access to multidisciplinary care and interventions to manage life-threatening complications of HLH.

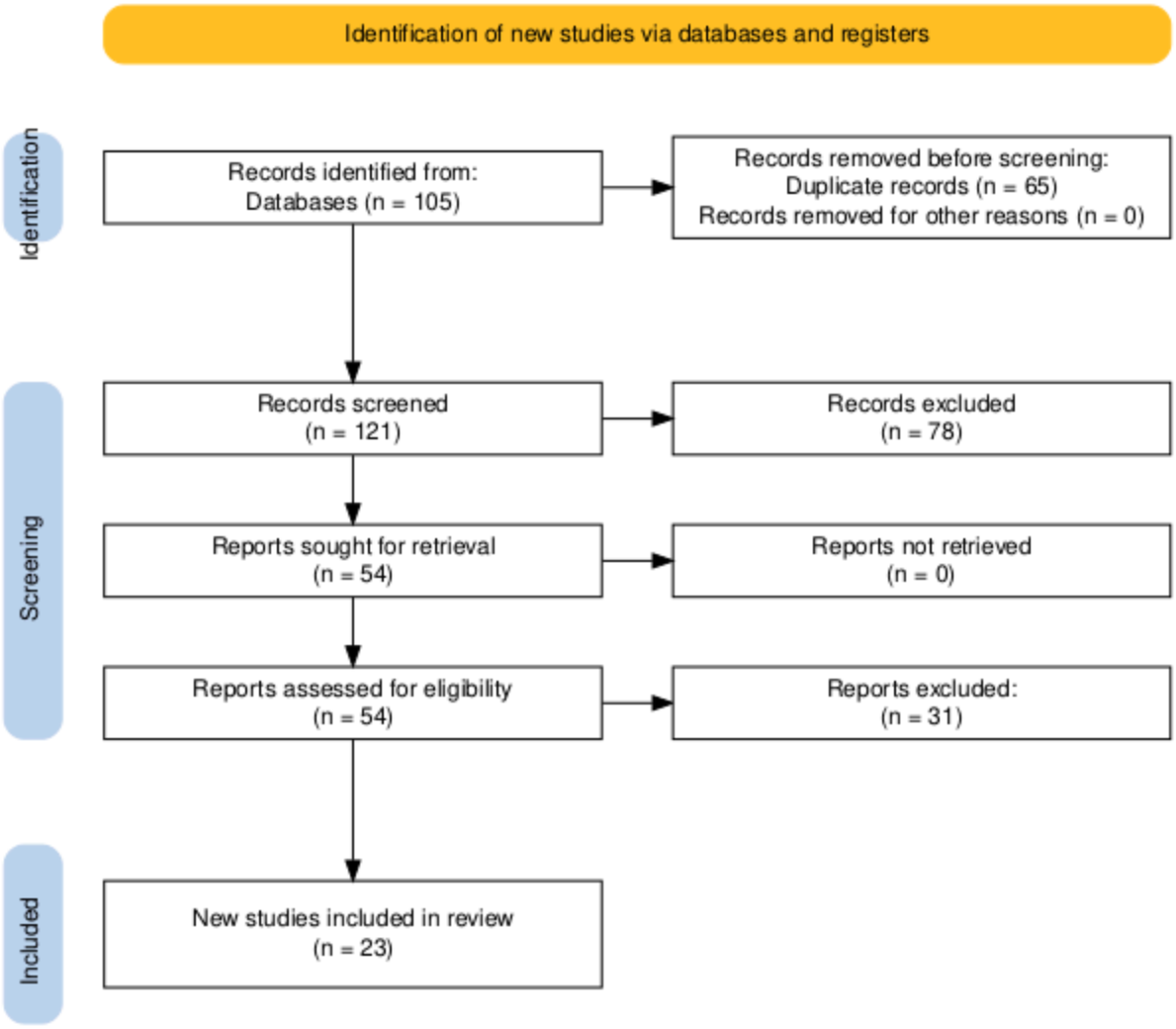

Figure 1: PRISMA Flow diagram of Search Strategy

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Fellow-Initiated Rheumatology EDUcation Project (FIRED UP)

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Abstract

Objective

Rheumatology is an important field with topics encountered commonly on board exams and clinically, yet it remains an area of low confidence for trainees. This innovative project seeks to increase resident competency in rheumatology topics, improve performance on rheumatology questions on standardized exams, and increase interest in rheumatology. As a part of this unique project, fellows also expand and gain confidence in their teaching skills.

Methods

Internal medicine residents rotating on the rheumatology consult service are asked to answer a pre-rotation quiz and survey to gauge initial rheumatology knowledge and interest. Rheumatology fellows develop and deliver weekly didactics to the residents based on high-yield topics.

The primary outcome is the residents' gain in rheumatology knowledge measured by self-assessment and pre-and post-quiz scores. The pre- and post-tests contain similar, but not identical, questions randomized from a set in order to best gauge learners' increase in knowledge rather than recalling the correct answers. We also evaluate changes in learner confidence in diagnosing common rheumatic diseases (gout, rheumatoid arthritis, lupus, scleroderma), and confidence in treating common or important diseases encountered in rheumatology (gout, osteoarthritis, scleroderma renal crisis).

