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A Case of Idiopathic Inflammatory Polymyositis – a rare phenomenon

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Abstract
Idiopathic inflammatory polymyositis is a disorder characterised by progressive muscle weakness, extra muscular manifestations , elevated serum levels of muscle enzymes , Electromyographic (EMG) abnormalities, inflammatory infiltrates in muscle biopsy . Extra muscular involvement such as pulmonary , cardiac and gastrointestinal involvement is correlated with increased mortality and morbidity in patients. Here we report a case of idiopathic inflammatory polymyositis.
Keywords: Polymyositis, Anti JO-1 antibodies, idiopathic inflammatory polymyositis , IIM .

Introduction

Polymyositis (PM) is a one of rare entity of IIM, presents with muscular and extra muscular organ involvement similar to dermatomyositis (DM), without a rash . Unlike DM which manifests in childhood, PM usually manifests in adults over 20 years and is more common in female. Proximal limb muscle weakness, myalgia and are the most common complaints experienced by the patient in polymyositis . Serum creatine kinase (CK) and myositis specific antibodies (MSA) help in differentiating clinical phenotype of IIM and to confirm the diagnosis. This disease may progress to other organ systems, such as the lung and the heart, with significant morbidity. Early diagnosis and timely medical intervention remains the gold standard in treating polymyositis. Here we report a rare case of idiopathic inflammatory polymyositis.

Case Description:

A 31 year old male patient with no known comorbidities presented to our department with muscle pain. He was chronic alcoholic for more than 10 years, consumed 180ml/binge on alternate days, (last binge 20 days back). He had complaints of lower back pain on and off since 2 months, , diffuse abdominal pain since 3 days , weakness and myalgia since 2 weeks . He experienced inability to perform day to day activities, inability to walk since two days, inability to get up from sitting position. He had history of inability to roll on the bed from one side to the other, inability to lift hands above the level of shoulder. He was unable to comb his hair . On physical examination , there was bilateral upper limb and lower limb proximal muscle weakness with normal tone . Other systems were clinically normal .

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Lab investigations revealed normal blood counts with Hb12.7 gm/dl, platelet 3 lakhs/cu.mm, total leucocyte count 10250 cells/cu.mm. LDH was highly elevated 985 IU/L (100-300 IU/L). Total and direct bilirubin was 0.63 and 0.22 respectively. Elevated SGOT level was noted and it was 663 U/L. SGPT - 246, ALP -103, albumin and electrolytes were normal. Serum calcium was 8.7 mg/dl.
Renal function tests were normal. Total CPK was highly elevated to the level of 32000 IU/L (20-200IU/L).

ANA test was positive with speckled pattern. Anti JO- 1 antibodies were highly positive (++) in myositis profile with ESR 120mm/hr (<30mm/hr), CRP 349(<10) and ASO titre 249.1(<200) . Urine myoglobin was negative. Electromyographic findings showed spontaneous activity with positive sharp waves and fibrillations noted in right deltoid, biceps and quadriceps. qmvp-multiphasic , normal to medium amplitude and normal duration. Early and complete recruitment showed myopathic pattern with positive sharps and fibrillations, suggestive of inflammatory myositis. MRI stir image obtained from bilateral thigh showed signs of increased uptake.

