https://doi.org/10.48047/AFJBS.6.Si4.2024.5620-5623

African Journal of Biological Sciences

A RARE CASE OF POLYMYOSITIS

DR SHWETHA BM[1*], DR MOUNA B M[2],DR MURTHY TV[3]

[1*,3]Department of General medicine ,Sri Siddhartha Medical College, Tumkur
[2]–Department of General medicine ,All India Institute of Medical Sciences, New Delhi

***Corresponding Author:** DR SHWETHA BM
*Department of General medicine ,Sri Siddhartha Medical College, Tumkur

Volume 6, Issue Si4, August 2024

Received: 13 July 2024

Accepted: 15 July 2024

Published: 8 August 2024

doi: 10.48047/AFJBS.6.Si4.2024.5620-5623

Abstract:
Idiopathic inflammatory myopathies (IIMs) are connective tissue diseases of unknown etiology, characterized by progressive mononuclear cell infiltration in muscles. Polymyositis (PM), a rare subset of IIMs, is clinically defined by progressive proximal muscle weakness with a symmetric distribution. We present a 50-year-old female with a 3-month history of progressive proximal muscle weakness. Laboratory studies showed elevated creatine kinase (CK) and inflammatory markers, with electromyography indicating irritable myopathy. Muscle biopsy confirmed polymyositis, showing characteristic histopathological features. The patient was treated with steroids, resulting in significant improvement in muscle strength and continued remission at a one-month follow-up. This case highlights the importance of early diagnosis and treatment to prevent severe complications.

Key words – polymyositis , inflammatory myopathies ,proximal muscle weakness, creatine kinase ,muscle biopsy

Introduction:

Idiopathic inflammatory myopathies (IIMs) are connective tissue diseases of unknown etiology, characterized by progressive mononuclear cell infiltration in muscles. Polymyositis (PM), a subset of IIMs, is a heterogeneous group of disorders that presents with symmetrical and proximal muscle weaknesses that worsens over several weeks to months.(1)Polymyositis as a stand alone entity is a rare disorder(2)It is considered a diagnosis of exclusion; therefore, multiple pathologies must be ruled out before diagnosis.Diagnostic evaluation includes laboratory studies showing elevated sarcoplasmic enzymes, with muscle biopsy being the gold standard for diagnosis. If left untreated, PM can progress to involve respiratory and pharyngeal muscles, potentially leading to life-threatening complications. Thus, early diagnosis and treatment of this rare condition are crucial for preventing severe outcomes. We present a case of a middle aged female patient who presented with symmetric proximal upper and lower limb weakness with no other neurological involvement and her laboratory and pathology studies were confirmatory of this rare disease.

Case Report: A 50 Year old female presented with progressive proximal muscle weakness for 3 months. She initially noted difficulty in standing from a seated position, which progressed to weakness with ambulation. One week prior to presentation, she experienced difficulty in combing

hair and lifting objects above her head. She had no history suggestive of distal muscle involvement.She denied constitutional symptoms.she had no history of chronic drug usage or any addictive habits. She had no prior co morbid conditions . On examination she demonstrated 3/5 proximal muscle weakness at both Upper and Lower limb with an otherwise unremarkable neurologic examination. There were no dermatological manifestations.

Creatine kinase (CK) was increased at 1668 IU/L with concomitant elevated inflammatory markers, ESR of 45mm/hr, CRP of 82 mg/dl, TSH of 2.8 mIU/L.Myositis antibody panel was negative. Electromyography demonstrated features consistent with irritable myopathy. Right biceps biopsy H and E sections showed Transversely cut skeletal muscle fibers with effaced fascicular architecture,rounded to polygonal myofibers with variation in size. Scattered myophagocytosis,regeneration and necrotic fibers were seen. Marked infiltration seen in the interstitium with a few inflammatory cells infiltrating into normal myofiber.Findings were consistent with diagnosis of Polymyositis.Right biceps biopsy confirmed the diagnosis of polymyositis(figure 1 .(A) and (B)).

Figure 1

(A)

Gross details	Received 2 bottles Bottle 1) Contains tiny muscle bit measuring 0.2x0.2cm. All processed – A1. Bottle 2) Contains muscle bit measuring 0.5x0.2cm. All processed-B1		
Biopsy (Fresh)	**Enzyme histochemistry (Yes)**	**Tissue preserved (Yes)**	**Unstained slides(Yes)**
Clinical diagnosis: Polymyositis/ Dermatomyosits	**Site of biopsy :** Right biceps.		