Results

In the initial pilot phase of the study, pre-rotation quiz scores were 62.5% correct compared with 83.8% correct post-rotation. The lowest area of confidence for learners was perception of the ability to treat scleroderma renal crisis (3.31 out of 10 confidence; n=13). The area of highest confidence for learners was perception of the ability to treat osteoarthritis (7.62 out of 10 confidence; n=13).

Preliminary results show an increase in confidence in all assessed areas, with the largest increase in treatment of scleroderma renal crisis (from 3.31 out of 10 confidence to 6.29 out of 10 confidence). Changes in fellows' confidence in teaching will be measured at the end of the academic year.

Conclusions

This project highlights the need for increased rheumatology education and provides a unique way to directly engage rheumatology fellows in the education of medicine residents.

WHERE YOU LIVE MATTERS: EXPLORING REGIONAL DIFFERENCES IN SLE AND IHD MORTALITY:

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Abstract

Background:
Over recent decades, regional disparities in mortality rates for systemic lupus erythematosus (SLE) and ischemic heart disease (IHD) have become apparent, driven by a variety of health determinants. This study analyzes national data to reveal mortality trends across U.S. regions, highlighting areas where the greatest health burdens persist and how they have changed over time.

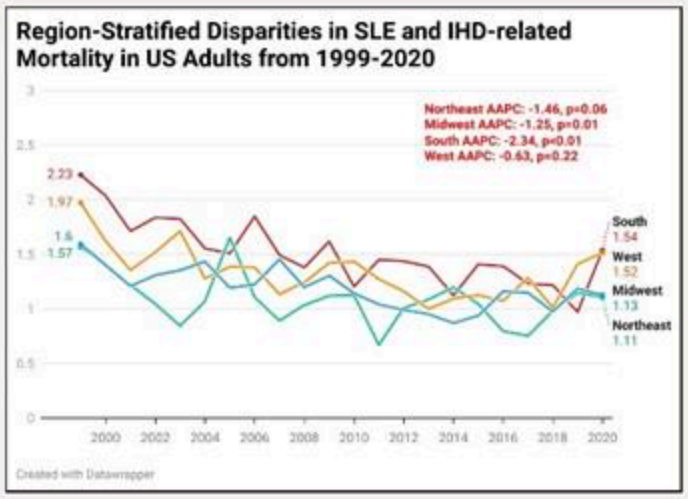
Methods:
We utilized the CDC WONDER database to analyze mortality data for U.S. adults aged ≥ 25 from 1999 to 2020. Age-adjusted mortality rates (AAMRs) per 1,000,000 were calculated, stratified by region, and analyzed using Annual Percent Change (APC) and Average Annual Percent Change (AAPC). Statistical significance was determined using p-value <0.05.

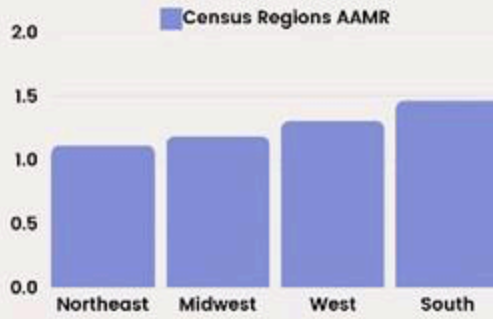
Results:
From 1999 to 2020, the South had the highest AAMR (1.48, 95% CI: 2.23-1.54), followed by the West, Midwest, and Northeast. Mortality rates decreased in all regions, with the South showing the smallest decline (AAPC: -2.34, $p<0.01$). Oklahoma had the highest state-level AAMR (2.21), while Massachusetts had the lowest (0.57).

Conclusion:
This study reveals stark regional differences in mortality rates for SLE and IHD. By identifying areas with the highest mortality burdens, targeted health strategies can be developed to reduce these disparities and improve outcomes nationwide.

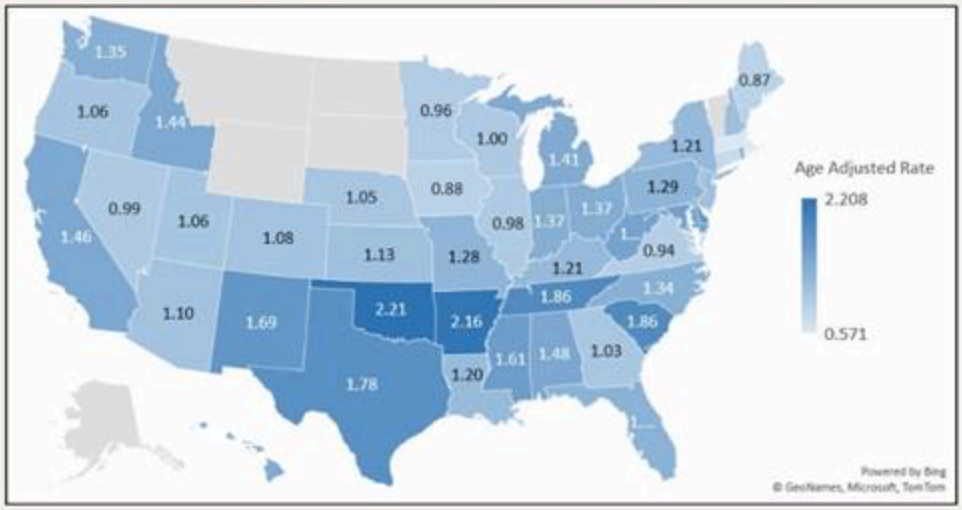
CENTRAL ILLUSTRATION: Regional stratification of Systemic Lupus Erythematosus and Ischemic Heart Disease-related Mortality among US adults from 1999-2020.

