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RESEARCH ARTICLE

SEVERE BULBAR-PREDOMINANT DERMATOMYOSITIS WITH MI-2A ANTIBODIES PRESENTING AS PROGRESSIVE QUADRI-PARESIS: A CASE REPORT

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Abstract

Background: Idiopathic inflammatory myopathies (IIMs) are autoimmune disorders characterized by skeletal muscle inflammation and progressive weakness. Dermatomyositis is a major subtype distinguished by characteristic cutaneous manifestations in addition to muscle involvement. Dysphagia may occur during the course of disease; however, severe bulbar involvement requiring enteral feeding is relatively uncommon and may delay diagnosis.

Case Presentation: We report a 45-year-old male farmer who presented with a two-month history of progressive proximal limb weakness and severe dysphagia. The illness initially began with bilateral facial swelling and violaceous periorbital rash that resolved spontaneously after several weeks. Subsequently, the patient developed progressive proximal quadri-paresis, hoarseness of voice, and severe oropharyngeal dysphagia requiring Ryle's tube feeding. Neurological examination revealed pure motor proximal-predominant quadri-paresis with bulbar involvement. Laboratory investigations demonstrated markedly elevated muscle enzymes. Electromyography showed myopathic motor unit potentials, and MRI of the thighs revealed diffuse symmetrical muscle edema suggestive of inflammatory myositis. A myositis-specific antibody panel was weakly positive for Mi-2 antibodies. Treatment with intravenous methylprednisolone followed by intravenous immunoglobulin and azathioprine resulted in significant improvement in muscle strength.

Conclusion: This case highlights an atypical presentation of dermatomyositis characterized by severe bulbar dysfunction and transient cutaneous manifestations. Early recognition and prompt immunotherapy are essential to prevent complications and improve functional outcomes.

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Introduction:-

Idiopathic inflammatory myopathies are a group of immune-mediated disorders characterized by chronic muscle inflammation leading to progressive weakness and disability. The principal subtypes include Dermatomyositis, Polymyositis, Inclusion Body Myositis, and immune-mediated necrotizing myopathy. These conditions differ in clinical presentation, immunological features, and prognosis. ¹Dermatomyositis is characterized by symmetric proximal muscle weakness accompanied by distinctive dermatological findings such as heliotrope rash, Gottron's papules, and photosensitive rashes. The disease pathogenesis involves complement-mediated microangiopathy affecting small blood vessels in skeletal muscle and skin. ²Bulbar involvement may occur due to weakness of the striated muscles of the pharynx and upper esophagus. Dysphagia has been reported in approximately 10–30% of patients and may lead to aspiration pneumonia and malnutrition. ³ Severe dysphagia requiring enteral feeding, however, is relatively uncommon. Myositis-specific autoantibodies have improved the classification of inflammatory myopathies. Among these, Mi-2 antibodies are highly specific for dermatomyositis and are often associated with favorable treatment response. ⁴We report a case of dermatomyositis presenting with progressive quadriparesis and severe bulbar dysfunction in which the characteristic cutaneous manifestations resolved prior to medical evaluation.

Case Presentation:-

A 45-year-old right-handed male farmer presented with a two-month history of progressive weakness and difficulty swallowing. The illness began with bilateral facial swelling associated with dark discoloration around both eyes. The swelling was painless and gradually subsided spontaneously over approximately four weeks. Approximately two weeks later, the patient developed progressive dysphagia to both solids and liquids, characterized by choking sensation and nasal regurgitation. The dysphagia worsened over time and resulted in pooling of saliva and inability to maintain adequate oral intake, necessitating Ryle's tube feeding. During the same period, the patient developed progressive weakness of the limbs. Initially he experienced difficulty rising from a squatting position and gradually required assistance for standing. Weakness progressed to involve the upper limbs, resulting in difficulty performing activities of daily living such as dressing and bathing. Subsequently, truncal muscle weakness developed, causing difficulty lifting the neck, turning in bed, and sitting up from a lying position. Hoarseness of voice was also present. There was no history of sensory symptoms, diplopia, fluctuating weakness, weight loss, fever, or joint pain. Past medical history and family history were unremarkable.

Examination:-

General examination showed stable vital signs. No active rash was present at the time of evaluation. Neurological examination revealed pure motor quadriparesis with proximal predominance. Proximal muscle strength was significantly reduced in both upper and lower limbs, while distal strength was relatively preserved. Truncal and neck flexor weakness were present. Bulbar examination revealed dysphagia and hoarseness of voice. Sensory examination was normal and deep tendon reflexes were preserved. Single breath count was 28 and chest expansion was 4 cm. Routine blood investigations were within normal limits. Autoimmune screening including antinuclear antibody and antineutrophil cytoplasmic antibody testing was negative. Serum creatine kinase was elevated to **3963 IU/L**, and lactate dehydrogenase was 919 IU/L, indicating active muscle injury. Nerve conduction studies were within normal limits. Electromyography showed short-duration low-amplitude polyphasic motor unit potentials with reduced recruitment, consistent with inflammatory myopathy. ⁵A myositis-specific antibody panel was weakly positive for Mi-2 antibodies. Upper gastrointestinal endoscopy performed to evaluate dysphagia was normal. Magnetic resonance imaging of the thighs demonstrated diffuse symmetrical T2 hyperintensity involving multiple proximal muscle groups, including the psoas, gluteal, adductor, and quadriceps muscles, consistent with inflammatory myositis. ⁶Malignancy screening was negative.

