Inflammatory Myositis—Secondary to SLE

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Abstract

Inflammatory myositis involving the proximal muscles has been reported to occur in 5% to 11% of SLE patients and may develop at any time during the course of the disease. It can be secondary to internal malignancies also. We report one such patient who presented with generalised muscle weakness for 7 months. Erythematous hyperpigmented scaly patches were present over the scalp, face, trunk, upper limbs. We discuss the inflammatory myopathies secondary to SLE and internal malignancies. Most cases respond to low-dose corticosteroid treatment.

Introduction

In SLE, generalized myalgia and muscle tenderness are common, especially during disease exacerbations. As per the Kelley’s Textbook of Rheumatology, 8th ed; Inflammatory myositis involving the proximal muscles has been reported to occur in 5% to 11% of patients and may develop at any time during the course of the disease. There have been many associations between the inflammatory myopathies and the presence of malignancy, but the etiology of the association is controversial. Here we report a lady who presented with inflammatory myositis, secondary to SLE, and discuss the possible causes of such a presentation.

Table 1: Suggested classification for inflammatory myopathies

Descriptions

Pure Dermatomyositis (DM)
Pure Polymyositis (PM)
Cancer-associated myositis (CAM): clinical paraneoplastic features without an overlap autoantibody or anti-Mi-2
Overlap myositis (OM): myositis with at least one clinical overlap feature or an overlap autoantibody

Bohan and Peter Definition of Myositis

1. Symmetric proximal muscle weakness
2. Typical skin rash of DM, including heliotrope rash, Gottron’s sign, and Gottron’s papules
3. Elevation of serum skeletal muscle enzymes
4. Muscle biopsy abnormalities of degeneration, regeneration, necrosis, phagocytosis, and interstitial mononuclear infiltrate
5. Electromyographic triad of short, small, polyphasic motor unit potentials; fibrillations, positive sharp waves, and insertional irritability; and bizarre, high-frequency repetitive discharges

Probable myositis: 3 criteria (without the rash) for PM; 2 criteria (plus the rash) for DM
Definite myositis: 4 criteria (without the rash) for PM; 3 or 4 criteria (plus the rash) for DM

Case Report

A 44 year old housewife, mother of two children, presented with c/o progressively increasing symmetrical weakness of all four limbs and neck muscles for 7 months, for which she consulted primary care physicians and received symptomatic treatment without a proper diagnosis (Details not available). There was no history of diabetes, hypertension and cardiovascular disease. She had history of hysterectomy eight years back for fibroid uterus. On examination erythematous hyperpigmented scaly patches were present over the scalp, face, trunk, upper limbs (Figures 1-3). She had multiple small joint arthritis (Figures 4-5). She also had bilateral hallus valgus which was an incidental finding (Figure 6). There were no cutaneous markers suggestive of Dermatomyositis. CNS examination showed generalised wasting, hypotonia in all four limbs, symmetrical weakness (Proximal>Distal) of all four limbs and neck muscles. All deep tendon reflexes were sluggish. There were no signs of involvement of sensory system and cranial nerves. Other systems were within normal limits.

Routine full blood count and biochemical analysis including thyroid function tests were normal. Chest X-ray, Electrocardiogram, USG Abdomen and CECT Abdomen were also normal. ESR was 20 mm/hr. She was negative for HIV test. We investigated her for muscle disorders secondary to autoimmune and neoplastic causes: CPK-10,730. LDH - 298. pANCA and cANCA- negative. CEA and CA-125 were Negative, USG B/L Breast was Normal study, Stool occult blood –negative. Rheumatoid factor was negative. Colonoscopy and Upper GI Endoscopy were normal.

Muscle Biopsy (Left vastus lateralis) showed perifascicular interstitial inflammation with perifascicular atrophy, ATPase enzyme histochemistry showed atrophic Type two fibres: Inflammatory myopathy-consistent with polymyositis and possible vasculitis. There were no histopathological features of dermatomyositis. Decreased fibre density with a few regenerating fibres on myelin stain and Mild to moderate chronic axonopathy was there in Left Sural nerve biopsy which can be secondary to SLE. NCS was suggestive of sensory motor axonal neuropathy. EMG was suggestive of neuropathic pattern of weakness. ANA was positive. ANA profile was (Anti U1 RNP, Anti Sm, Anti Ro, Anti SSA, Anti SSB, Anti Jo-1, Anti Sc-I-70 autoantibodies) negative, except that Anti ds DNA was positive. (The pattern is attached).