Locations (Age-Adjusted Mortality Rate per 1,000,000 persons among Adults)





State map representation (Age-Adjusted Mortality Rate per 1,000,000 persons among Adults)



States above 90th Percentile according to AAMRs:
Oklahoma
Arkansas
South Carolina
Tennessee

States below 10th Percentile according to AAMRs:
Massachusetts
Connecticut
Maine
Iowa

Sarcoid, Infection, Sarcoma?

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Abstract

We present a case of a 32 year old non-smoking male who presented initially with a dry cough. He was then found to have multiple lung nodules on CXR. Patient had followed up with a pulmonologist who ordered a CT chest – this confirmed numerous nodules. A nodule was biopsied with non-specific results. With concern for a metastatic process, he had gotten a full body CT scan as well as a brain MRI. Brain MRI was concerning for metastatic disease. The patient had admitted to having some headaches, but otherwise no neurologic symptoms. The patient was admitted for brain biopsy. Rheumatology was involved to help rule out an autoimmune etiology. The rheumatologic differential included: vasculitis (ANCA labs negative), IgG4 (IgG4 level mildly elevated at 104, brain biopsy with negative staining), and sarcoidosis (IL-2 elevated at 1064, lysozyme normal, ACE was collected but did not result, no hilar lymphadenopathy). He also had a positive ANA of 1:80 with a negative profile.

The patient had two brain biopsies – the first with some features consistent with the rare Erdheim Chester Disease (foamy histiocytes) that were not confirmed on repeat biopsy. The biopsy was also stained for IgG4 and returned negative. The patient had also had a thorough infectious disease workup which returned negative. The leading differential was malignancy. The patient chose to continue his care at MD Anderson. There he had a wedge resection of his lung that was consistent with Alveolar Soft Part Sarcoma.

Alveolar Soft part Sarcoma is a rare cancer that usually has primary lesions in the long bones of the legs. It is one of the rarest sarcomas (0.2%-1% of all soft tissue sarcomas) and usually affects younger patients. Our patient was found to have some 4mm sclerotic foci on both of his femurs on MRI. The therapy is usually resection and radiation. Our patient underwent gamma knife radiation for his brain lesions. Chemotherapy is not thought to be effective for this condition. This was a good learning case as it had a broad differential and a definitive ending diagnosis of a rare disease.

Atypical case of adult-onset onset stills disease in a patient with alpha-gal syndrome

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Abstract

Adult-onset Still's disease is a rare systemic inflammatory disorder of unknown etiology presenting with inflammatory polyarthritis, daily fever, and transient salmon pink maculopapular rash.

-Alpha-gal syndrome is a condition in which people are allergic to alpha-gal, usually found in mammals other than primates. It presents with pain in the abdomen, breathing difficulty, and diarrhea.

Presentation

-A 44-year-old Caucasian female presented with recurrent fever, swelling in the hands and feet, salmon pink maculopapular rash, migrating arthralgias, dysphagia, sore throat, cold hands, and feet for a few months before she presented.

-Her alpha-gal syndrome usually presents with fevers, myalgias, diarrhea, and stomach pain triggered by eating mammalian meat. However, she reported that she has not eaten any recently.

-Initial labs showed leucocytosis of 22.44 $*10^3$, with mild transaminitis. ESR - 61, CRP -338.9, RF 14, Ferritin 23431. Her IgG was positive for EBV, CMV, while viral load remained negative. Workup for hepatitis and other infectious panel, including blood cultures and urine cultures, was negative. ANA, anti-phospholipase antibodies, RF, anti-CCP, and other autoimmune workup were negative.

- CT abdomen: showed moderate pericardial effusion, mild left-sided pleural effusion, and hepatosplenomegaly.
- Biopsy of the skin showed perivascular inflammation with dermal edema
- Rheumatology recommended anakinra, after which she showed significant improvement in joint pains, swelling. However, her fever spikes were not controlled, so prednisone was added later on.

Criteria: As discussed in Figure 1, other inflammatory disorders, such as Rheumatoid arthritis and HLH, must be ruled out. MAS is a subtype of HLH that presents with high ferritin, cytopenias, transaminitis, and elevated ESR and CRP. She did not get a bone marrow biopsy. Regardless, this case has met 4 major and 4 minor criteria.

