Original Research Paper

Volume - 13 | Issue - 02 | February - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Rheumatology

SYSTEMIC LUPUS ERYTHEMATOSUS WITH MYOSITIS OVERLAP SYNDROME IN MALE: A RARE CASE

Rajkumar Sharma*	Department of Internal Medicine, Army Hospital (R&R), New Delhi. *Corresponding Author
Rajat Shukla	Department of Internal Medicine, Army Hospital (R&R), New Delhi.
Anuj Singhal	Department of Internal Medicine, Army Hospital (R&R), New Delhi
KK Kompella	Department of Internal Medicine, Army Hospital (R&R), New Delhi
G Shivaram	Department of Internal Medicine, Army Hospital (R&R), New Delhi

ABSTRACT Systemic lupus erythematosus (SLE) is an inflammatory systemic disease that damages organs by depositing autoantibodies and complement-activating immune complexes or by vascular occlusion resulting from procoagulant states linked with antiphospholipid antibodies(APLA). SLE is a chronic autoimmune disorder that affects women of childbearing age in the vast majority of cases [5]. SLE is diagnosed by the presence of clinical manifestations (clinical domains) and auto-antibodies (Immunological domains). SLE is becoming more prevalent. Here, we report a rare case of a young male with SLE who presented with add-on features of inflammatory myositis.

KEYWORDS : overlap syndrome, systemic lupus erythematosus, inflammatory myositis, corticosteroid

Introduction:

The term overlap syndrome includes conditions characterized by the simultaneous occurrence in the same patient of signs, symptoms, and immunological features of two or more CTDs. In SLE, generalized myalgia and muscle tenderness are frequent signs & symptoms, particularly when they experience exacerbations.[2] According to Kelley's Textbook of Rheumatology, the overlap syndrome (SLE with Inflammatory myositis) has been reported to occur in 5% to 11% of patients with SLE and may be developed at any time during the course of the disease[1]. Myositis is a rare complication of systemic lupus erythematosus (SLE) and occurs in almost 4–16% of cases of SLE.[3] There have been various associations between inflammatory myopathies and the presence of malignancies, but the etiology of this is controversial. SLE is a chronic autoimmune disorder that affects women of childbearing age in the vast majority of cases [5]. It typically affects younger women, but up to 20% of patients 50 years of age or older may experience it. Systemic lupus erythematosus (SLE) affects almost every body system to varying degrees [6]. Many latest studies have shown that systemic lupus erythematosus (SLE) is becoming more prevalent. The ratio of female-to-male ranges from 4.3 to 13.6.[7] The Lupus Foundation of America estimates that approximately 5 million individuals worldwide have some form of Lupus. A young male with overlap syndrome (SLE & inflammatory myositis) is the subject of this case report. This article discusses the challenges encountered during the diagnosis and treatment of such a case.

Case History

A 35-year Male, presented with generalized fatigue, muscular weakness, and myalgia involving upper & lower limbs muscles, lower limbs oligoarthralgia, skin rash over the right arm & upper back, and frothyuria for two months. He had no history of chest pain, palpitation, syncope, cough, expectoration, flank pain, hematuria, ascites, altered sensorium, loss of consciousness, seizure, weight loss, blood loss, altered bowel & bladder habit, recurrent oral ulcers, sicca symptoms and consumption of alternative medications. No history of similar illness in the past and family members.

On examination, he was sick looking male, conscious, and well-oriented to time, place, and person. He was afebrile and his vitals were normal. He had pedal edema, periorbital puffiness, petechial over the hard palate (Figure 1(a)), and old-heeled hyper-pigmented skin rashes seen over the lateral aspect of the right arm and right scapular region (Figure 1(b)). He had also muscle tenderness in both thighs. Central nervous system (CNS) examination revealed hypotonia in all four limbs present (lower limbs more than upper limbs), with symmetrical, proximal grade IV muscle weakness but no muscle wasting. His MMT Score was 118/150. There was no evidence of involvement of the sensory system and cranial nerves. Other systemic examinations were within normal limits.

