**Myositis in a Young Pregnant Woman: A Case Report**

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Abstract

This case report describes a presentation of a young healthy pregnant woman who presents early in the third trimester with generalized weakness, rash and shortness of breath. Evaluation and work-up demonstrate elevated muscle enzymes, multiple pathognomonic antibodies for systemic lupus and myopathic findings on MRI imaging. The discussion reviews myositis presentation and work up, a review of the literature regarding lupus versus non-lupus myositis findings and prognostic implications regarding the etiology of myositis associated with connective tissue disease.

Keywords: Myositis; Pregnant; Systemic lupus erythematosus; Prognosis; Overlap syndrome

Case Presentation

A 19-year-old African American G2P0101 female with past medical history of depression presented at 27 weeks gestation with progressive diffuse aching pain, generalized fatigue, and weakness for 2 months duration. Patient also reported a rash on her face and upper extremities for 1 year without pruritus or photosensitivity. She had mildly increased dyspnea on exertion which she indicated was worse than during her previous pregnancy. On physical exam, vital signs where significant for tachycardia but were otherwise unremarkable. The patient appeared drowsy with hyper pigmented well-demarcated plaques on nose, face, anterior arms and hyper pigmented erythematous macules on palms and toes. Power was 3+/5 throughout (proximal and distal) though limited by pain or effort. Her exam demonstrated no synovitis. Laboratory findings (Table 1) were significant for anemia, leukopenia, hypocomplementemia, hypoalbuminemia and proteinuria. Transaminases, Creatinine Kinase (CK), aldolase, C-Reactive Protein (CRP), and erthrocyte sedimentation rate were elevated. Urinalysis disclosed large protein, trace blood, with 3-5 RBCs. Serologic tests indicated positive assays for anti-nuclear, anti-Smith, anti-RNP, anti-Histone AB, and anti-DNA antibodies. VDRL was negative. An echocardiogram showed a small pericardial effusion. MRI of the left thigh showed diffuse patchy increased signal intensity within the quadriceps group, medial and lateral hamstrings, and adductor group. A muscle biopsy of the left vastus lateralis showed no evidence of active myositis. Skin biopsy of the rash showed dermal infiltrate with melanophages but without evidence of vasculopathy. Patient was started on hydroxychloroquine 400mg/day and prednisone 60mg/day with significant improvement in weakness, pain, and rash as well as down trending of CK and normalization of her hypocomplementemia. Pulmonary Function Tests (PFTs) were unable to be completed during the hospitalization and the patient was lost to follow up.

Discussion

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease involving multiple organ systems. The prevalence of SLE is significantly higher in women compared to men with the age of onset peaking around 20-30 years old during reproductive years [1,2]. SLE manifestations vary from arthralgias, malgias, fatigue, and rash to life-threatening end-organ damage involvement of the kidneys, nervous system and heart [3]. In addition, SLE may present with overlap syndromes including clinical features characteristic of SLE and rheumatoid arthritis or SLE and idiopathic inflammatory myositis [3].

Myalgia is a common presenting symptom of SLE, occurring in 70% of patients [4]. Inflammatory myositis is relatively uncommon in SLE, only occurring in 7-15% of patients. Inflammatory myositis typically presents with muscle weakness of the shoulder and pelvic girdle [5]. In a study evaluating muscle involvement in SLE, 8% of 228 patients with SLE had significant muscle disease. Among this group, a majority of patients had myositis at the time of diagnosis and though CK was largely normal, aldolase was frequently elevated.

The importance of differentiating between SLE with myopathy and SLE-myositis overlap...
syndrome lies in the prognostic implications associated with an overlap syndrome. In a study comparing SLE patients with SLE-myositis overlap syndrome patients, it was observed that the age of onset is typically younger in overlap patients (31 years old vs. 39 years old). In addition, patients with overlap all had pulmonary involvement on presentation with evidence of confirmed Interstitial Lung Disease (ILD) on imaging and pulmonary function tests earlier in the course of the disease [6]. Consequently, when SLE patients present with myositis, it is appropriate to evaluate them for pulmonary disease.

In SLE-myositis patients, pathology may show perivascular and perifascicular mononuclear cell infiltrates in 25% of patients. However, histology may be nonspecific in a majority of these patients [7]. One study compared SLE patients with myositis, SLE patients without myositis, and fibromyalgia patients in an attempt to evaluate data that might differentiate between the diseases. They observed that the presence of lymphocytic vasculitis in quadriceps muscle biopsies was found exclusively in SLE patients [8]. In addition, they observed that there was a statistically significant difference of anti-RNP and CK elevation between SLE patients with myositis and SLE patients without myositis.

The differentiation between myalgias and myositis in SLE patients is important for prognosis and management. Making the proper diagnosis of SLE/Myositis overlap syndrome allows physicians to remain vigilant about pulmonary involvement in these patients.

Differential diagnostic evaluation of these SLE patients by obtaining CK, aldolase, and anti-RNP antibodies may suggest the diagnosis of myositis. It is also important to recall that elevated transaminases may reflect myositis, rather than liver disease in these patients. In addition, MRI and muscle biopsy may help confirm myositis in patients with SLE. Interestingly, in our case, despite imaging and laboratory evidence of myositis, biopsy of the involved muscle was non-diagnostic; this likely occurs due to the patchiness of myositis and sampling error.

Acknowledgements

We would like to thank Dr. Stanley Ballou for his helpful suggestions.

References