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Polymyositis: A Case Report

KEYWORDS: Polymyositis, inflammatory myopathy, endomysial inflammation, lymphocytic infiltration, perifascicular atrophy, proximal weakness, immunosuppressive therapy.

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ABSTRACT *Polymyositis is an idiopathic inflammatory myopathy characterised by progressive symmetric proximal muscle weakness resulting in increasing difficulty with everyday tasks. Raised total CPK levels are commonly seen during the acute phase. The EMG characteristically shows a myopathic pattern. Affected muscles histopathologically demonstrate endomysial inflammation, lymphocytic infiltration zonalmyofibrillar loss and perifascicularatrophy.Patients respond to immunosuppressive therapy, although some degree oflong term muscle weakness is not uncommon. We report a female with aclassicalpresentation ofpolymyositis.*

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.[1,2,3].Affected patients experience increasing difficulty in performing everyday tasks like climbing stairs, getting up from squatting position, raising arms above the head. Neck and pharyngeal muscle affection may also occur, resulting in head drop and dysphagia respectively[1,2].We present a female patient withacharacteristic presentation of polymyositis.

CASE REPORT

A 60 year old female patient presented to us with a 5 month history of progressive weakness in both the lower limbs, followed by weakness in both the upper limbs for about 2 months. The patient reported difficulty in getting up fromsquatting position, climbing stairs, so also sitting up or turning sideways in the bed. The weakness had progressively worsened. This was followed a few months later by upper limb weakness. The patient had difficulty wearing clothes, combing her hair. There was no history of any sensory loss. There was no history of dysphagia, double vision or skin rash. Family history was non-contributory. On examination, the patient had demonstrable proximal limb weakness. Power in the upper limbs was Grade III proximally and Grade III-IV distally. Muscle power in the upper limbs was Grade II proximally and Grade III distally. Neck and trunk muscles were also weak. 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 7590 IU/L; ESR was 20 mm. EMG revealed a characteristic myopathic pattern. Thigh muscle biopsydemonstrated multifocal endomysial inflammation, lymphocytic inflammatory infiltration and focal individual fibre degeneration (Image 1). The histopathological appearancewas consistent with polymyositis. ECG, 2Dechocardiography and HRCT thorax werenormal. The patient was initially given pulse methyl-prednisolone therapy for 3 days and then put on oral prednisolone andazathioprine. A few weeks into treatment, the patient reported slight improvementin mucle power. She was discharged on immunosuppressive therapy.

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. Sensations were intact.There are reports from India of case series of patients with inflammatory myopathies, mainly polymyositis[6]; so also a report of a patient with polymyositis presenting predominantly with respiratory failure[7].

Other systemic manifestations that associated withpolymyositis includefever, malaise, weight loss, arthralgia, Raynaud's phenomena, atrio-ventricularconduction defects, tachyarrhythmias, congestive heart failure, interstitial lungdisease and gastrointestinal symptoms such as dysphagia, dysphonia [1,2] . Our patientdid not have any cardiac, respiratory or gastrointestinal affection.

Polymyositis may co-exist 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 prognos-

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tic indicators[4] .

In patients with polymyositis, total CPK is frequently raised during the acute phase[4].Our patient had a total CPK value of 7590 IU/L. EMG characteristically demonstrates a myopathic pattern[1,4] , as seen in our patient. Histopathological examination of affected muscle reveals endomysial inflammation with lymphocytic infiltration, zonalmyofibrillar loss and peri-fascicularatrophy[4,5,8]. A similarhistopathological appearance was seen in muscle biopsyof 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. Our patient was administered corticosteroids and reported improvement in muscle weakness.

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 lupuserythematosus, 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 respondwell to therapy, but someamount of residual weakness is not uncommon.

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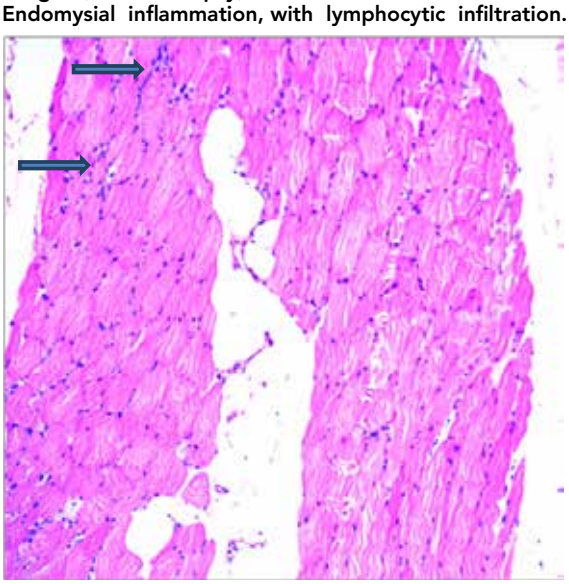
Image 1(Muscle Biopsy):
Endomysial inflammation, with lymphocytic infiltration.



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