Management: For mild disease, NSAIDS remain the mainstay of treatment. While for moderate to severe disease, or disease extending for more than 2 weeks, anakinra and prednisone remain the first-line treatment.

Conclusions

For cases with such diffuse non-specific symptoms, differentials should be broadened to include stills disease after ruling out infectious and other rheumatological etiologies. Once diagnosed multidisciplinary approach should be considered and treated with steroids and immunomodulators

Successful Use of Pegloticase with Concurrent Immunosuppressive Therapy

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Abstract

Introduction:

Pegloticase has been demonstrated to be effective in the treatment of refractory gout. It has successfully controlled uric acid levels and resolved at least one tophi in 40% of patients.[1] Co-treatment with disease modifying anti-rheumatic drug (DMARD) therapy has reduced reactions and increased response rates.[1,2] This case of tophaceous gout was successfully treated using Azathioprine with Pegloticase.

Case Presentation:

This 63 year old Caucasian male with a PMH of CAD, CHF and hypertension was diagnosed with gout over 30 years ago. On presentation in August 2023 he complained of constant joint pain in his hands, elbows, knees and feet. His previous treatment history included allopurinol, colchicine, indomethacin and prednisone, without improvement. He reported frequent gout attacks, used alcohol weekly.

His BMI was 36. Both hands were disfigured with multiple tophi on all fingers and knuckles and a left index finger amputation. His elbows and feet had similar findings.

Being ineligible for febuxostat, he was started on prednisone 10mg daily, azathioprine 50mg daily and Pegloticase 8mg infusions every 2 weeks. His uric acid decreased from 10.6mg/dL to less than 1.0mg/dL after five infusions. No reactions reported. On annual follow up, pain and tophi had significantly reduced in all sites.

Summary:

Pegloticase is a recombinant polyethylene glycol form of uricase used in treatment refractory disease. The overall response rate in clinical trials is 42% with monotherapy, of which 89% of patients developed antibodies.[3]

Most common side effects were gout flares and infusion reactions (IR) out of which 5.3% were anaphylactic; mostly in those that developed anti-drug antibodies.[4] Achievement of higher response rates and reduction of adverse effects is beneficial and easily attained through use of immune modulating co-therapy such as DMARDs. Methotrexate, azathioprine and mycophenolate mofetil have all been used to increase response rates to 63.6% to 87.5%.[2,5]

We have successfully treated this patient using azathioprine to prevent treatment failure, therefore improving tolerability, reducing reactions and increasing disease control. [2] Maximizing these effects is essential.

Figure 1: Right hand before treatment

Figure 2: Right hand after treatment

Figure 3: Left hand before treatment

Figure 4: Left hand after treatment

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From Autoimmunity to Remission: Unlocking the Potential of CAR-T Therapy in Psoriatic Disease

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Abstract

Psoriatic arthritis (PsA) is a chronic inflammatory disease characterized by a multifaceted relationship between immunological, genetic, and environmental factors. The pathogenesis involves both the innate and adaptive immune systems, with significant involvement of autoreactive T-cells (1). Chimeric antigen receptor (CAR) T-cell therapy is emerging as a treatment for connective tissue diseases, especially those with substantial autoimmune elements (2). The therapy primarily targets autoreactive B-cells, which play an important role in the pathogenesis of these disorders. CAR T-cell therapy, particularly targeting CD19, leads to a profound and sustained depletion of B-cells in autoimmune diseases. This results in a reduction of autoantibody production and an “immune reset”, in which the immune system’s unregulated mechanisms are recalibrated (3). The potential application of CAR T-cell therapy in the treatment of PsA is an area of budding interest, although specific clinical trials are lacking. Blocking CD19 in a T-cell-mediated disease like PsA may seem counterintuitive, nevertheless, recent insights into the disease mechanism suggest B-cells also contribute to the pathogenesis of PsA. Additionally, by removing the B-cell-mediated immune activation, CAR T-cell therapy may also incidentally modify the pathogenic T-cell responses (4). This case presents a 74-year-old female with a past medical history of psoriasis diagnosed in 2005, PsA diagnosed in 2008, non-Hodgkin’s lymphoma stage IV status post R-CHOP and radiotherapy in 2012, who presented for management of active PsA. She was on an IL-17 inhibitor, and while her peripheral joint pain had improved she continued to complain of worsening axial pain. During follow-up with her oncologists for her malignancy history, she was found to have diffuse large B-cell lymphoma. The recommended treatment by oncology was anti-CD19 CAR T-cell therapy, which she began in 2023. Her PsA and lymphoma have since remained in remission off all therapy. This case highlights how CAR T-cell therapy reduces the inflammatory processes driven by autoreactive B-cells. By exhausting CD19 B-cells, it may be possible to reinstate the balance between pro-inflammatory and regulatory immune responses, thereby improving joint and skin symptoms in PsA. Ultimately, CAR T-cell therapy could offer a new treatment approach for PsA.

