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CASE REPORT

Polymyositis

[1]Yuvraj S Cheema, [2]Parveen Gupta, [3]Sahil Attri, [4]Vipin Jamdagni, [5]Deven Mittal

ABSTRACT

Polymyositis is an uncommon inflammatory disease that causes muscle weakness affecting both sides of the body, typically the proximal muscles. Most commonly, it affects adults in their 30s, 40s, or 50s. Signs and symptoms usually develop gradually, over weeks or months. Raised creatinine phosphokinase (CPK) levels are commonly seen during the acute phase. The electromyogram characteristically shows a myopathic pattern. The affected muscles histopathologically demonstrate endomysial inflammation, lymphocytic infiltration, zonalmyofibrillar loss, and perifascicular atrophy. We report a male patient with a classical presentation of polymyositis.

Keywords: Endomysial inflammation, Inflammatory myopathy, Lymphocytic infiltration, Perifascicular atrophy, Polymyositis.

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INTRODUCTION

Polymyositis is an inflammatory myopathy associated with progressive symmetric proximal muscle weakness. The overall disease incidence is 1:100000 with a female/male ratio of 2:1.[1-3] The affected patients experience increasing difficulty in performing everyday tasks like climbing stairs, getting up from squatting position, and raising arms above the head. Extramuscular manifestations can also be present ranging from systemic symptoms (fever, malaise, weight loss, arthralgia, and Raynaud's phenomena) to joint contractures, dysphagia, gastrointestinal symptoms, cardiac symptoms, pulmonary dysfunction, and subcutaneous calcifications.[1] Electromyogram and creatinine phosphokinase (CPK) levels aid in diagnosis but muscle biopsy is the most sensitive and specific test for establishing the diagnosis of inflammatory myopathy.[1] We present a male patient with a characteristic presentation of polymyositis.

[1]Postgraduate Student (3rd Year), [2]Professor, [3,5]Postgraduate Student (1st Year), [4]Postgraduate Student (2nd Year)

[1]Department of Internal Medicine, Maharishi Markandeshwar Institute of Medical Sciences and Research, Ambala, Haryana India

[2-5]Department of Medicine, Maharishi Markandeshwar Institute of Medical Sciences and Research, Ambala, Haryana, India

Corresponding Author: Yuvraj S Cheema, Postgraduate Student (3rd Year), Department of Internal Medicine, Maharishi Markandeshwar Institute of Medical Sciences and Research Ambala, Haryana, India, Phone: +919888889218, e-mail: yuvrajsinghcheema@hotmail.com

CASE REPORT

A 42-year-old male patient presented to us with an 8-month history of progressive weakness in both the lower limbs, followed by weakness in both the upper limbs for about 6 months. The patient reported difficulty in getting up from squatting position, climbing stairs, and doing routine daily activities. The weakness had progressively worsened, followed a few months later by upper limb weakness. There was no history of any sensory loss. There was no history of trauma. Family history was noncontributory. On examination, the patient had demonstrable proximal limb weakness. Power in the upper limbs was grade II proximally and grade III to IV distally. Muscle power in the lower limbs was grade II proximally and grade III + IV distally. Neck and trunk muscles had mild weakness. Deep tendon reflexes were depressed and plantars were flexors. There was no sensory deficit. On investigating the patient, total CPK level was significantly increased to 332 IU/L; ESR was 100 mm; hemoglobin 13.08; total leukocyte count 8000 mm^3; serum albumin 3.0 gm/dL; protein 6.5 gm/dL; urea 17.0 mg/dL; creatinine 0.45 mg/dL; sodium 138 meq/L; and potassium 4.3 meq/L. Electrocardiogram was normal. Thigh muscle biopsy demonstrated variations in the muscle fiber size within the bundles with moderate degree of lymphocytic and histiocytic cells in the endomysium. Small area of inflammatory infiltrate concentrating around the muscle fibers with necrosis and phagocytosis is also present. Fat infiltration also present. IMPRESSION – POLYMYOSITIS.

The patient was discharged on oral prednisolone 1 mg/kg and physiotherapy exercises. Further progression or improvement of disease would be assessed in the coming weeks. Treatment would be adjusted accordingly.

