Co-occurrence of Polymyositis and systemic lupus erythematosus: Polymyositis-systemic lupus erythematosus overlap syndrome: a case report from Ethiopia

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Introduction

The term *overlap syndrome* includes a large group of conditions characterized by the coexistence of signs, symptoms, and immunological features of two or more connective tissue diseases occurring simultaneously in the same patient.1 It is very rare to see an overlap of inflammatory myositis and SLE and it has not been well studied.[2] With variable epidemiological data, most often diagnosed concurrently with SLE.[3] In contrast to myalgia which can affect nearly half of patients with SLE, true myositis is relatively rare as shown by some studies.[11]Myositis can occur before, after SLE, or sporadically both diseases can be present simultaneously.[10]

Myositis associated with overlap syndromes is usually of paroxysmal variety and has been associated with one or another of connective tissue disorders.[1] Raised serum creatine kinase is found to correspond with underlying myositis in patients with SLE.[12] Furthermore, the presence of myositis-specific antibodies such as anti-U1RNP, anti-Ro/SSA, anti-La/SSB, anti-Sm or anti-PM-Scl is suggestive of an overlap myositis.[13,12]

Case presentation

History and physical examination

A 28-year-old female patient, 2 years back diagnosed with polymyositis and interstitial lung disease after she was historically presented with diffuse bilateral joint pain, extremities weakness which she described as difficulty with combing her hair, walking upstairs, and difficulty swallowing. The symptoms stayed for nearly one year before she sought medical attention. On physical examination which was done 2 years back during her first visit; There was mild tenderness over bilateral hand, wrist, elbow, ankle, and foot joints, with a passive limited range of motion.

Investigation and treatment course

Investigations during her first visit showed elevated CK-total(2227IU/L), elevated anti-PM scl-75. High-resolution chest CT; bilateral septal thickness, few honeycombing patterns (NSIP) with bilateral pleural effusion, and Echocardiography showed borderline left ventricular wall hypertrophy, and EKG was normal.

With the diagnosis of polymyositis, the patient was initially put on 1mg/kg of prednisolone, tapered over more than a month, and continued with 10mg oral daily dose and azathioprine 100mg oral daily. The symptoms have subsided over time.

Currently, she presents with a 2-month history of worsening joint pain. 2 weeks ago she had a decreased amount of urine and cola-colored change of urine and bilateral flank pain. The current physical finding showed elevated blood pressure i.e. 170/90mmhg, pale conjunctiva, and bilateral costovertebral angle tenderness. Recent investigations have shown moderate anemia hemoglobin of 9.2g/dl, positive ANA qualitative study,

total CK 2154IU/L(more than 15x elevated), decreased C3 level (0.275g/dl), normal c4 (0.275g/dl), and elevated ant-dsDNA-IGG (141.7 IU/L). The investigation summary is shown in Table 1 below

With the final diagnosis of Polymyositis-SLE overlap syndrome, she was started on induction therapy with high dose methylprednisolone followed by prednisolone and cyclophosphamide.

Outcome and follow-up

At the 4th month of induction therapy with methylprednisolone and cyclophosphamide, her creatinine normalized and proteinuria resolved. The patient is currently on her maintenance therapy with azathioprine, chloroquine and low-dose prednisolone with complete clinical and laboratory remission..

Discussion

Myositis identifies a group of patients in whom muscular weakness is the principal clinical feature often associated with muscle pain, tenderness, wasting, or other forms of connective tissue diseases; the muscle biopsy generally demonstrates areas of muscle fiber necrosis accompanied by interstitial and/or perivascular cellular infiltrates.1 Its incidence is estimated to be around 4.27–7.89 cases per 100,000 people per year.[5] Inflammatory myositis (IM) is broadly classified into five categories: polymyositis, dermatomyositis, immune-mediated necrotizing myopathy, sporadic inclusion body myositis, and overlap myositis.[6] Clinically, a patient can present with acute or subacute-onset muscle weakness of different patterns, often accompanied by raised creatinine kinase (CK).[7]

In severe cases, respiratory and oesophageal muscles can be affected.[8] The diagnosis of inflammatory myositis (IM) requires careful clinical evaluation paired with serological markers, neurophysiological testing, and a muscle biopsy. In history taking, we need to take a relevant family history, myopathic drugs, alcohol, and features of endocrinopathy.[9]