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Development of MDA-5 Dermatomyositis After Initiation of Adalimumab

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Abstract

TNF inhibitors are agents commonly used to treat inflammatory conditions such as rheumatoid arthritis. However, TNF inhibitors can also have a paradoxical effect in which they can induce autoimmune conditions such as dermatomyositis. We present the case of a 47 year old female who developed new-onset dermatomyositis after being initiated on adalimumab for treatment of rheumatoid arthritis.

A 47 year old female with a history of seronegative rheumatoid arthritis had been on treatment with methotrexate and sulfasalazine. Due to persistent inflammation, adalimumab was added to the patient's medication regimen for her rheumatoid arthritis. Approximately one month after starting treatment with adalimumab, despite improvement in rheumatoid arthritis symptoms, the patient noted new a onset rash with erythema present at her cheeks, nose, and periorbital region. This was initially thought to be secondary to possible drug eruption, however after consultation with dermatology, myositis labs were sent.

Lab work including aldolase and creatine kinase were within normal limits, however myositis specific panel was notable for MDA5 antibody with a titer >15. In light of these findings, patient was instructed to stop adalimumab due to concerns of TNF inhibitor induced dermatomyositis. CT chest was done and notable for focal reticular and groundglass opacities in the medial right lower lobe. PFTs were ordered, however still pending. The patient was switched to rituximab for further treatment as she still had ongoing synovitis.

This case highlights a rare yet clinically important phenomenon that may occur after initiation of TNF inhibitor therapies. The exact mechanism behind this is unclear, however it has been proposed that TNF inhibition promotes the expression of type 1 interferon which plays a role pathogenesis of dermatomyositis. Another hypothesis is that TNF inhibition interferes with apoptosis, leading to increased formation of autoantibodies contributing to autoimmunity. Specifically, as noted in this patient, MDA5 antibodies have been identified in patients with clinically amyopathic dermatomyositis and rapidly progressive lung disease. Thus, clinicians should be aware of the adverse effects of TNF inhibitor therapy, including the paradoxical effect of TNF-inhibitor induced dermatomyositis – regardless if there is muscular involvement or not.

Revealing the MAS(k) of Angioedema

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Abstract

This case highlights diagnostic uncertainty and early recognition of clinical deterioration. A 37-year-old female with history of HELLP syndrome, untreated hypertension, and latent tuberculosis on rifampin, presented to the ED for evaluation of periorbital and angioedema for two months in duration. Underwent maxillofacial CT demonstrating retropharyngeal fluid collection. ENT performed I&D revealing minimal fluid. Blood cultures positive for MSSA. Intravenous antibiotics given without resolution of angioedema. Rheumatology consulted. She was afebrile and hemodynamically stable without concerning cardiopulmonary findings, abdominal or CVA tenderness, or synovitis of large or small joints. An erythematous, papular rash was present. Lab work was significant for ANA 1:1280 titer in an atypical/speckled pattern with extractable nuclear antigen test positive for anti-SM/RNP, chromatin, and ribosome P antibodies, and, anti-RNP. p- and c-ANCA negative. Hypocomplementemia, bicytopenia (3.76 x 10^3/uL WBC and 4.01 x 10^6/uL RBC) present with ESR 61 mm/h, CRP 1.4 mg/dL. Urinalysis revealed minimal protein and no hematuria, in addition to electrolyte disturbances. For angioedema, CH50 and C1q returned low, consistent with acquired secondary process due to an undifferentiated connective tissue disorder, likely systemic lupus erythematosus (SLE). She discharged on prednisone 40 mg daily and HCQ 200 mg twice daily. Within 2 weeks, she was admitted for sepsis complaining of new fevers and general malaise, with worsening flank, joint, and pleuritic pain, malar rash, and angioedema. Antibiotics were discontinued in favor of high-dose intravenous corticosteroids for SLE flare. After several days of stability, she developed fevers, hypotension, tachycardia, and tachypnea. Labs showed ferritin 115,700 ng/ml, soluble IL-2 receptor 1781.9 pg/ml, and triglycerides 226 mg/dL. Hemophagocytic activity was confirmed on bone marrow biopsy; in addition to hepatic dysfunction and anemia met diagnostic criteria for MAS-HLH. No acute infection or malignancy identified as a secondary cause of HLH. Ferritin down-trended on cyclosporine and IL-1 receptor antagonist therapy. There are several etiologies of secondary HLH, including SLE and Adult-onset Still's disease. Interdisciplinary teams inclusive of Internal Medicine, Rheumatology, Infectious Disease, Hematology/Oncology, Interventional Radiology, and Pathology are utilized to secure the diagnosis and select the appropriate treatment. Angioedema can be misdiagnosed and SLE can be missed, prolonging time to diagnosis.

Rare case of systemic lupus erythematosus presenting as erythromelalgia

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Abstract

Erythromyalgia (EM) is a rare disorder that is characterized by episodes of increased temperature, erythema and burning pain that most commonly affects the distal extremities. It is under recognizied and can sometimes be missed as a preceding or clinical manifestation of systemic lupus erythematosus (SLE). EM can present as primary, usually due to neural dysfunction, or secondary due to etiologies such as myeloproliferative or autoimmune disorders. It is fairly rare without a definite diagnostic criteria and is mainly dependent on clinical history and physical exam findings. The pathophysiology is still unknown but a proposed theory is that EM is due to microvascular arteriovenous formation leading to tissue hypoxia which causes impaired blood flow based on temperature changes.

