Volume : 3 | Issue : 1 | January 2014 • ISSN No 2277 - 8179

Research Paper

Medical Science

KEYWORDS : Inflammatory myopathy, plasmapheresis

Role of Plasmapheresis in a Treatment of Idiopathic Inflammatory Myopathy

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ABSTRACT

Idiopathic inflammatory syndrome, a rare autoimmune disorder of unknown etiology characterised by muscle inflammation. We report an elderly lady with acute onset , progressive , proximal muscle weakness which developed over a period of one month duration. Clinical features and laboratory results led us to the diagnosis of idiopathic inflammatory myopathy. Her muscle weakness did not improve with steroids. She, then received plasmapheresis , in addition to oral azathioprine which was not found to be beneficial. She improved slowly but marginally, after about one month of combination therapy of oral azathioprine and methotrexate , which was continued.

Introduction –

Idiopathic inflammatory myopathies(IIM) are a group of rare, systemic diseases that result in proximal muscle weakness, muscle enzyme elevations and extramuscular manifestations such as fever, weight loss and rash. The most common clinical forms are polymyositis, dermatomyositis and inclusion body myositis ; however, other clinically useful phenotypes with different risk factors and prognoses are also defined. Treatment remains challenging because of varied clinical presentation , responses to the treatment and prognoses. We are presenting our experience of plasmapheresis in a patient with idiopathic inflammatory myopathy.

Case Report--

A 65 year old lady was admitted to our hospital complaining of about 1-month duration of myalgia of the shoulder and limb muscles and progressive difficulty in getting up from sitting or supine position ,combing her hair, climbing stairs, dressing and undressing. She had no fever, arthralgia , arthritis, cough, dyspnoea, rash, difficulty with swallowing, paresthesias, or any other symptoms. There was no history of any medical illness or consumption of any medication.. The patient did not smoke, drink alcohol or use illicit drugs and her family history was negative for neuromuscular diseases.

Neurological examination revealed -4/5 muscle strength in the upper and lower extremities more in proximal muscle groups. Her neck holding was poor. The tendon reflexes, muscular tone and sensation were normal throughout and plantars were flexor. There was no evidence of any skin lesions.

Laboratory investigations showed Hb- 11gm%, fasting blood sugar level 86mg/dl, Sr Na -138mmol/L, Sr K-4.2mmol/L , Sr Calcium – 8.8 mg/dl, Sr albumin 4.2gm/dl, Sr creatinine – 0.9mg/dl and TSH- 2.3 mU/L . She was HIV negative. Urine examination was normal. Her serum creatinine kinase level was 8990 IU/ L.

Electrocardiogram , 2D Echocardiogram, ultrasonography abdomen and pelvis, computed tomography chest studies were done, which revealed no abnormality.

Her ANA titre was negative. Antibodies tested against antigens Sm, SS-A, SS-b, Ro-52, Scle 70, PM –scle, JO-1, CENP-B, dsDNA were negative.

The Electromyography of upper and lower limbs showed short – duration, low-amplitude polyphasic potentials and positive sharp waves suggesting myopathic changes in all the examined proximal muscles. Nerve conduction study was normal.

Left quadriceps muscle biopsy (which was not tested with EMG) section study showed muscle bundles/fascicles consisting of myofibers. There was moderate variation in size and shape of myofibers. Some fibers were pale and vacuolated. Focal necrosis of myocytes was noted showing moth eaten appearance of cytoplasm. Regenerating myocytes having basophilic cytoplasm and vesicular nuclei with prominent nucleoli were seen. A sparse lymphocytic infiltrate was seen around the myocytes. Histopathological findings were consistent with inflammatory myopathy.

The patient was started on prednisone 1mg /kg/d . There was no improvement in muscle pain or power even after 15 days of treatment . Then she underwent 7 cycles of plasmapheresis with 1.5 lit of plasma exchange per cycle and simultaneously azathioprine was started in a dose of 2 mg/kg/d along with calcium and vitamin D supplementation. There was subjective improvement in myalgia and muscle tenderness decreased. Serum creatinine kinase level decreased. Improvement in muscle strength was not significant. So she was started on oral methotrexate, weekly. One month after this combination therapy, patient became ambulatory with support with improved neck holding .

Discussion –

Although the IIM are rare, they are the most commonly acquired chronic muscle diseases in adults, with an estimated prevalence of 10–20 per 100 000. Their aetiology remains unknown but these diseases probably result from chronic inflammation induced by a combination of the necessary and sufficient genetic and environmental risk factors.(1) The diagnosis of these disorders is based on the combination of clinical examination, electromyographic data, serum muscle enzyme levels, various autoantibodies, and the muscle biopsy findings. The muscle biopsy offers the most definitive diagnostic information in the majority of the cases. However, false-negative biopsy results are common, as the disease is typically patchy in distribution. MRI scans of muscles may be helpful in selection of a site for muscle biopsy in patients with suspected inflammatory myopathy when a first muscle biopsy turns out to be negative. It may also provide diagnostic information through the form and anatomic distribution of the pathology. Connor A et al reported 2 cases illustrating the advantages of whole-body short tau inversion recovery magnetic resonance imaging before muscle biopsy which helped in identification of the muscles most suitable for biopsy. (2,3) In our case report , though muscle biopsy findings were consistent with inflammatory myopathy, changes were very minimal making diagnosis difficult. Patient did not consent for repeat muscle biopsy.

The primary treatments for myositis include corticosteroids and other immunosuppressive agents. Apart from azathioprine and methotrexate , many other immunosuppressive agents are studied like hydroxychloroquine, mycophenolate, cyclosporine, cyclophosphamide, and intravenous immunoglobulin. Rituximab also showed benefit in some patients.(1) Physiotherapy and exercise to rebuild muscle strength and function , however , play important role in long term care.

Plasmapheresis is used in the treatment of refractory autoimmune diseases to remove circulating autoantibodies and immune complexes. Since idiopathic inflammatory myositis is associated with the production of autoantibodies, plasmapheresis has been tried in refractory cases.

Our patient received seven cycles of plasmapheresis but results were not satisfactory. Though there was subjective improvement in myalgia but that effect can not be dissociated from that of the concomitantly administered immunosuppressive drugs.

Dau P C et al described beneficial role of plasmapheresis in a study 35 patients with IIM . They had inadequate clinical responses to steroids and cytotoxic immunosuppressive drug therapy. All patients received cyclophosphamide or chlorambucil in addition to plasmapheresis. Muscle power improved in 32 patients during combined therapy.(4) Subsequently, Clarke CR also described ameliorative effects of plasmapheresis in three patients with dermatomyositis-polymyositis resistant to other therapy, particularly prednisolone. The remission induced by plasmapheresis was temporary in one case and lasting, with low doses of prednisolone, in two.(5) However, the results of a subsequent randomized, double-blind controlled trial in 39 patients with polymyositis or dermatomyositis were disappointing.(6) Due to lack of efficacy, the cost and the potential for complications, there is little justification for using plasma exchange in patients with IIM.

An open-label trial suggested that combination methotrexate and azathioprine may benefit patients with treatment resistant myositis, including those who previously had inadequate responses to either methotrexate or azathioprine alone.(7) In the present case report ,after starting combination therapy of oral methotrexate and azathioprine, patient's power improved marginally.

Conclusion –

Plasmapheresis as a treatment modality did not help in our case of inflammatory myopathy. Combination therapy of oral methotrexate and azathioprine may benefit patient with treatment resistant idiopathic inflammatory myopathy.

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