Management and Outcome:-

The patient was treated with intravenous methylprednisolone pulse therapy followed by intravenous immunoglobulin therapy as per the body weight with 2 mg per kg dosage. Long-term immunosuppressive therapy with azathioprine was initiated. Supportive management included swallow therapy, speech therapy, early mobilization, and physiotherapy. The patient showed gradual improvement. The Medical Research Council (MRC) sum score improved from 44 at admission to 60 at discharge. He was discharged ambulatory without support, although Ryle's tube feeding was continued. At one month follow up patient was completely asymptomatic and Ryle's tube was also removed.

Discussion:-

Dermatomyositis is characterized by immune-mediated microvascular injury leading to muscle inflammation and weakness.² Cutaneous manifestations typically precede muscle weakness and are often essential diagnostic clues.¹ However, in our patient the skin manifestations resolved spontaneously before evaluation, obscuring the diagnosis. Dysphagia occurs in approximately 10–30% of patients with dermatomyositis due to involvement of pharyngeal musculature.³ Severe dysphagia requiring enteral feeding is less common but may result in aspiration and nutritional compromise. Bulbar-predominant presentations may mimic other neuromuscular disorders such as Myasthenia Gravis.⁵ In the present case, the absence of fluctuating weakness and the presence of markedly elevated muscle enzymes supported inflammatory myopathy. Myositis-specific antibodies play an important role in diagnosis and prognosis. Mi-2 antibodies are strongly associated with dermatomyositis and favorable therapeutic response.⁴ Magnetic resonance imaging has emerged as a valuable non-invasive tool for identifying muscle inflammation and guiding diagnosis.⁶ Early treatment with corticosteroids and immunomodulatory therapies is essential to prevent complications and improve functional outcomes.⁷ The favorable response observed in our patient highlights the importance of early recognition and prompt therapy.

Conclusion:-

Dermatomyositis can present with diverse and atypical manifestations. This case illustrates an unusual presentation characterized by severe bulbar dysfunction and progressive quadriparesis with transient cutaneous manifestations. Clinicians should maintain a high index of suspicion for inflammatory myopathy in patients presenting with progressive proximal weakness and dysphagia, even in the absence of active skin lesions. Early diagnosis through integration of clinical findings, muscle enzymes, electrophysiology, autoantibody testing, and MRI is crucial. Prompt immunotherapy and multidisciplinary management can significantly improve functional recovery and reduce disease-related complications.



Figure 1: Axial pelvic MRI demonstrating hyperintensity within bilateral piriformis muscles suggestive of active myositis.

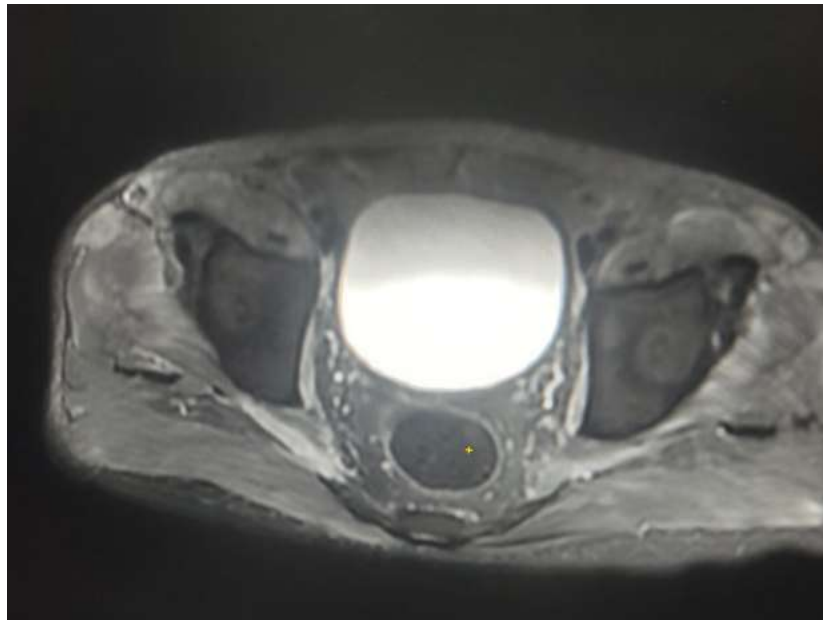


Figure 2: Axial T2-weighted fat-suppressed MRI showing diffuse hyperintensity in obturator and adductor muscle groups.

