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Case Report: Rare Case of Systemic Lupus Erythematosus (SLE) Presenting as CIDP in Male

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Abstract

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare but recognized neurological manifestation of systemic autoimmune disorders, including Systemic Lupus Erythematosus (SLE). However, SLE presenting primarily as CIDP, especially in a young male, is extremely uncommon. We report a 32-year-old male with a previous diagnosis of CIDP who presented with progressive blackish discoloration of fingers and toes, sensory-motor neuropathy, alopecia, and acute dyspnea. Examination showed digital gangrene, mild muscle wasting, neuropathic deficits, and heart failure with reduced ejection fraction (LVEF 25%). Autoimmune evaluation revealed high-titer ANA (1:3200, speckled), strongly positive U1-RNP/Sm, positive anti-Sm, positive nucleosome and histone antibodies, low complements (C3/C4), and low-positive antiphospholipid antibodies.

Nerve conduction studies showed sensory-motor polyradiculoneuropathy. Echocardiography revealed global LV hypokinesia and pericardial effusion. The patient improved with high-dose steroids, anticoagulation, and standard heart failure therapy; EF improved to 49% on repeat echocardiogram. This case highlights the need to suspect SLE in males presenting with CIDP-like neuropathy, especially when systemic features evolve. Digital gangrene, myocarditis, and immune- mediated polyradiculoneuropathy may coexist as initial manifestations of lupus.

Keywords: SLE, CIDP, Gangrene, Heart Failure, ANA

Introduction

Systemic lupus erythematosus is a well explored autoimmune disease causing systemic inflammation and presents primarily as fatigue, fever, joint pain, a butterfly-shaped rash on the face, and hair loss. Other symptoms vary by individuals, affecting the skin (rashes, sun sensitivity), nervous system (headaches, memory loss, seizures), and organs like the kidneys and heart.[1] SLE is more common in women of reproductive age and disproportionately affects individuals of African and American descent. The sex predilection is 10 times more in women, having clear gender and race predilection, making other races and ethnicity less prone to and less reported in the literature.[2] The associated complications have also been more common in the susceptible races and ethnicities.

Neuro-lupus or the neuropsychiatric systemic lupus erythematosus (NPSLE), is very common in SLE patients and above 90% of SLE patients experience at least one neural manifestation. Common manifestations of neural SLE are headache, cognitive dysfunction, and mood disorders, more severe cases may show seizures, psychosis, and stroke.[3] These manifestations are due to both autoimmune inflammation and vascular issues, affecting the central and peripheral nervous systems. However, all these manifestations are reported in later stages of SLE and they are seen neither in the initial stages nor as a pathegnomonic feature. Therefore, chronic inflammatory demyelinating polyneuropathy (CIDP) as an initial manifestation is quite rarely reported in the literature.[4]

In CIDP, the immune system attacks the myelin sheath leading to progressive weakness, deficiency in coordination, neuropathic pain and tingling. It has been associated with many pathologies like diabetes, malignancies, chronic infections like hepatitis,HIV and also as adverse effects of medications. Idiopathic CIDP must be distinguished from secondary CIDP by identification of underlying pathologies.[5]

Coming to vascular pathology in SLE, interplay of autoimmune responses and antiphospholipid antodies (aPL) drives the vasculitis and thrombosis. It leads to inflammatory cell activation.[6]

Complement system involvement, and direct effects of aPL on the endothelium and coagulation.[7] Collectively these mechanisms lead to vasculitis and thrombosis. This can rarely lead to digital ischemia and consequent gangrene. With regard to cardiac involvement, it can manifest as pericarditis, myocarditis, cardiomyopathy.

However, all the manifestations spoken above are rarely seen as initial manifestation, that too in a male patient. The difficulty in diagnosis SLE is further amplifies especially in a misleading presentation as CIDP or peripheral neuropathy, digital gangrene and heart failure. CIDP as the initial presentation of SLE is extremely rare and often leads to diagnostic delay.[8,9] We present a unique case of male SLE presenting initially as CIDP, later complicated by digital gangrene and lupus

myocarditis. This paper highlights the importance for clinicians to recognize unusual multisystem presentations.