Myositis associated with overlap syndromes is usually of paroxysmal variety and has been associated with one or another of connective tissue disorders.[1] Raised serum creatine kinase is found to correspond with underlying myositis in patients with SLE.12Furthermore, the presence of myositis-specific antibodies such as anti-U1RNP, anti-Ro/SSA, anti-La/SSB, anti-Sm or anti-PM-Scl is suggestive of an overlap myositis.[13,12]Myositis, lymphocytic vasculitis, type II muscle, atrophy, vessel wall thickening, and vacuolar myopathy are various histopathological findings observed in the muscle biopsies of patients with SLE.[14]However, histopathological findings of lymphocytic vasculitis and/or myositis are confirmatory of true myositis in SLE.[12]

SLE is an autoimmune disease that affects virtually any organ in the body. It affects females more commonly than males.4 Skeletal muscle complications are common in SLE patients, which are seen in the form of myalgia, muscle weakness, and atrophy.15 Myalgia is the most skeletal muscle manifestation, affecting 40–80% of patients.[16]

Treatment can prove difficult, as both conditions respond to a variety of immunosuppressive and cytotoxic agents. Corticosteroids were used as first-line therapy, and additional immunosuppressive agents such as cyclophosphamide, methotrexate, rituximab, and mycophenolate mofetil have been used with varying degrees of clinical response and remission rates.[13,17]

The reports on the prognosis of SLE-myositis overlap syndrome are conflicting. Some of them suggest that it follows a benign course, while others suggest no difference between them in terms of morbidity and response to therapy.[13,18] Again, another study suggests that those with SLE-myositis overlap syndrome have a poorer outcome with early death.[14] Although myositis in SLE responds well to corticosteroids, the association of pulmonary involvement leads to early mortality with an increased standardized mortality ratio compared to SLE alone. This indicates a poor prognosis for SLE-myositis overlap syndrome.[19]

Our patient was initially diagnosed with polymyositis 2 years ago after she presented with characteristic proximal muscle weakness and difficulty swallowing with associated multiple joint pain, and laboratory investigations revealed elevated CK-total (2227 IU/L) and anti-PM scl-75 (elevated). With an initial treatment

with corticosteroids and a continual treatment with azathioprine, the patient showed symptomatic improvement. After 2 years of treatment, she presents with a history of worsening joint pain, a decreased amount of urine, a cola-colored change of urine, and bilateral flank pain. Physical findings showed elevated blood pressure of 170/90 mm hg, pale conjunctiva, and bilateral costovertebral angle tenderness. Laboratory investigations showed moderate anemia (Hgb of 9.2 g/dl), positive ANA qualitative study, total CK 2154 IU/L (more than 15x elevated), elevated serum creatinine level (7 mg/dl), decreased C3 level (0.275 g/dl), normal C4 (0.275 g/dl), and elevated anti-dsDNA Ab (141.7 IU/L). Considering the evidence, she was diagnosed with SLE-polymyositis overlap syndrome, and she was started on methylprednisolone 500 mg IV for 3 days, followed by prednisolone 60 mg oral daily, cyclophosphamide 750 mg IV monthly, and amlodipine 10 mg oral daily. At the 4^{th} month of induction therapy with methylprednisolone and cyclophosphamide, her creatinine normalized and her proteinuria resolved. The patient is currently on maintenance therapy with azathioprine, chloroquine, and low-dose prednisolone, with complete clinical and laboratory remission.

In our patient, the occurrence of myositis, or polymyositis in our case, before SLE makes it unique and to the best of my knowledge, this pattern of clinical presentation has never been reported before.

Conclusion

Overlap of SLE and PM is a very rare clinical scenario and is an important cause of morbidity and mortality. The overlap syndromes have a poorer prognosis. Timely identification and treatment with immunosuppressive is the mainstay of the management of the disease**.**

Data Availability Statement

The data that support the findings of this case report are available from the corresponding author upon reasonable request

Ethical Approval

The author's institution does not require ethical approval for the publication of a single case report.

Consent for Publication

The patient provided written informed consent for the publication of details including, history, physical findings, laboratory reports, and imaging results.

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No funding was used in this case report.

Abbreviations

ANA anti-nuclear antibody

Ant-dsDNA anti-double-stranded deoxyribonucleic acid

CK creatine kinase

Hgb hemoglobin

IM inflammatory myositis

SLE systemic lupus erythematosus

PM Polymyositis

Authors

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