He was founded to have pancytopenia (Hb-8.3gm/dl, TLC 1200/cumm, Platelet– 46000/cumm). PBS showed microcytic and normocytic hypochromic (McHc + NcHc) RBCs, no evidence of hemolysis, and a retic count of 1%. His urine analysis revealed protein 2+, RBCs 2-3, with spot uPCR of 1.99, and 24hrs urine protein was 9534 mg/day. His renal function test (BUN/Creatinine) was within normal limits. He had transaminitis with a maximum AST/ALT level - 843/178 U/L with normal other metabolic parameters. His viral markers were negative and coagulation parameters & thyroid profile were within normal limits. He has raised LDH- 1369 U/L, CPK- 6703 U/L, CRP- 1.16 mg/dl and ESR- 108 mm fall in 1st hr. Immunological workup showed DCT 2 +, ANA by IIF positive with AC-1 nuclear homogeneous /AC-19 cytoplasmic dense fine speckled seen, ENA by LIA showed dsDNA (++), nucleosomes (+++), Rib-PO (+++), RO (++) with normal immunoglobulin profile, C3/C4 level- 0.101/0.0219 gm/l (hypocomplementemia) and RF (rheumatoid factor) was normal. Serum myositis profile (16 antigens) revealed positive (+) Ku antigen.

USG Abdomen, chest X-ray, and CECT (chest & abdomen) showed no significant abnormality. Bone marrow aspirate revealed normoblastic erythroid series, no atypical cells and biopsy showed reactive bone marrow. His blood and urine cultures were negative. ECG was normal sinus rhythm(NSR) and 2DEcho showed EF 60%, no RWMA, valves normal, and no clot/vegetation /pericardial effusion. Renal biopsy suggested focal lupus nephritis(LN) and membranous nephropathy (ISN/RPS- LN Class III+IV) without significant tubule-interstitial chronicity. MRI of bilateral thighs and legs showed features suggestive of diffused myositis involving all compartments muscles without atrophy (Figure 2). His workup for malignancy including tumor markers was negative.

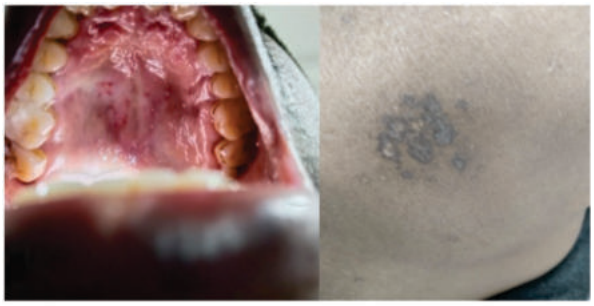
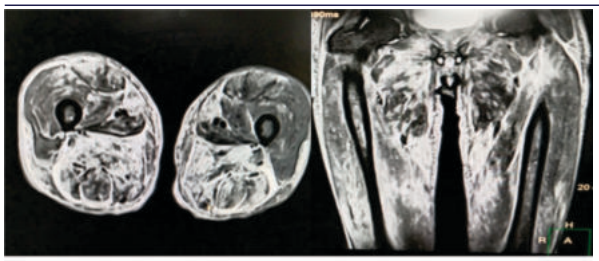


Figure 1: Image of (a) petechial rash over the hard palate and (b) heeled hyper-pigmented rash over the scapular region



(a) (b)

Figure 2: Image of (a) MRI bilateral thighs and legs (STIR axial and coronal section) showing diffuse hyperintensity involving all visualized muscles with minimal subcutaneous and intermuscular edema

The patient was diagnosed with a case of inflammatory polymyositis with SLE. During the hospital stay, the patient was treated with IV methylprednisolone pulse 1gm IV for 3 days followed by oral steroid prednisolone 60mg daily (1mg/kg/day) along with IV pulse cyclophosphamide therapy (as per NIH protocol), HCQ 200mg, and Losartan 50mg and supportive therapy. With treatment, his weakness and other parameters were getting better.