We present the case of a 26 year old female with no prior past medical history who presented to the hospital for pain in distal digits of upper and lower extremities bilaterally, including palms and soles of her feet. The pain was described as burning or “fire-type”, worse in warm temperatures and improved in the cold, with exam significant for erythematous rash on the distal digits, concerning for EM, initially unclear if primary or secondary. Lab work-up was remarkable for serological evidence of lupus given positive ANA and dsDNA, alongside cutaneous SLE on physical examination. During this time the patient also had subsequent acute onset chest pain that was concerning for non-ST elevation myocardial infarction and was started on heparin drip, nifedipine and lopressor with plans to undergo a left heart catheterization (LHC). The LHC was unremarkable aside from mild diastolic dysfunction, thus cardiac MRI was pursued that showed a mixed pattern of myocardial fibrosis with microvascular obstruction and near transmural fibrosis, which was concerning for lupus myocarditis. Hospital course was then complicated by declining kidney function with urine studies showing 2.6 grams of proteinuria, thus, given her age and presentation, a kidney biopsy was pursued which confirmed the diagnosis of lupus nephritis. She was started on mycophenolate, hydroxychloroquine, and pulse dose steroids, Solumedrol 500 mg IV for three days with oral taper. The patient’s burning pain in the distal digits did improve with steroids and, given her age, it was less likely that the EM was secondary to a myeloproliferative disorder over the underlying autoimmune disease. Thus, hematology and rheumatology recommended the patient be started on aspirin therapy for EM, and on follow-up she has been doing well on regimen of plaquinil, prednisone, mycophenolate, nifedipine and aspirin.

In conclusion, EM is a rare condition but an important finding that needs more awareness as it can be a preceding or only clinical manifestation of SLE. There are no current guidelines for the diagnosis or management of EM, and treatment is based on conservative management with trigger avoidance. However, it is important for providers to have early recognition of this disease as it may offer a better chance of remission and prevention of long-term complications as well as work-up for underlying etiology.

MIMICKING RHEUMATOID NODULES: A HIDDEN SIDE EFFECT OF MIGRAINE THERAPY

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Abstract

Introduction

Palisaded neutrophilic granulomatous dermatitis (PNGD) is one of the reactive granulomatous conditions that have been found in association with autoimmune disorders (e.g., rheumatoid arthritis, systemic lupus erythematosus), drug reactions (e.g., tumor necrosis factor inhibitors), malignancies (e.g., hematologic malignancies), and infections. PNGD includes conditions such as granuloma annulare, necrobiosis lipoidica, and rheumatoid nodules.

Case description

38-year-old white female, non-alcoholic and non-smoker with past medical history of migraine presents with painful nodules on dorsal surface of fingers and right feet for one year and noticed increase in size, frequency over the last 3 to 4 months. No history of autoimmune disease and recent infections. Physical examination notable for multiple nodules on dorsal surface of several proximal interphalangeal joints (PIP) as well as right foot. Her RAPID 3 (routine assessment of patient index data) scoring is 1 and the disease severity is near remission. Her lab work was overall unremarkable for any inflammatory arthritis as were her x-rays negative. She was referred to Ortho for biopsy of lesions. The biopsy of right index finger and ring finger soft tissue showed palisading granulomatous dermatitis necrobiosis with scattered neutrophils and basophilic debris, pathology reported it could be seen with rheumatoid nodules. She started on Prednisone taper 15mg once daily for 2 weeks followed by 10mg once daily for 2 weeks followed by 5mg once daily for 2 weeks. The patient had no improvement and on medication review it was found out that she takes Topiramate 150mg per day in divided doses for her migraine which has increased from100mg per day one-year ago. Topiramate was weaned down and discontinued, she had complete resolution of her soft tissue nodules. She currently takes Rimegepant for her migraine and follows neurology.

Discussion

This case illustrates Topiramate causing granulomatous dermatitis mimicking rheumatoid nodule in absence of autoimmune disease and is very rare. Discontinuation of Topiramate led to complete resolution of nodules underscoring importance of medication review.

Conclusion

This case emphasizes the role of medication induced etiology of soft tissue nodules difficult to distinguish from rheumatoid nodules from pathological perspective. Early recognition and withdrawal of medication can prevent unnecessary interventions and lead to complete resolution.

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Lupus Nephritis or ANCA Associated Vasculitic Nephritis: A Diagnostic Conundrum of Crescentic Glomerulonephritis

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Abstract

Introduction

Crescentic glomerulonephritis (CGN), or rapidly progressive glomerulonephritis (RPGN), is a critical renal condition marked by crescent formation in over half of the glomeruli and rapid kidney function decline. It arises from various causes, including immune complex-mediated and pauci-immune glomerulonephritis, the latter often linked to ANCA-associated vasculitis.Systemic lupus erythematosus (SLE), a systemic autoimmune disease, typically causes immune complex-related lupus nephritis, but in some cases, features overlap with ANCA-associated vasculitis, presenting with minimal immune deposits and positive ANCA tests. These overlapping presentations complicate diagnosis and treatment, particularly when CGN dominates the pathology.