Case Report

A 32-year-old male, Indian, daily wage laborer, presented with a complaint of Blackish discoloration of fingers and toes for a duration of 3 weeks. He also reported pain and cold sensitivity in digits along with progressive lower limb pain and weakness. There was also decreased sensation in both feet. He clearly reported difficulty rising from squatting and climbing stairs. He previously had CIDP diagnosed in 2019 and was treated with steroids over 7-8 months with symptomatic improvement. On examination patient conscious, oriented, dyspneic and tachypneic at rest, pulse rate 110/mt, regular rhythm, low volume, peripheral pulses were normal, Blood pressure was 100/80 mmHg, Respiratory rate of 26/minute, no pallor, no pedal edema were seen. Alopecia, Digital gangrene of multiple fingers and toes, Small-muscle wasting of hands and feet, Tenderness of digits without warmth. Neurological examination showed sensory-motor deficits, reduced reflexes, inability to walk without support.

Routine investigation shows he had dimorphic anemia, Neutrophilic leukocytosis and Reactive thrombocytosis (5.5 lakhs), Renal workup showed Urine albumin to be 2-3+, Spot PCR to be 30–49 mg/g, 24-hour urine protein to be 1 g/day. (Renal biopsy was planned but deferred). Autoimmune Profile shows ANA (HEP-2) was positive, speckled (1:3200, 4+) U1- RNP/Sm was Strongly positive, Anti-Sm ws positive, Nucleosomes and Histones were positive, C3 and C4 were low (56 mg/dL & 9.8 mg/dL), Cardiolipin IgA/IgM was positive but low, Lupus anticoagulant was negative, p-ANCA was positive at 1:10, and c-ANCA was negative. Echocardiography showed Severe left ventricle systolic dysfunction (Ejection fraction 25%). Global hypokinesia and Mild-moderate pericardial effusion was seen. After treatment, the ejection fraction improved to 49% with mild pericardial effusion. Nerve Conduction Study showed sensory-motor polyradiculoneuropathy (demyelinating) Based on clinical feautures and investigations we diagnosed the patient as high-titer ANA, U1-RNP/Sm positivity, low complements, neuropathy, digital ischemia, myocarditis, and proteinuria, the final diagnosis was Systemic Lupus Erythematosus (SLE), complicated by Immune-mediated sensory-motor polyradiculoneuropathy Digital gangrene (likely vasculitic/immune-mediated) Lupus myocarditis with acute LV systolic dysfunction Low-grade proteinuria.

We treated the patient with Pulse methylprednisolone 1 g/day × 5 days, Heparin 5000 IU QID x 5 days, Aspirin 150 mg OD, Atorvastatin 10 mg HS, Enalapril 2.5 mg BD, Metoprolol 25 mg, Frusemide, Vitamin supplementation and supportive care. Anticoagulation was continued considering digital ischemia and positive anticardiolipin antibodies. Marked improvement in dyspnea was seen. Ejection Fraction improved from 25% to 49%. Stabilization of digital gangrene was achieved with no new lesions. Neurological symptoms improved gradually and Renal parameters stabilized. The patient was discharged with immunosuppression plan and follow-up in rheumatology and neurology

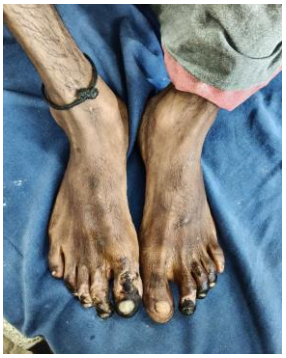


Figure 1 & 2: Digital gangrene

Figure 3: Digits of hand

Figure 4: Alopecia

Discussion

This case report has presented an exceptionally rare and diagnostically challenging case of SLE in a young male. It initially showed itself as a CIDP in the past as shown by the medical history. The underlying cause was unrecognized and hidden for several years. When it presented itself in a more severe and complicated form of digital gangrene and acute lupus myocarditis. The salient point here is the while neurological symptoms are well-known in SLE, the demyelinating polyradiculoneuropathy is quite rare and uncommon. More so if it presents as the first or dominant manifestation of SLE. This rarity is further accentuated by demographic profile of the patient who is a male and Asian.

