Sarcoidosis presenting as Proximal Myopathy

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Introduction

Sarcoidosis is a multi-system inflammatory disorder of unknown cause. Most patients with this disease present with respiratory symptoms as lungs are frequently affected organs. Symptomatic muscle involvement is rare[1] and presentation of this disease as myopathy is even rarer.[2] We describe a case of a lady who presented with severe disabling proximal myopathy as the only feature of underlying sarcoidosis. Finding of non-caseating granulomas on muscle histopathological examination and raised angiotensin converting enzyme levels (ACE) clinch the diagnosis. To our knowledge such a presentation of sarcoidosis has never been reported from Pakistan.

Case Report

A middle aged lady presented with 4 months history of difficulty in rising from squatting position along with walking difficulty and painful thighs. Swelling of ankle and feet for 6 months preceded these symptoms. There was no history of fever and weight loss or any other focal symptoms. On examination she was overweight, had normal BP, pulse and temperature. Systematic examination revealed marked proximal muscle weakness. Her Hb was 12.4gm/dl, white cell count of 7000/mm^3 with 56% of neutrophils and 35% of lymphocytes and ESR 25mm/first hour. Blood sugar, electrolytes, uric acid, thyroid, liver, kidney functions and urine analysis and x-ray chest were normal. Creatinine phosphokinase (CPK) was 72 IU/L and serum calcium 9.0mg/dl. RA factor was negative. EMG demonstrated myopathic pattern and muscle biopsy from involved muscle showed chronic granulomatous inflammation and possibility of sarcoidosis was questioned. Angiotensin converting enzyme levels was 198U/L (normal 8-52.0). Her eye examination was also unremarkable. In spite of restrictive pattern of pulmonary function tests, she had no respiratory symptoms. She was given steroid and within few days., she was able to walk and her proximal muscle weakness improved considerably. The initial tapering of steroids made her symptoms worse so they were tapered rather slowly at the cost of making her cushingoid and glucose intolerant. Addition of methotrexate at this stage helped in tapering of steroids. Her glucose values became normal and ACE levels returned to normal, her symptoms improved remarkably. For the last 2 years she is on 5mg prednisolone and 7.5mg/week of methotrexate and doing well.

Discussion

Sarcoidosis is a systematic granulomatous inflammatory disorder of unknown origin. It has protean manifestations and can affect any organ including bones, joints, muscles and vessels.[3] Most patients present with acute or chronic respiratory symptoms as lungs are frequently affected organs. Symptomatic muscular involvement is rare[1] and presentation of this disease as isolated case of myopathy is even rarer.[2]

The usual presentation of those with symptomatic disease is severe disabling proximal muscle weakness and pain as in this patient. Most patients are less than 40yrs old. These patients may also present with diffuse muscular atrophy or tumor like growth.[4] Diagnostic workup include muscle enzymes, EMG and muscle biopsy. Most of the patients have raised creatinine phosphokinase (CPK) although some may have normal (CK) as in this case.

EMG usually shows a myopathic pattern. Histologically, non-caseating granulomas are typically present but are non-diagnostic (Figures 1, 2, 3). It is essential to look for other organ involvement like eyes, joints, lymph, nodes and lungs. Angiotensin enzyme level is not diagnostic but supports the diagnosis. It helps in follow-up if high initially, it was raised in this patient and came down with treatment. Muscle MRI is considered to be useful in diagnosing muscle sarcoidosis though was not done in this case.[5] Asyptomatic pulmonary involvement is quite common as in our case where PFTs were abnormal but the patient was asymptomatic. Corticosteroid remain the first line therapy. Alternative to corticosteroids are introduced either because of steroid intolerance and or in attempt to reduce steroid dose and side effects. In this case low dose methotrexate was used as steroid sparing agent because of development of glucose intolerance and cushing syndrome. The results were excellent as were shown by others.[6] Our case highlights the fact that although symptomatic muscle sarcoidosis is rare disease, it should be considered in patients presenting with muscular pain and weakness.

References

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