

A Rare Presentation of Acute Neck Muscle Weakness in A Male Patient with Systemic Lupus Erythematosus

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Abstract

Introduction: Systemic lupus erythematosus is a chronic autoimmune disease which may affect every organ in the body. According to reports, 5% to 11% of SLE patients experience inflammatory myositis that affects the proximal muscles. This condition can appear at any point throughout the disease's progression. Additionally, it may develop as a result of internal cancers. **Methodology:** We hereby report a 57 year SLE patient, on hydroxychloroquine who presented with neck muscle weakness for one week duration. Imaging revealed myositis in the neck region and autoimmune profile positive ANA, Anti SSN, Ro 52, and dsDNA and low serum complement levels. Other investigations were within normal limits. **Results:** Hence the diagnosis of inflammatory polymyositis secondary to systemic lupus erythematosus was made and patient was managed successfully with steroids, hydroxychloroquine and immunosuppressants. **Conclusion:** Proximal muscle weakness in SLE may be due to a drug-related myopathy secondary to corticosteroid, statin medications use or antimalarial, concurrent hypothyroidism or secondary to SLE. Thus prompt evaluation can be life saving to the patient.

Keywords: Systemic lupus erythematosus, muscle weakness, inflammatory myositis.

INTRODUCTION

A chronic, multi-dimensional autoimmune inflammatory condition that may affect any area of the body is systemic lupus erythematosus (SLE)^[2]. SLE has a number of presenting characteristics and manifestations but no definitively identified aetiology. Improved diagnosis techniques and increased awareness of the condition have increased the number of cases that have been identified significantly in recent years. Depending on the region of the body is affected, symptoms might be mild, moderate and severe^[2].

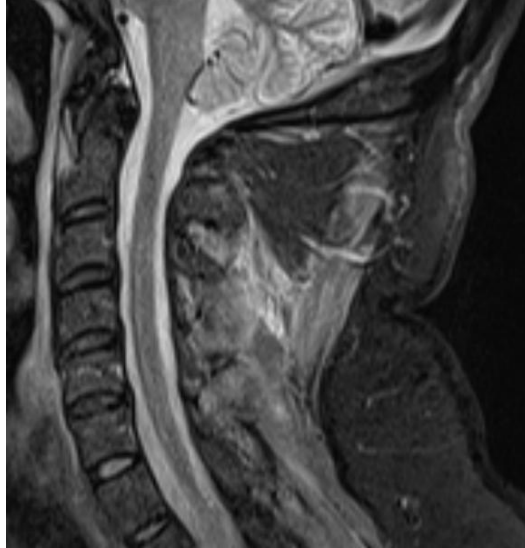
Generalized muscle soreness, muscle tenderness, and muscle weakness are common symptoms in SLE patients. Proximal muscle inflammation in myositis affects 5% to 11% of patients. And it may appear at any stage of the illness^[1]. The origin of this connection is still up for debate, despite certain links between inflammatory myopathies and systemic lupus erythematosus^[1]. In this case report we describe the example of a woman who had inflammatory myositis as a result of SLE.

CASE REPORT

A 57-year-old female was brought with complaints of progressively increasing, symmetrical muscle weakness and stiffness of neck region for one week duration. Patient was a known case of systemic lupus erythematosus on tablet hydroxychloroquine with no other known comorbidities. On clinical examination, generalised muscle wasting was seen. No cutaneous markers suggestive of dermatomyositis were present. Muscle tenderness over the neck region was present. In proximal muscles of neck, the power ranged from grade 3 to 4. Deep tendon reflexes were sluggish and there was no sensory system and cranial nerve involvement. Other system examination was normal. Baseline investigations, complete blood count and biochemistry analysis were normal. ECG and chest radiograph were normal. CRP was 3.3. She tested negative for HIV, HbsAG and HCV. We then investigated her for muscle disorders secondary to autoimmune and neoplastic causes. CPK value 3916, LDH -456, MRI neck

was done which showed myositis. Thyroid function tests were within normal limits. Autoimmune profile was done and revealed positive ANA, Anti SSN, Ro 52, and dsDNA while Anti Sm, Anti – Jo 1, PCKA, CENP were negative . The serum complement level was low. Anti-cardiolipin Ab and Lupus anticoagulant was negative. Thus, the diagnosis of inflammatory polymyositis secondary to systemic lupus erythematosus was arrived at. Patient was managed conservatively with steroids, hydroxychloroquine and immunosuppressants. The weakness improved with treatment.

Figure 1: MRI neck of the patient with systemic lupus erythematosus who came with acute neck muscle weakness



MRI neck of the patient showing hyperintensities in the paravertebral region denoting myositis.

DISCUSSION:

Being an autoimmune disorder, SLE is marked by microvascular inflammation in multiple systems of the body and production of various types of autoantibodies, specifically antinuclear antibodies². It can affect people of all ages, sexes, and ethnicities. But SLE presents itself in the reproductive years in greater than 90% of cases^[2].

The term myositis be it polymyositis or dermatomyositis refers to a cohort of patients whose primary clinical feature is muscle weakness, which is frequently accompanied by agony in the muscles, wasting, tenderness, or other forms of connective tissue diseases^[6]. Biopsy from the muscle typically reveals patchy necrosis of the muscle fibres along with interstitial or/and perivascular cellular infiltrates. Inflammatory myositis associated with overlap syndromes has been linked to systemic sclerosis (SSc), rheumatoid arthritis (RA), Sjögren's syndrome, or systemic lupus erythematosus (SLE), which generally occurs in bouts as remissions and relapses^[6].

