

Case Report

A rare case of inflammatory myopathy

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ABSTRACT

Idiopathic inflammatory myopathies (IIMs) happened to be the group of heterogeneous, systemic rheumatic diseases including adult polymyositis (PM), adult dermatomyositis (DM), myositis accompanied with another connective disease or cancer. A 52 years old male patient with known history of type 2 diabetes mellitus presented with complaints of muscle pain and swelling over left arm. These cases was successfully treated by using Corticosteroids. This cases study described the clinical presentation and features of inflammatory myositis. Although this is a rare case, its clinical features and treatment procedure helps in management of similar cases.

Keywords: Corticosteroids, Inflammatory polymyositis, Type 2 diabetes

INTRODUCTION

The idiopathic inflammatory myopathies (IIM) are often described as group of heterogenic disorders presented as muscle inflammation and progressive muscle weakness. This group of disorder comprises of five diseases including dermatomyositis (DM), nonspecific or overlap myositis (NSM), necrotizing autoimmune myopathy (NAM), and sporadic inclusion body myositis (IBM).¹ The literature available shows that, the incidence of PM, DM and IBM is approximately 1 in 100,000.^{2,3}

The studies pertaining to this type of myositis show that, the viral, bacterial infections and some drugs are known to trigger this disease in the presence of genetic factors and autoimmune mechanisms.^{4,5} Measurement of serum enzyme levels, electromyography and muscle biopsy are the critical tests for establishing and confirming the diagnosis.⁶ Creatinine kinase is most sensitive enzyme, levels of aspartate aminotransferase, alanine aminotransferase and lactate dehydrogenase are usually elevated along with aldolase, myoglobin and creatinine.⁷

Short duration, low amplitude polyphasic units on voluntary activation and increased spontaneous activity

with fibrillations, complex repetitive discharges and positive sharp waves are usually found by using needle electromyography.⁶ definitive diagnosis can be established by using muscle biopsy. This case report may help in establishing the clinical signs and symptoms, laboratory features, electromyography and muscle biopsies and treatment of patients with inflammatory polymyopathies.

CASE REPORT

A 52 Years old male known case of type 2 diabetes mellitus since 12 years on insulin presented to our hospital with complaints of muscle pain and swelling over left arm for over 1month and over his both thighs since last 1 week. He also complaints of difficulty in getting up from squatting/lying down position since 1 week, no complaints of weakness of upper limbs. General examination revealed swollen and tender biceps, deltoids, quadriceps and hamstrings bilaterally, vitals were stable, afebrile, no tachypnae, tachycardia. Systemic examination cardiovascular, respiratory and abdominal examination revealed no significant abnormality, Motor system examination revealed reduced power in the proximal hip muscles, adequate higher mental functions,

cranial nerve examination was normal, sensory system was intact. On evaluation with blood investigations revealed normal complete haemogram, renal parameters, serum electrolytes, liver function tests, urine routine was normal, CRP-Negative, ESR-60mm/hr, Total Creatinine kinase was elevated -15850 U/L and CPK-MB- 200U/L.

In view of elevated creatinine kinase value history was taken regarding any medications that causes myopathy but patient was not on statins or other drugs causing myopathy, no history of intake of antipsychotics or clinical features suggestive of malignant neuroleptic syndrome was ruled out, no prior history of similar complaints, no significant family history. He was provisionally diagnosed with inflammatory myopathy, anti-U1RNP, ANA was sent and were negative, autoimmune etiology for myositis was ruled out. He empirically initiated on injectable intravenous methylprednisolone pulse therapy, adequate analgesia was also given. 2D Echocardiography was done and no revealed no significant abnormality. Muscle biopsy specimen was obtained from left vastus lateralis and was sent for histopathological examination.

Muscle biopsy showed skeletal muscle bundles showing mild endomysial inflammation and focal myofibre inflammation. Foci of fibrosis was noted, an impression of features correlating with inflammatory myositis was made. On day 3 of intravenous methylprednisolone pulse therapy creatinine kinase decreased to, patient showed definitive evidence of clinical improvement. 5 days of pulse steroid was continued and later he was switched over to oral prednisolone 1mg/kg. He was discharged from hospital with oral steroids, oral antidiabetic drugs and advised for follow up on OP basis. Patient was reviewed in OPD after 2 weeks and was found to have significant clinical improvement and was advised to continue oral steroids.

DISCUSSION

The defined diagnostic criteria for dermatomyositis and polymyositis is conflicting as evident by the primary studies in this regard.^{8,9} Patient in this study presented with muscle pain, myalgia and muscle tenderness which happen to be common signs and symptoms like other studies and more often in dermatomyositis than polymyositis.^{10,11} Literature shows that, the disease subacute in onset and muscle weakness develops within weeks to months in a symmetrical and proximal distribution in the arms and legs. Hip flexors and/or shoulder abductors are commonly affected which may extend up to neck flexors and distal muscles. Myopathies can also involve other systems, rash and periorbital edema.¹²

The diagnosis is mainly based on the clinical, laboratory and pathological examinations. The laboratory findings including elevated serum creatine kinase (CK), positive antinuclear (ANA) bodies and antibodies to the

extractable nuclear antigens (ENA) e.g., myositis associated antibodies (MAAs) including anti SS-A, Anti SS-B, Anti ribonucleoprotein (Anti - RNP), anti- PM - Scl, anti scl 70 and anti centromere antibodies may indicate a concomitant connective tissue disorder. Specific syndromes can lead to the myositis specific antigens.^{13,14}

A muscle biopsy may be necessary to confirm the diagnosis. In 10 - 20% the muscle biopsies may be inconclusive which may be sampling error. The histopathology may show the complement mediated microangiopathy with perifascicular atrophy (usually in later stages and more so in children than adults) and inflammatory cell infiltrates composed of macrophages, B - cells, plasmacytoid dendritic and CD4 positive T - helper cells, located predominantly at perivascular sites in the perimysium.^{15,16}

Use of corticosteroids is the generally accepted treatment for the inflammatory polymyopathies followed by tapering over a period of 1 - 1.5 year. Alternative immunosuppressive treatments including azathioprine or methotrexate are recommended in case of relapse. In severe and refractory cases, intravenous immunoglobulin (IVIG), intravenous methylprednisolone, rituximab or entercept can be considered.¹⁷⁻²⁰

CONCLUSION

Prompt diagnosis and early treatment of inflammatory myopathies is crucial in early recovery of patients with inflammatory myopathies. Delay in diagnosis can lead to suffering of the patient.

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