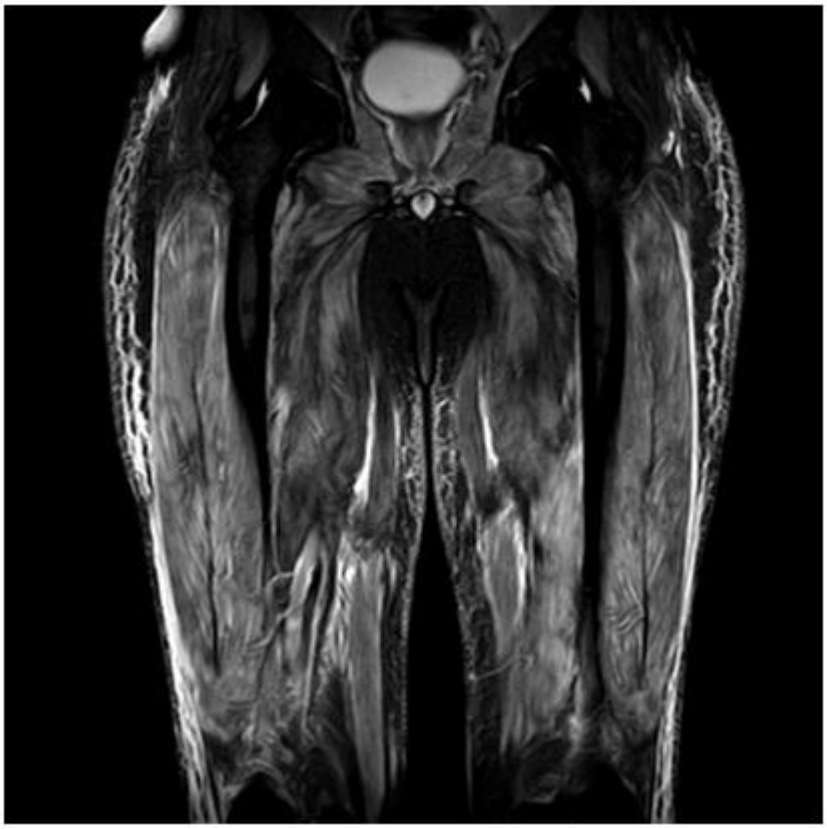

Figure 1 . MRI STIRR IMAGE OF BILATERAL THIGH OF THE PATIENT SHOWING INCREASED UPTAKE .

Diagnosis of Polymyositis was confirmed and patient was started on pulse therapy with oral steroids. He showed improvement within few days and pain reduced . He was able to walk and carry out normal day to day activities . Patient is now on regular follow up with no specific complaints.

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Discussion

Polymyositis is increasingly being recognised as a bucket term for idiopathic inflammatory myositis. Patient typically presents in a manner similar to dermatomyositis with subacute proximal symmetric muscle weakness; however, polymyositis patients are without characteristic rashes of dermatomyositis .[9] Progressive muscle weakness, elevated serum levels of muscle enzymes, Electromyographic (EMG) abnormalities, inflammatory infiltrates in muscle biopsy and extra muscular manifestations are the hallmarks of polymyositis. Serum CK levels may be elevated up to 50 times and our patient's CK was around 32000 IU/L (<200IU/L).

The incidence of PM is 3.8 and the prevalence is 9.7 per 100,000 people with female preponderance. PM increases with advancing age and reaches a peak at age 50–59 years .[11, 12] EMG is useful in demonstrating the myopathies. Around 90% of the patients will have abnormal EMG .[13] Our patient showed spontaneous activity with positive sharp waves and fibrillations in right deltoid, biceps and quadriceps. Anti-Jo-1 antibodies occur in syndromes that includes muscle weakness and pain. Our subject was highly positive for anti-Jo 1. Confirmation of IIP is done by all these markers.

Patient was initially evaluated for myositis. Since he had history of alcoholism, differential diagnosis was initially alcohol induced rhabdomyolysis, but urine myoglobin was negative. Then ANA was done and was positive with speckled pattern, with jo1++. Then patient's diagnosis was made as acute bilateral symmetrical proximal muscle weakness involving shoulder and hip with no sensory, cranial nerve, bladder and bowel involvement. Further electromyography also confirmed features suggestive of polymyositis. Corticosteroid (high-dose) is the first line of treatment for adult onset of dermatomyositis and polymyositis .[14] So patient was started on pulse therapy followed up with oral steroids and showed great improvement within 4 days , patient was able to walk and pain reduced significantly . Patient is now in regular follow up and has no complaints.

Conclusion

Polymyositis has highest mortality rate of around 28%. Malignancies, arthropathy, cardiomyopathy and interstitial lung disease are the common reasons behind the critical illness and mortality.. Intensive immunosuppressive treatment may result in high risk of infection so care should be taken in aiding the suitable treatment in appropriate time. Our patient was diagnosed and treated appropriately and his condition is improving. So Early diagnosis and therapy remains the gold standard for optimal prognosis.

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