However, true myositis is relatively uncommon, unlike myalgia, which is complained of by 50% of patients with systemic lupus erythematosus. The former, can develop prior to or after the onset of SLE, or both diseases may be present intermittently⁶. In an examination of 30 cases, Garton and Isenberg found that patients with overlap myositis associated with SLE and those with myositis of primary variety have very similar clinical characteristics. On extended follow-up, there were no appreciable differences in morbidity or mortality, and the clinical trajectory for the two groups was largely identical^[3].

Younger ages are recorded for those with SLE myositis, and length of the illness is negatively correlated for myositis in SLE, indicating that the condition may present early. Dermatomyositis has been more frequently documented, even though polymyositis has also been linked to SLE⁹. Both the upper and lower limbs are affected by the symmetrical, proximal muscular weakness. Trunk muscles are commonly seen in advanced phases. When myositis in SLE is treated promptly, patients have been demonstrated to have a decreased incidence of respiratory muscle involvement, neck flexor weakness, and pharyngeal paralysis^[9].

The clinical features and lab results of 290 patients with SLE without myositis and 10 patients with SLE complicated by myositis were compared in a recent study⁴. According to the findings, erosive joint disease, baldness, mouth ulcers, and pulmonary illness were commoner in patients with myositis associated with SLE overlap, but renal dysfunction was rarely noticed. The study also discovered that those with SLE/myositis overlap were more prone for death at younger ages, the anti RNP autoantibodies playing an important role in the course of their illness⁴.

Myositis specific antibodies are found in high rates in patients with polymyositis, dermatomyositis and their associated overlap syndromes namely anti-Ro/SSA, anti-U1 RNP, anti-La/SSB, and anti-Sm⁵. Autoimmune rheumatic diseases must be thoroughly pondered into when encountered with a strong positive ANA⁵.

In individuals with SLE, elevated blood CK was observed to be associated with underlying myositis, and these patients also more frequently experienced proximal weakening symptoms and/or anti-RNP antibodies⁷. Type II fibre atrophy and artery wall thickening, on the other hand, did not correlate with any of the clinical or laboratory variables examined and seem to be more general findings⁷.

Although the appearance of muscle complaints in systemic lupus erythematosus is relatively prevalent, diagnosis of inflammatory myopathy is rare⁸. Few reports of muscle biopsy data exist, and those that do tend to describe myofiber necrosis or regeneration or perivascular mononuclear cell infiltrates⁸.

Since many of the medications for myositis are also used to treat other autoimmune illnesses, the treatments are essentially the same. First-line therapies often included corticosteroids, and other immunosuppressive drugs were frequently employed⁶. Myositis prognosis varies considerably, and overlap syndrome SLE/myositis should be treated similarly to IIM in terms of prognostic variables. Age of the patient, degree of myositis, the presence of cardiopulmonary disease or dysphagia and the patient's early reaction to corticosteroid therapy constitute a few of these⁶.

Glucocorticoids continue to be the backbone of treatment for SLE with myositis overlap, and care is the same as for inflammatory myositis⁹. The majority of SLE patients respond well to glucocorticoids, but they will need an alternative DMARD to taper off steroids properly. An alternate immunosuppressant medication is urgently needed in individuals with SLE who have not responded adequately to steroids, who have experienced an adverse pharmacological reaction to steroids, or who have any other major organ involvement. In cases of refractory myositis with SLE, methotrexate, azathioprine, cyclosporine, cyclophosphamide, and MMF are typically recommended second-line immunosuppressants⁹.

CONCLUSION

With overlap syndromes, the heterogeneity of SLE's presentation and progression becomes more complex, and the key to addressing this complexity is routine clinical follow-up with attention to iatrogenic consequences, comorbidities, and infections that mimic disease activity. For a better knowledge of the natural history, prognosis, and necessary therapeutic approach for overlapping syndromes, specialised clinical trials in patients with overlap syndromes are needed⁹.

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ETHICAL CONSENT

Patient included in the study had provided informed consent.

FUNDING

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CONFLICT OF INTEREST

The authors declare that was no conflict of interest.

REFERENCES

1. Inflammatory myositis- Secondary to SLE , Sreejith V Ravi , Selvakumar .MSacrates, Shobhana , Arjunan Mahesh , JAPI August 2019 Volume 67 ISSN 0004-5772
2. Manifestations of Systemic Lupus Erythematosus
Manole COJOCARU, MD, PhD,a Inimioara Mihaela COJOCARU, MD, PhD,b Isabela SILOSI, MD, PhD,c and Camelia Doina VRABIE, MD, PhDd
3. Garton MJ, Isenberg DA. Clinical features of lupus myositis versus idiopathic myositis: a review of 30 cases. *Br J Rheumatol.* 1997;36:1067–74. [PubMed] [Google Scholar]
4. Dayal NA, Isenberg DA. SLE/myositis overlap: are the manifestations of SLE different in overlap disease? *Lupus.* 2002;11:293–8. [PubMed] [Google Scholar]
5. Mimori T. Structures targeted by the immune system in myositis. *Curr Opin Rheumatol.* 1996;8:521–7. [PubMed] [Google Scholar]
6. Maazoun F, Frikha F, Snoussi M, Kaddour N, Masmoudi H, Bahloul Z. Systemic lupus erythematosus-myositis overlap syndrome: report of 6 cases. *Clin Pract.* 2011;1:e89.
7. Lim LM, Abdul-Wahab R, Lowe J, Powell RJ. Muscle biopsy abnormalities in systemic lupus erythematosus: correlation with clinical and laboratory parameters. *Ann Rheum Dis.* 1994;53:178–82.
8. Miller JB, Paik JJ. Overlap syndromes in inflammatory myopathies. *Curr Treat Options in Rheum.* 2017;3:289–98.
9. Shah S, Chengappa K G, Negi VS. Systemic lupus erythematosus and overlap: A clinician perspective. *Clin Dermatol Rev.* 2019;3:12–7.