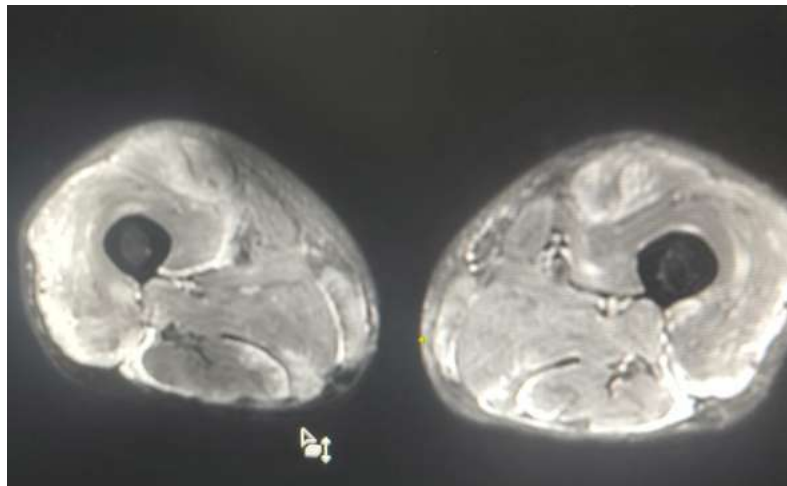


Figure 3: Axial T2-weighted fat-suppressed MRI of the thighs demonstrating symmetrical inflammatory changes involving multiple muscle groups.

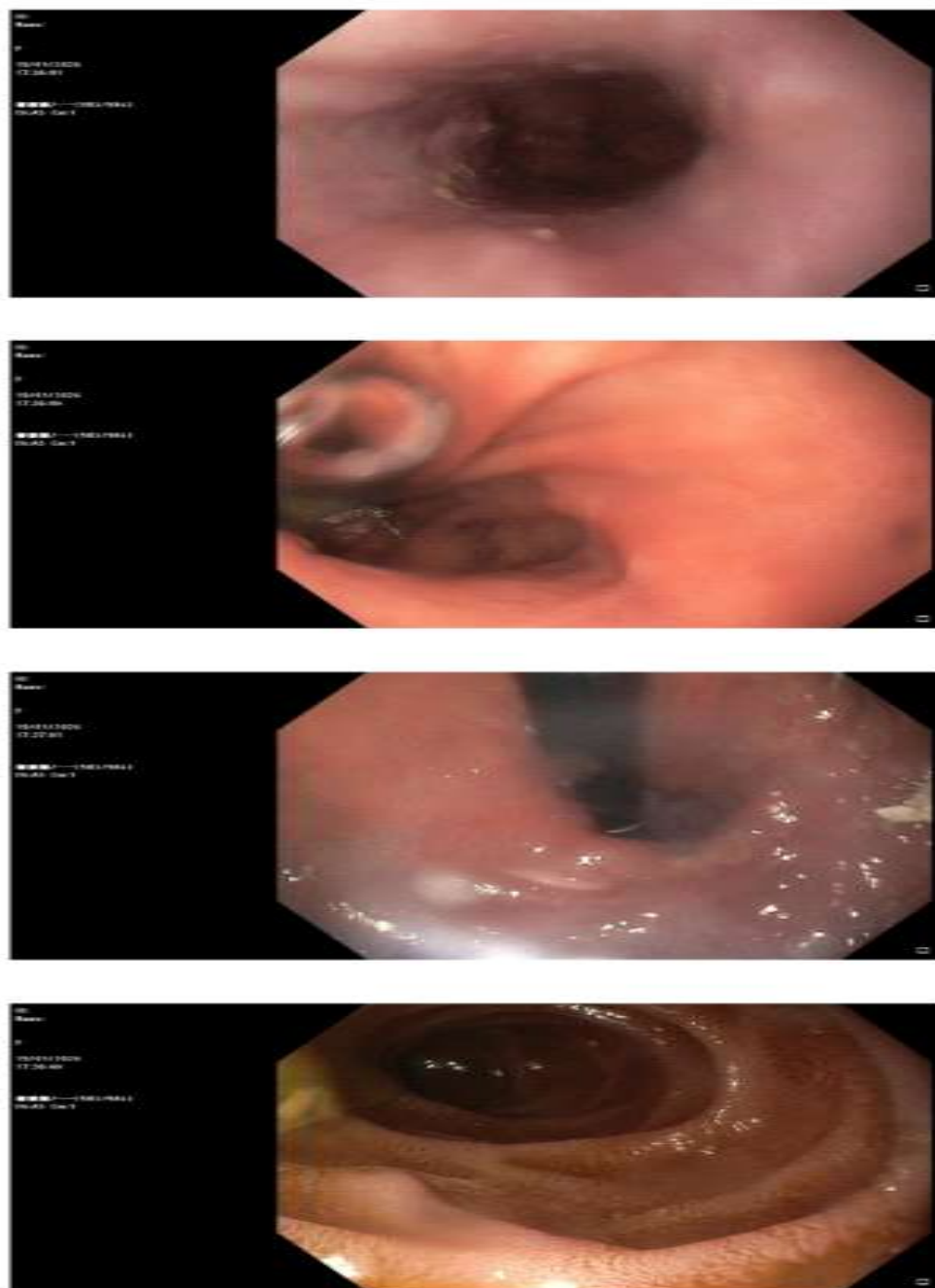


Figure 4: Upper gastrointestinal endoscopy which was within normal limits.

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