Microscopic findings:

PARAFFIN SECTIONS	
Quality of fixation	Optimal
Haematoxylin& Eosin (HE)	Transversely cut skeletal muscle fibers with partly preserved fascicular architecture. Myofibers are rounded with variation in fiber size. Scattered atrophic fibers are seen. Marked lymphocytic infiltration is seen in the interstitium. Few fibers show splitting and internalized nuclei.Many scattered regenerating and myophagocytic fibers seen.
Masson's trichrome (MAT)	

(B)

CRYOSECTIONS	
Quality of fixation	Optimal
Haematoxylin& Eosin (HE)	Transversely cut skeletal muscle fibers with effaced fascicular architecture. Myofibers are rounded to polygonal with variation in size. Atrophic fibers are seen. Marked infiltration is seen in the interstitium. Scattered myophagocytosis, regeneration and necrotic fibers seen. A few inflammatory cells are seen infiltrating into normal myofiber. No perifascicular atrophy.
Modified Gomoritrichrome (MGT)	No ragged red fibers or inclusions
Oxidative enzyme -SDH	No ragged blue fibers.
Oxidative enzyme- NADH	
ATPase (pH 9.4, 4.6)	Predominantly type II fiber atrophy.
COX – SDH:	Few COX deficient fibers seen. (secondary)

Impression: Inflammatory myopathy - polymyositis; right biceps.

A CT scan of the abdomen, chest, and pelvis revealed no signs of malignancy.During her initial hospitalization, she was treated with steroids; there was improvement in her muscle strength over a period of one week. She was discharged with steroids and was seen in follow up one month later and continued to be in remission

Discussion: The inflammatory myopathies represent the largest group of acquired and potentially treatable causes of skeletal muscle weakness. They are classified into 3 major groups. Polymyositis,Dermatomyositis , Inclusion body myositis.Polymyositis represents 5% of autoimmune myositis.(3)Polymyositis mimics several other myopathies and is a diagnosis of exclusion.It is a subacute inflammatory myopathy that affects adults and rarely children,who do not have any of the following: rash, involvement of the extra ocular and facial muscles, family h/o neuromuscular

disease, h/o exposure to myotoxic drugs or toxins, neurogenic disease, endocrinopathy, muscular dystrophy, biochemical muscle disorder. PM occurs in association with a systemic autoimmune or connective tissue disease or with a known viral or bacterial infection. Patients with Polymyositis present with progressive and symmetric muscle weakness. They usually report increasing difficulty with everyday task requiring the use of proximal muscles, distal muscles are affected only late in the course of PM.Pharyngeal and neck flexors are often involved which can result in aspiration pneumonia.In advanced cases respiratory muscles may also be affected. (4)Diagnosis of PM is made after excluding other causes of proximal muscle weakness with elevated muscle enzymes. Diagnosis is confirmed by analysis of serum muscle enzymes, EMG findings and conducting a muscle biopsy(4).CK can be elevated as much as 50-fold.Needle EMG shows myopathic potentials characterized by short duration, low-amplitude polyphasic units on voluntary activation and increased spontaneous activity with fibrillations, complex repetitive discharges and positive sharp waves.Inflammation is the histologic hallmark. Treatment includes steroids and other immunosuppressive drugs.High-dose glucocorticoids are the initial treatment for polymyositis. Oral prednisone 1 mg/kg is recommended, and in severe and rapidly worsening disease, intravenous methylprednisolone 1 g/dose once daily for 3-5 days may be used(5).In patients with a rapidly worsening disease, severe life-threatening weakness, dysphagia with a high risk for aspiration, or resistant disease, intravenous immunoglobulin G (IVIG) is recommended . When glucocorticoids and intravenous immunoglobulin are not effective, treatment escalation with rituximab, a monoclonal antibody against CD20, should be considered.

Conclusion: A rare disease like Polymyositis must be suspected in evaluating a case of proximal muscle weakness with elevated creatine kinase and inflammatory markers. If left untreated, PM can progress to involve the muscles of respiration and deglutition, potentially leading to life-threatening complications. Thus, early diagnosis and treatment of this rare condition are crucial for preventing severe outcomes. 5 year survival rate for treated patients with PM is 95% and 10 year survival rate is 84%. Most patients improve with therapy and many make full functional recovery, which is often sustained with maintenance therapy

References

1. Polymyositis, dermatomyositis and inclusion body myositis.in:Loscalso J Fauci AS Kasper DL Hauser SL Longo DL Jameson JL Harrison's principles of internal medicine. McGraw-Hill, New York 2021: 2819-2820
2. Inflammatory muscle disease - An update. Baig S, Paik JJ. Best Pract Res Clin Rheumatol. 2020;34:101484. [PubMed] [Google Scholar]
3. Epidemiology of adult idiopathic inflammatory myopathies in a U.S. managed care plan. Furst DE, Amato AA, Iorga ŞR, Gajria K, Fernandes AW. Muscle Nerve. 2012;45:676-683. [PubMed] [Google Scholar]
4. Polymyositis and dermatomyositis. Bohan A, Peter JB. New Eng Jr Med. 292:344-347. [PubMed] [Google Scholar]
5. Inflammatory myopathies: how to treat the difficult cases. Mastaglia Mastaglia, F, & Zilko, P. . (2003. Journal of Clinical Neuroscience. 10:99-101. [PubMed] [Google Scholar]
6. Current classification and management of inflammatory myopathies. Schmidt J. J Neuromuscul Dis. 2018;5:109-129. [PMC free article] [PubMed] [Google Scholar]