A Case Series of unusual presentations of Immune-Mediated Polymyositis

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Introduction
Idiopathic inflammatory myopathies involve four major subtypes that include: Polymyositis, Dermatomyositis, Inclusion Body Myositis and Necrotizing myopathy. Polymyositis is an autoimmune and chronic inflammatory disease characterized by symmetrical proximal muscle weakness due to the involvement of endomysial layers and skeletal muscles.
Polymyositis develops due to abnormal activation of cytotoxic T lymphocytes (CD8 cells) and macrophages against muscular antigens.

Polymyositis mostly affects individuals already suffering from some kind of disease due to viral infections like HIV, HTLV-1, Hepatitis C or malignancies such as lung carcinoma, lymphomas [1].
Polymyositis can also occur in patients meeting criteria for other collagen vascular disease example rheumatoid arthritis, systemic lupus erythematosus or progressive systemic sclerosis.[7]

Rarely patients on statin therapy can develop Polymyositis.

The main symptoms include:
- Difficulty in swallowing
- Difficulty speaking
- Arthralgia and fatigue
- Shortness of breath
- Pain in multiple joints.

The predominant symptom of Polymyositis is muscle weakness. The weakness is symmetrical and affects proximal muscles of extremities as well as neck flexors. Patient experiences difficulty in raising arms, getting up from sitting position or climbing stairs.[2]
The signs and symptoms of polymyositis typically develop gradually in middle aged adults and more women are affected than men.[6]

Main diagnostic tests include:
1. Muscle enzymes- lab markers of muscle injury include elevation in the blood level of CK (creatinine kinase), aldolase, AST (aspartate transaminases), ALT (alanine transaminases) and LDH (lactate dehydrogenase). CK is the most reliable enzyme to measure.
2. Autoantibodies- A variety of autoantibodies may be present in the serum of patients with Polymyositis. Some of these such as ANA (antinuclear antibodies) and antibodies to ribonucleoprotein (RNP).
3. MRI- it can demonstrate muscle inflammation clearly in Polymyositis.
4. Muscle biopsy- it is the gold standard for Polymyositis diagnosis. It can demonstrate muscle fibers in various stages of inflammation, necrosis, regeneration, endomysial infiltration by mono nuclear cells, capillary obliteration, endothelial cell damage and increased amounts of connective tissue. [2]

EMG should always be considered as an extension of the clinical examination since it can be misinterpreted. EMG findings include increased insertional activity, polyphasic short, small MUAP with low amplitude and short duration, positive sharp and high frequency repetitive discharge and low level of contraction. These changes are nonspecific but are useful in distinguishing myopathic causes of weakness from neuropathic disorders. It is also used for selecting appropriate muscles for biopsy.[3]

Case series

Sr. No.	Date	Case Details	Signs and Symptoms	Outcome
1	25th November 2024	A 49-year-old female patient, came with history of progressive generalized weakness and decreased oral intake. Weakness affected proximal upper limb and lower limb symmetrically. She had previous history of dengue NS1 positive 1 month ago.	Weakness affected both proximal upper and lower limb. She had generalized anasarca. Her reflexes were +1 in both upper and lower limb and plantars were flexors. Her power was 3/5 in bilateral upper and lower limb. She also had complains of dysphagia and past history of high grade fever.	Her muscle biopsy was suggestive of Polymyositis. Patient was given 1 gm Methyl Prednisolone pulse therapy for 5 days and then started on tapering dose of steroids. Patient came on follow up and was able to stand and walk on her own. Patient was discharged on Azathioprine (50) BD for 1 month
2	24th December 2024	A 23-year-old male patient with previous history of Dengue NS1 positive 2 weeks ago came with complaints of difficulty walking, lifting heavy weights and raising his arms above his shoulders. His CPK Total was more than 1800 and CPK-MM was 46,440. His muscle biopsy was suggestive of polymyositis	Patient had bilateral symmetrical proximal muscle weakness and truncal weakness. Patient also had previous history of high grade fever associated with chills and rigor, joint pain and retro orbital pain. He had generalized anasarca, +1 reflexes and 3/5 power in both upper and lower limb.	Patient was given Methyl Prednisolone 1gm in pulse therapy and discharged on tapering dose of steroid and azathioprine (50) BD. Patient on follow up had significant improvement in walking and reduction of anasarca.
3	6th May 2023	A 28-year-old female patient experiencing progressive weakness in her shoulders, hips and thighs with difficulty in rising up from chair or climbing stairs. Patient had elevated muscle enzyme and biopsy was suggestive of polymyositis. Her ANA profile was suggestive of Sjogren's Syndrome	Patients had symptoms of proximal upper and lower limb weakness with generalized anasarca with symptoms of dry eyes and dry mouth.	Patient was started on methyl prednisolone 1 gm pulse therapy along with tapering dose of steroids. Patient was also started on hydroxychloroquine (200) BD. Patient had significant symptomatic relief when she came on follow up
4	18th August 2023	A 23-year-old female patient with 8 months of amenorrhea and previous history of dengue (recovered) 1 month ago came with progressive proximal muscle	Progressive proximal muscle weakness with tiny petechial rash in the truncal area with past history of high grade fever. Her reflexes were +2 and 2/5 power in both upper and lower limb.	Patient underwent elective Cesarean Section and later was given 1 gm Methyl Prednisolone for 5 days . Patient was discharged on

