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Electrodiagnostics

Atypical Electrodiagnostic Findings in a Patient with Immune-Mediated Necrotizing Myopathy: A Case Study

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Autoimmune Necrotizing Myositis (NAM)/Immune-Mediated Necrotizing Myositis (IMNM), a progressive myopathy, lacks skin lesions and exhibits unique pathologic traits. Risk factors like statin use and malignancy are implicated, while most IMNMs are idiopathic. Manifesting as insidious proximal muscle weakness, they often lead to difficulties in functional tasks early in the disease. This article presents a case of a 65-year-old male with Immune-Mediated Necrotizing Myopathy exhibiting atypical needle electromyography findings with widespread denervation in multiple limbs in a proximal distribution. The patient complained of bilateral hip/thigh weakness and symmetric numbness in the feet, among other symptoms. Evaluation revealed significant findings including elevated creatine kinase (CK) levels, Anti 3-hydroxy-3-methylglutaryl-coenzyme A (HMG-CoA) antibodies, and abnormal electromyography indicating motor axon/myopathic involvement. Urgent referrals led to diagnosis confirmation and subsequent immunotherapy. This case highlights the challenges in diagnosing myopathies, emphasizing the importance of comprehensive patient history and clinical examinations. Electrodiagnostic studies complement clinical assessments, aiding in confirming myopathy and suggesting alternative diagnoses. Statin-induced necrotizing immune myopathy, associated with anti HMG-CoA antibodies, poses serious risks, warranting statin cessation and cancer screening. Recognizing when to refer patients promptly is crucial to minimize diagnostic delays. Prognosis for IMNM patients is challenging, with persistent weakness despite intensive treatment, emphasizing the need for a multifaceted approach in diagnosis, management, and patient care. Understanding clinical presentations, utilizing diagnostic tools judiciously, and recognizing risk factors are vital for optimizing outcomes in these complex neuromuscular disorders.

BACKGROUND

Inflammatory myopathies (IM) constitute a group of complex disorders within the realm of autoimmune diseases, characterized by inflammation and loss of skeletal muscle fibers.[1] These conditions, are relatively rare with a global incidence between 1.6-19 in 1 million and 1 in 100,000 in the United States. Myopathic conditions pose significant challenges to both patients and healthcare providers due to their diverse clinical presentations, variable disease courses, and the intricacies surrounding their diagnosis and management.[2,3] Autoimmune Necrotizing Myositis (NAM) / Immune-Mediated Necrotizing Myositis(IMNM) is a unique progressive myopathy with distinct pathologic features. NAM/IMNM presents as an insidious and progressive onset of proximal muscle weakness without skin lesions.[3,4] Several risk factors including the use of statins and malignancy have been identified, whereas a majority of cases of IMNM are idiopathic in nature.[5] The most common symptom of a myopathic condition is weakness with a predilection for proximal muscles although variations and unusual patterns of weakness are seen in some muscular dystrophies and other neuromuscular conditions.[3] Due to the slow progressive onset of proximal muscle weakness in some myopathies, the manifestations early in the disease typically involve difficulty with functional tasks such as rising from a chair.[6]

Electrodiagnostic testing (EDX), which includes nerve conduction studies and needle electromyography, is a diagnostic tool, used as an extension of the clinical examination, to help assist the practitioner in determining location, severity, and extent of neuromuscular conditions including myopathy. While EDX and other diagnostic tests such as neuromuscular ultrasound, muscle biopsies, and blood panels are certainly important in the evaluation process in patients with suspected neuromuscular disease, a thorough patient and family history as well as a detailed neurologic examination are a crucial first step. We present a case of a 65-year-old male with IMNM who was seen for an electrodiagnostic assessment with atypical needle electromyogra-

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