Case Presentation

An 81-year-old man with a history of myelofibrosis, stage 3b chronic kidney disease, and neutrophilic dermatosis presented with acute kidney injury (creatinine 4.9 mg/dL from a baseline of 1.6), leg swelling, and decreased urine output. A review of prior records showed he had been diagnosed with CKD six months earlier, thought to be related to glomerulopathy from his underlying myeloproliferative disorder.

Urinalysis on this presentation revealed marked hematuria (RBCs >50) and proteinuria, with a protein-to-creatinine ratio of 1.17 (normal <0.1). Autoimmune testing showed positive antinuclear antibodies (ANA 1:160), low-positive anti-dsDNA, anti-histone antibodies, rheumatoid factor, and persistently low complement C3 levels. These findings led to a kidney biopsy, which revealed focally crescentic glomerulonephritis with mild immune complex deposition (IgM, C1q, and C3), but no significant proliferative changes or fibrinoid necrosis. The biopsy also demonstrated severe interstitial fibrosis, acute tubular injury, and numerous red blood cell casts.

During evaluation, the patient's kidney and lung function worsened, ultimately necessitating initiation of hemodialysis. He was started on high-dose pulse corticosteroid therapy, which led to marked improvement in both renal and respiratory function. At a follow-up visit four weeks later, he was clinically improved and no longer required dialysis.

Discussion

This case underscores the diagnostic challenges of crescentic glomerulonephritis with overlapping features of lupus nephritis and ANCA-associated vasculitis. The patient's presentation with rapidly rising creatinine, hematuria, and proteinuria prompted concern for a primary glomerular process. His autoimmune serology was notable for findings that are commonly associated with SLE. However, the kidney biopsy did not show the classic immune complex-mediated proliferative features typically seen in lupus nephritis. Instead, it demonstrated focal crescent formation with only mild immune deposits and absence of significant endocapillary hypercellularity or wire-loop lesions, findings more consistent with

pauci-immune glomerulonephritis.

The coexistence of lupus-related serologies and pauci-immune histology raises the possibility of an ANCA-lupus overlap syndrome, a rare but increasingly recognized entity. Although ANCA was not explicitly reported in this case, the pauci-immune biopsy pattern combined with the absence of dense immune complex deposition strongly suggests an ANCA-associated mechanism. The patient's rapid deterioration in renal and pulmonary function likely represented a vasculitic flare affecting multiple organ systems. Initiation of pulse-dose corticosteroids led to significant clinical improvement, highlighting the role of early, aggressive immunosuppression in such overlap cases, even in elderly or frail patients. Notably, the patient was able to discontinue dialysis within four weeks, reinforcing the potential reversibility of crescentic lesions when treated promptly.

A Breathless Clue: MPO-ANCA Vasculitis Through Respiratory and Renal Involvement

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Abstract

Objectives

Pulmonary-renal syndrome (PRS) is a rare but life-threatening condition characterized by diffuse alveolar hemorrhage (DAH) and rapidly progressive glomerulonephritis (RPGN). MPO-ANCA-associated vasculitis is a common etiology of PRS and can initially present with non-specific symptoms. This case highlights a patient with no prior vasculitis history who presented with PRS as the first manifestation of MPO-ANCA vasculitis.

Case presentation

A 61-year-old woman with known rheumatoid arthritis (on certolizumab), stage 3A chronic kidney disease, and interstitial lung disease (ILD) presented with acute kidney injury and systemic symptoms, including fever, vomiting, and respiratory decline. Laboratory workup and imaging studies were reviewed to determine the etiology of her condition. Bronchoscopy and immunologic testing were performed, while biopsies were deferred due to respiratory and hemodynamic compromise.

Results

Initial labs revealed a creatinine of 3.83 mg/dL, up from a baseline of 1.21 mg/dL. Imaging showed findings consistent with usual interstitial pneumonia (UIP). During hospitalization, she developed worsening hypoxia and hemoptysis. Bronchoscopy confirmed DAH. Autoimmune workup was positive for P-ANCA and anti-MPO antibodies, with negative PR3, anti-GBM, and anti-dsDNA antibodies. Urinalysis revealed hematuria, proteinuria, and granular casts, raising concern for RPGN. Treatment included corticosteroids, rituximab, plasmapheresis, and continuous renal replacement therapy. Her course was complicated by Aspergillus and Pseudomonas infections and ventilator-associated pneumonia. Despite maximal therapy, she succumbed to respiratory failure.

Conclusions

PRS should be considered in patients with bilateral pulmonary infiltrates and signs of renal dysfunction, particularly when accompanied by anemia, hematuria, or systemic symptoms. MPO-ANCA-associated vasculitis can present de novo with PRS. Early recognition, immunologic testing, and prompt treatment initiation are critical, even in the absence of biopsy. Infections remain a leading cause of mortality in these immunocompromised patients. Clinicians must maintain a high index of suspicion, especially in patients with underlying ILD or autoimmune disease.