DISCUSSION

Polymyositis is an inflammatory myopathy, resulting in progressive proximal limb muscle weakness.[1,2,4,5] This causes progressive difficulty in performing everyday tasks. Neck and pharyngeal muscle affection can occur, resulting in head drop and dysphagia respectively. Ocular

muscles are spared, deep tendon jerks are preserved, and sensations are normal.[1,4] Our patient presented with classical progressive proximal muscle weakness, with mild affection of neck and trunk muscles. Sensory sensations were intact. There are reports from India of case series of patients with inflammatory myopathies, mainly polymyosits[6]; also a patient with polymyositis presenting predominantly with respiratory failure.[7]

Other systemic manifestations that are associated with polymyositis include fever, malaise, weight loss, arthralgia, Raynaud's phenomena, atrioventricular conduction defects, tachyarrhythmias, congestive heart failure, interstitial lung disease and gastrointestinal symptoms, such as dysphagia and dysphonia.[1,2] Our patient did not have any cardiac, respiratory, or gastrointestinal involvement.

Polymyositis may coexist with systemic connective tissue disorders, such as scleroderma, systemic lupus erythematosus, mixed connective disease, or underlying malignancy.[1,2,4] In some patients antibodies to RNA synthetases (anti-Jo 1) may be documented; their presence frequently correlates with the presence of interstitial lung disease.[1] Our patient did not have any associated systemic connective tissue affection. Advanced age, long-standing weakness, and associated malignancy, or heart and lung involvement are poor prognostic indicators.[4] In patients with polymyositis, total CPK is frequently raised during the acute phase.[4] Our patient had a total CPK value of 1777 IU/L. Electromyogram characteristically demonstrates a myopathic pattern.[1,4] Histopathological examination of affected muscle reveals endomysial inflammation with lymphocytic infiltration, zonalmyofibrillar loss, and perifasciculartrophy (Fig. 1).[4,5,8] A similar histopathological appearance was seen in muscle biopsy of our patient. Early initiation of therapy is essential in patients with polymyositis, since patients often respond well to immunosuppressive therapy.[1,2,4,5,9,10] Treatment options include corticosteroids, azathioprine, methotrexate, cyclophosphamide, rituximab, and IV immunoglobulins. The following sequential empirical approach to treatment of polymyositis is suggested: Step 1 – high-dose prednisolone; Step 2 – azathioprine, mycophenolate, or methotrexate for steroid sparing effect; Step 3 – IV Ig; Step 4 – a trial with rituximab, cyclosporine, cyclophosphamide, or tacrolimus.[1]

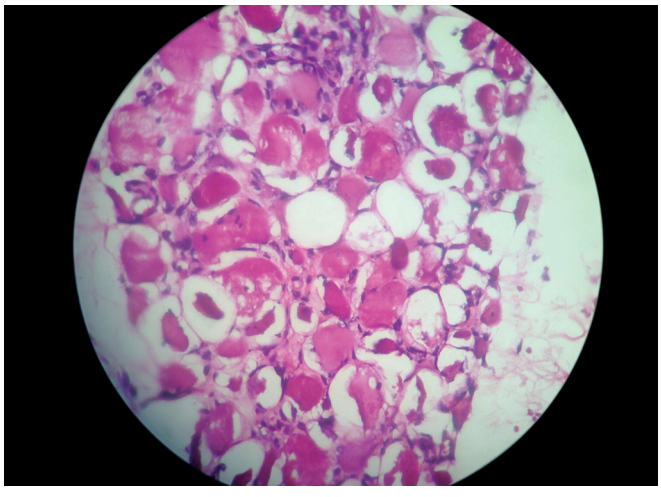

Fig. 1: Endomysial inflammation with lymphocytic infiltration. (A) Muscle biopsy high power; and (B) muscle biopsy low power

CONCLUSION

Polymyositis is an inflammatory myopathy causing progressive proximal limb weakness without any sensory affection. Other systemic manifestations in the form of arthritis, Raynaud's phenomena, cardiac rhythm disturbances, cardiac failure, interstitial lung disease, or gastrointestinal affection may be present. At times, polymyositis may be associated with other connective tissue disorders, such as systemic lupus erythematosus, Sjogren's syndrome, or mixed connective disorder. Definitive diagnosis is made by muscle biopsy which reveals endomysial inflammation with lymphocytic infiltration and zonalmyofibrillar loss. Treatment is by administration of immunosuppressive therapy. Patients respond well to therapy, but some amount of residual weakness is not uncommon.

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