DISCUSSION:

Generalized myalgia and muscle tenderness are frequent signs and symptoms, particularly when exacerbating SLE. A drug-induced myopathy secondary to steroids, DMRDs, statin, and other drugs is a cause of proximal myopathy in patients with SLE. Concurrent hypothyroidism can cause proximal myopathy and also elevate creatinine phosphokinase (CPK) levels. Inflammatory and drug-induced myopathies can be distinguished by characteristics such as muscle biopsy, electromyography (EMG) study, and raised enzyme level (CPK or aldolase). Patients with myositis in SLE have less typical histological features than idiopathic polymyositis. Histological features in patients with myositis are muscle atrophy, microtubular inclusions, and a mononuclear cell infiltrate. The necrosis of muscle fibers is not a common feature, but deposition of immunoglobulin is virtually always present despite less frequent concurrent inflammation. Patients with connective tissue diseases(CTDs) including SLE can be presented with normal enzyme(CPK) levels; in presence of signs and symptoms of myositis with normal creatinine phosphokinase(CPK) enzyme level should not stop the physician from diagnosing the myopathy. In diagnosing myositis in CTD patients, both features of myositis and raised enzyme levels are crucial. Patients with SLE can also manifest with skin lesions of dermatomyositis. Patients with SLE have reported chest pain or discomfort due to costochondritis; however, other illnesses such as angina pectoris, pericarditis, and esophageal spasm should first be ruled out.

It has been known that inflammatory myopathies-related malignancies might manifest many years after the diagnosis of muscle illnesses; hence, continue surveillance and repeated routine screening for malignancy are recommended. On average, the prevalence of malignancy in association with inflammatory myopathies has been approximately 25%.[8] Although it appears to be lower than in dermatomyositis, the relative risk of acquiring internal malignancies in patients with polymyositis is consistently increased than would be predicted in the general population. Many common malignancies areas are related to inflammatory myopathies, such as stomach, ovarian, and breast adenocarcinomas. The risk of malignancies increases in a patient with polymyositis or dermatomyositis. In patients with dermatomyositis, there is also a higher rate of mortality from malignancies[9].

This association is rare with the predominance in females, the various clinical presentations, and the benign course of myositis. Certainly, there are undoubtedly differentiations between overlap and non-overlap patients (SLE & myositis). The overlap (SLE & myositis) should not be considered a mild disease and treated as more aggressively as primary myositis[10]

CONCLUSION:

A high index of clinical suspicion is to be kept while dealing with multisystem diseases. Generalized myalgia and muscle tenderness are frequent problems particularly when exacerbations of SLE. The drug-induced and concurrent hypothyroidism were included in the differential diagnosis of proximal myopathy in SLE. Inflammatory and drug-induced myopathies can be distinguished by characteristics such as muscle biopsy, electromyography (EMG) study, and raised enzyme level (CPK or aldolase). Inflammatory myositis in patients with SLE has less typical histological features than idiopathic polymyositis.

REFERENCES

1. Sreejith V Ravi, CJ Selvakumar, Inflammatory Myositis-Secondary to SLE, Journal of the association of physicians of India; vol 77,2019
2. Gary S. Firestein, Ralph C. Budd, Sherine E. Gabriel, Iain B. McInnes, James R. O'Dell, Kelley's Textbook of Rheumatology,2013,
3. Hilton-Jones D, Squier MV. Risk of cancer in dermatomyositis or polymyositis (letter). N Engl J Med 1992; 327:207-8.
4. Harvey AM, Shulman LE, Tumulty PA, et al. Systemic lupus erythematosus: a review of the literature and clinical analysis of 138 cases. Medicine 1954; 33:291-437.
5. Schroeder JO, Euler HH. Recognition and management of systemic lupus erythematosus. Drugs 1997; 54:422-34.
6. Samuel L Gurevitz, Jennifer A Snyder, Systemic lupus erythematosus: a review of the disease and treatment options, 2013 Feb;28(2):110-21.
7. Petri M. Epidemiology of systemic lupus erythematosus. Best Pract Res Clin Rheumatol 2002; 16:847-858.
8. DeVere R, Bradley WG. Polymyositis: its presentation, morbidity, and mortality. Brain 1975; 98:637-66.
9. Sigurgeirsson B, Lindelof B, Edhag O, Allander E. Risk of cancer in patients with dermatomyositis or polymyositis. Apopulation-basedd study. N Engl J Med 1992; 326:363-7.
10. Fatma Maazoun, Faten Frikha, Systemic lupus erythematosusmyositis overlap syndrome: report of 6 cases, 2011 Sep 28.