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Mycophenolate Mofetil in the treatment of Patients with Isolated Cardiac Sarcoidosis

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Abstract

Cardiac involvement is seen in approximately a fourth of patients with sarcoidosis, with 25% having isolated cardiac sarcoidosis (iCS). Patients usually present with heart failure (HF), ventricular arrhythmias (VA) or atrioventricular block (AVB) and have worse outcomes compared to those with systemic sarcoidosis. Given known side effects with long-term glucocorticoids (GC), there is an increasing role for steroid sparing agents (SSA) like mycophenolate mofetil (MMF) in iCS treatment. However, most data focus on general cardiac sarcoidosis (CS), with limited information on iCS treatment and outcomes. In our study, we aimed to characterize a cohort of iCS patients to assess their clinical and radiologic outcomes in response to treatment with MMF.

Methods

We conducted a retrospective chart review of adult patients diagnosed with CS between January 2021 and January 2022, who were treated with MMF. Patients meeting the Japanese Circulation Society's 2016 criteria for iCS were included. Clinical outcomes included symptomatic worsening of HF, new VA or AVB, need for left ventricular assist device (LVAD), heart transplant (HT) or death. Radiologic outcomes were based on the change in myocardial uptake on PET scans and left ventricular ejection fraction (EF) on echocardiogram after treatment with MMF. Statistical analyses were performed using Microsoft Excel.

Results

Of 36 reviewed cases, 16 (44.4%) patients met criteria for iCS. The demographics and clinical presentation are detailed in Table 1. All patients received GC, followed by MMF as the first SSA (Table 2). Three (18.7%) patients developed MMF-related adverse effects requiring an alternative SSA. One (6.2%) and 5 (31.2%) patients had worsening HF and VA, respectively, while no patient required LVAD, HT, or died. Partial or complete cardiac resolution on PET scans within and after 1 year of diagnosis were seen in 7 (53.8%) and 9 (56.3%) patients, respectively. Only 1 (7.7%) patient had worsening EF on follow-up echocardiogram.

Conclusion

The true prevalence of iCS is difficult to determine due to its variable presentation and diagnostic challenges. However, our study shows a higher iCS prevalence (44.4%) among CS patients compared to the reported 25%, reflecting underdiagnosis. To our knowledge, this is the first study to explore MMF as an SSA in iCS. Our findings indicate that MMF, in combination with GC, is generally well tolerated and leads to radiologic improvement in most patients. Larger studies are needed to better characterize iCS and optimize treatment strategies to reduce its associated morbidity and mortality.

Sweet Confessions of a Myositis Mimicker

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Abstract

Introduction: Diabetic amyotrophy (DA) is a rare diabetic neuropathy that causes pain, atrophy, and weakness of the proximal lower extremity muscles, and thus, it can mimic idiopathic inflammatory myopathies (IIM).[1,2] Symptoms of DA are related to muscle denervation (from an immune-mediated microvasculitis causing ischemic nerve injury) rather than direct muscle injury.[1-3] Nevertheless, findings of systemic inflammation and edema on MRI can be seen in both DA and IIM.[1,2] In this case report, we present a patient with acute onset of myositis-like symptoms with no history of DM and later diagnosed with DA.

Case Presentation: A 77-year-old male with a history hyperlipidemia and hypertension was hospitalized for one month of bilateral anterior thigh pain and weakness. On exam, he had mild atrophy of his quadriceps with 4/5 strength. Pertinent labs included WBC 18.61 [4.86-11.05 10*3/uL], platelets 587 [150-500 10*3/uL], CRP 343 [≤4.9ng/mL], ESR 104 [≤20mm/hr], CPK 28 [≤190U/L], serum glucose 342 [74-109mg/dL], A1C 6.7% [4.8-5.9%]. Blood/urine cultures were negative. ANA, RF/CCP, SSA/SSB, ANCA, cN1A, HMGCR, and myositis specific antibodies were negative. MRI of the bilateral femurs showed diffuse, heterogeneous edema of the muscles of pelvis/thighs. EMG/NCS indicated a severe sensorimotor axonal polyneuropathy. A right quadriceps biopsy showed no evidence of endomysial or perivascular inflammation, infection or vasculitis.

Based on the patient's physical exam findings, elevated inflammatory markers, normal CPK, negative muscle biopsy and EMG/NCS indicating polyneuropathy, the patient likely had a systemic inflammatory process, and in the setting of persistent hyperglycemia, he was thought to have DA. He was treated supportively with acetaminophen, methocarbamol, and physical therapy and was started on insulin for new-onset DM. At his one-month follow-up, he had significant improvement of his weakness with minor residual sensory deficits.

Discussion: Early distinction between DA and IIM is critical because DA usually occurs in patients with new-onset diabetes mellitus (DM), and care for DA is largely supportive.[1-3] If systemic corticosteroids are used too early for suspected IIM, it may obscure the diagnosis of DA.

Conclusion: Workup for diabetes should be performed in patients presenting with proximal lower extremity pain and weakness as part of an IIM evaluation.

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