		weakness. Her muscle biopsy was suggestive of polymyositis	Her plantars were flexors.	tapering dose of steroids.
5	11th September 2023	A 30-year-old female patient came with chief complaint of generalized anasarca and progressive muscle weakness. Her serum TSH was 0.001(1st time diagnosed hyperthyroidism) and free T3 & free T4 were in normal range. Her muscle biopsy was suggestive of polymyositis	Generalized anasarca with swelling of proximal joints and progressive muscle weakness with bulging eye appearance. She also complains of unexplained weight loss despite increased appetite, tachycardia, excessive sweating and trouble sleeping.	Patient was started on Tablet Neo-mercazole (10) BD, Tablet Propranolol- Long Acting (10) BD and Methyl Prednisolone pulse therapy. Her symptoms gradually resolved over a period of month.

DISCUSSION

Polymyositis is a rare disease whose etiology is not fully understood and requires a meticulous diagnostic approach.

In addition to causing disability and affecting the quality of life it is also associated with 10% mortality rate and a grave prognosis.

It affects distal musculature of esophagus in the later stages leading to inability to swallow as well as regurgitation problems that can cause aspiration pneumonia.

Heart abnormalities are frequent in Polymyositis, most of which were subclinical. Heart failure is the most frequent (32% to 72%). The efficacy of glucocorticoids and immunosuppressants here is uncertain.[4]

The muscle biopsies of all our patients were characteristically showing features of Polymyositis. So muscle biopsy is easily diagnostic tool of choice in these cases.

The goals of therapy are:

1. To reduce muscle weakness
2. To avoid the development of extra muscular diseases of vital organs.

Corticosteroids are the 1st line of therapy for Polymyositis. The usual starting dose is 1mg/kg/day for 6 weeks. The response to therapy should be addressed every 2-4 weeks by monitoring the proximal muscle strength, muscle enzyme levels and patient functionality.

In a steroid responsive patient, the goal is to attain the lowest dose of steroids that will adequately manage the disease. Azathioprine is given orally at a dose of 1.5-3 mg/kg/day. [2]

Most patients with Polymyositis respond to methotrexate, however this agent should be avoided in patients with interstitial lung disease and test positive for anti JO-1 antibodies due to possible result of pulmonary fibrosis [5]

Ivig (Immunoglobulins) can be given in patients who are corticosteroid resistant specially when it is rapidly progressing or life threatening. The initial dose is 2g/kg/ month.[2]

The development of diagnostic protocols that couple laboratory, histopathological and imaging procedures has greatly facilitated the precise identification of myopathies with inflammatory etiology which in turn allows hiring of an effective therapeutics.[8]

Incorrect interpretation can lead to mismanagement, delayed treatment, disability and high risk of complications related to management and death.

CONCLUSION

Thus patients of immune mediated Polymyositis can have unusual presentations as seen in this case series, which needs to be diagnosed and managed aggressively to prevent further deterioration.

We look forward to contributing further insights to this article in the future as we encounter additional Cases of Polymyositis.

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