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Clinical Features, Diagnosis and Treatment Outcomes in Necrotising Autoimmune Myopathy: A Rare Entity in the Spectrum of Inflammatory Myopathies

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Abstract : Inflammatory myopathies (IMs) have long been recognised as a heterogenous family of myopathies with acute, subacute, and sometimes chronic presentation and are potentially treatable. Necrotizing autoimmune myopathies (NAM) are a relatively new subset of myopathies. Patients generally present with subacute onset of proximal myopathy and significantly elevated creatinine kinase (CK) levels. It is being increasingly recognised that there are limitations to the independent diagnostic utility of muscle biopsy. Immunohistochemistry tests may reveal important information in these cases. The traditional classification of IMs failed to recognise NAM as a separate entity and did not adequately emphasize the diversity of IMs. This review and case report on NAM aims to highlight the heterogeneity of this entity and focus on the distinct clinical presentation, biopsy findings, specific auto-antibodies implicated, and available treatment options with prognosis. This article is a meta-analysis of literatures on NAM and a case report illustrating the clinical course, investigation and biopsy findings, antibodies implicated, and management of a patient with NAM. The main databases used for the search were Pubmed, Google Scholar, and Cochrane Library. Altogether, 67 publications have been taken as references. Two biomarkers, anti-signal recognition protein (SRP) and anti- hydroxyl methylglutaryl-coenzyme A reductase (HMGCR) Abs, have been found to have an association with NAM in about 2/3rd of cases. Interestingly, anti-SRP associated NAM appears to be more aggressive in its clinical course when compared to its anti-HMGCR associated counterpart. Biopsy shows muscle fibre necrosis without inflammation. There are reports of statin-induced NAM where progression of myopathy has been seen even after discontinuation of statins, pointing towards an underlying immune mechanism. Diagnosisng NAM is essential as it requires more aggressive immunotherapy than other types of IMs. Most cases are refractory to corticosteroid monotherapy. Immunosuppressive therapy with other immunotherapeutic agents such as IVIg, rituximab, mycophenolate mofetil, azathioprine has been explored and found to have a role in the treatment of NAM. In conclusion,given the heterogeneity of NAM, it appears that NAM is not just a single entity but consists of many different forms, despite the similarities in presentation and its classification remains an evolving field. A thorough understanding of underlying mechanism and the clinical correlation with antibodies associated with NAM is essential for efficacious management and disease prognostication.

Keywords : inflammatory myopathies, necrotising autoimmune myopathies, anti-SRP antibody, anti-HMGCR antibody, statin induced myopathy
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