She was diagnosed to have inflammatory polymyositis secondary to SLE: was managed conservatively with dexamethasone 8 mg iv, as she had gastritis at admission which later changed to prednisolone at a dose of 0.5 mg/kg/day and azathioprine 50 mg/day¹ and physiotherapy and is on follow up for internal malignancy. Her weakness was improving with treatment.

Discussion

Myositis

Polymyositis (PM), Dermatomyositis (DM), and inclusion body myositis (IBM) are the classic idiopathic inflammatory myopathies (IIMs) (Table 1), yet the same clinical picture and
investigational findings may be found in patients with SLE, scleroderma, MCTD, and Sjögren’s syndrome. Inflammatory myositis involving the proximal muscles has been reported to occur in 5% to 11% of SLE patients and may develop at any time during the course of the disease.

**Inflammatory Myopathies**

The inflammatory myopathies in adult populations encompass a group of illnesses characterized by an idiopathic immune-mediated attack on skeletal muscle that results in muscle weakness. There have been many associations between the inflammatory myopathies and the presence of malignancy, but the etiology of the association is controversial. Dermatomyositis has classically been associated with occult malignancies, whereas the associations between polymyositis and inclusion body myositis are less clear. A further issue is whether the inflammatory myopathy predates the malignancy and can be considered a primary rheumatic disease with known risks of developing malignancy, or whether it simply represents a manifestation of a paraneoplastic process.

On average, the prevalence of malignancy in association with the inflammatory myopathies has been approximately 25%. The frequency of malignancy has ranged, however, from 6% to 60% in patients with dermatomyositis and from 0 to 28% in patients with polymyositis. Other estimates have placed the incidence of cancer in patients with polymyositis at five to seven times that of the general population. In polymyositis, the relative risk for developing internal malignancies seems to be lower than that for dermatomyositis, but it is consistently increased over that expected in the general population. Studies have found a 14% to 30% prevalence of cancer among patients with polymyositis.

Studies have suggested that imaging of the chest, abdomen, and pelvis may increase the potential for discovery of underlying malignancy. Other studies have suggested the use of serum tumor markers (CA125 and CA19-9) to augment detection of patients with dermatomyositis or polymyositis at highest risk for associated malignancy. Malignancies associated with inflammatory myopathies have been known to develop many years after the diagnosis of muscle disease, so continued vigilance and repeated screening for malignancy are warranted. Although the pathogenesis is unknown, the types of malignancy associated with the inflammatory myopathies have been varied, including adenocarcinomas of the breast, ovaries, and stomach.

**SLE with Myositis**

Generalized myalgia and muscle tenderness are common, especially
During disease exacerbations. Inflammatory myositis involving the proximal muscles has been reported to occur in 5% to 11% of patients and may develop at any time during the course of the disease. The differential diagnosis of proximal muscle weakness in SLE includes a drug-related myopathy secondary to corticosteroid, antimalarial, or statin medications use. Concurrent hypothyroidism also can cause an increase in creatine phosphokinase and proximal myopathy. Muscle biopsy, electromyographic studies, and elevation of the serum creatine phosphokinase or aldolase levels help to differentiate between inflammatory and drug-related myopathy. The histologic features of myositis in SLE may be less striking than in idiopathic polymyositis. Histologic features include muscle atrophy, microtubular inclusions, and a mononuclear cell infiltrate. Fiber necrosis is an uncommon finding, but immunoglobulin deposition is almost always present despite the rarity of concurrent inflammation. A low serum creatine phosphokinase value can be found in patients with connective tissue disease including SLE; a normal creatine phosphokinase value in the presence of symptoms and signs of myositis should not dissuade the physician from a diagnosis of myopathy. The skin lesions of dermatomyositis also can appear in patients with SLE. Chest pain or discomfort secondary to costochondritis has been reported in SLE, and other conditions, such as angina pectoris, pericarditis, and esophageal spasm, must be ruled out first.

**Definition of clinical Paraneoplastic Features**

Cancer within 3 yr of myositis diagnosis, plus absence of multiple clinical overlap features; plus, if cancer was cured, myositis was cured as well

**References**