Peripheral neuropathy in SLE occurs in about 2–8% of patients commonly involving axonal or small-fiber neuropathy, while demyelinating neuropathy that presents like CIDP is very rare.[10] Such demyelinating neuropathy of SLE can resemble idiopathic CIDP even after investigations leading to a delayed diagnosis, as seen in the medical history of this case. Earlier symptoms were effectively managed with steroids that might have transiently suppressed the evolving lupus.

In current presentation, it was observed after biochemical investigations that the CIDP was not idiopathic and had an entire plethora of changes remaining hidden in the past. Alopecia, Raynaud-like symptoms, proteinuria, and very high-titer ANA with SLE-specific antibodies (viz. anti-Sm, nucleosome, histone) have offered strong evidence that points to CIDP being an early neurologic manifestation of SLE.[11] More specifically, immune- mediated neuropathy in lupus may be the consequence of autoantibody-driven myelin injury and the vasculitis affecting the vasa nervorum. This may explains the response of conventional CIDP treatments being effective in this case but did not permanently cure the disease.

Digital gangrene is the result of ischemia and is rare in SLE. This patient showed low-positive anticardiolipin antibodies, however, the symptoms combining immune markers, cold sensitivity, and rapid progression strongly shows immune-mediated vasculopathy. Intact peripheral

pulses confirm microvascular origin of ischemia.[12]

Cardiac involvement in SLE may be common, but myocarditis accounts occurs only in less than 1 in 10 cases. It shows rapid progression of heart failure and therefore is of significant morbidity.[13] The patient's significant lowering of ejection fraction (25%), global hypokinesia and pericardial effusion, strongly confirm lupus myocarditis. The swift improvement in ejection fraction due to high-dose corticosteroid therapy shows its immune- mediated origin.

From the diagnostic point of view, high ANA titer and specific SLE-associated markers clinched the diagnosis. The presence of strong U1-RNP/Sm positivity may cause a doubt about overlap syndromes which was solved by absence of anti-Scl-70, anti- centromere antibodies. Clinical features could eliminate mixed connective tissue disease. Low complement levels aided in confirming the immune complex–mediated disease.

Coming to the treatment aspect, it needed both control of neuroinflammation, vasculitis and myocarditis along with antiphospholipid-mediated ischemia. Administration of High-dose methylprednisolone led to rapid improvement on multiple aspects. Anticoagulant therapy was for digital ischemia and low-positive antiphospholipid antibodies. Long-term management may require steroid-sparing immunosuppressants depending on organ progression.

This case clearly highlights effect of treating recurrent neuropathy as CIDP relapse. Had investigations have not been carried out for systemic autoimmune causes, it may have led to delayed recognition of life- threatening lupus manifestations.

Unusual Initial Presentation in a Male SLE calls for High Vigilance, especially when neuropathy, myocarditis, and digital ischemia occurring together. In this aspect Digital Gangrene is an alarming Early Sign. Here the Importance of Comprehensive Autoimmune Evaluation in Atypical CIDP is clearly shown and Immunosuppression along with conventional CIDP therapy is shown to bring in successful recovery. This case clearly confirms that neuropathy with other systemic signs requires immediate investigation for autoimmune disease.

Conclusion

This case highlights the importance of considering systemic autoimmune diseases like SLE in patients-especially males-presenting with CIDP-like neuropathy. Early identification prevents delays in diagnosis and organ damage. Multisystem involvement such as myocarditis and digital gangrene may be the first clue pointing toward lupus.

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