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Images in Rheumatology

Immune-mediated necrotizing myopathy associated with antibodies to the signal recognition particle: A rare cause of hyperCKaemia

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A 73-year-old Caucasian male patient presented to the emergency department with 3 months evolution of severe progressive proximal weakness and atrophy. Weakness was characterized by difficulty in standing up from sitting position, combing hair, and changing clothes. No history of dysphagia, dyspnea, skin rashes, or weight loss was noted. Our patient was not taking any myotoxic drugs. Clinical examinations revealed right upper and lower limb proximal and distal muscle weakness and atrophy, with a clinical power grade of 4/5. Laboratory investigation revealed an elevated sedimentation rate of 61 mm/h, with markedly elevated serum creatine kinase (CK) levels, 9562 (normal range 30-200) units/L. Antisignal recognition particle (SRP) was positive. Electrophysiologic studies evidenced diffuse myopathy with fibrillations. Necrotic fibers and regeneration, in the absence of inflammatory infiltrates, were seen in the muscle biopsy (Figure 1. a-d). The patient was initially started on prednisolone 1 mg/kg/day. His weakness improved, with a CK level of 2198 U/L at 1 month after beginning the treatment. Steroid-sparing agent was initiated with azathioprine 2 mg/kg/day, and 1 month after, he achieved a steady functional recovery.

Immune-mediated necrotizing myopathy (IMNM) is a rare clinicopathologic entity that is composed of three serologically subtypes: antihydroxy-3-methylglutaryl-CoA reductase (HMGCR) myopathy, anti-SRP myopathy,

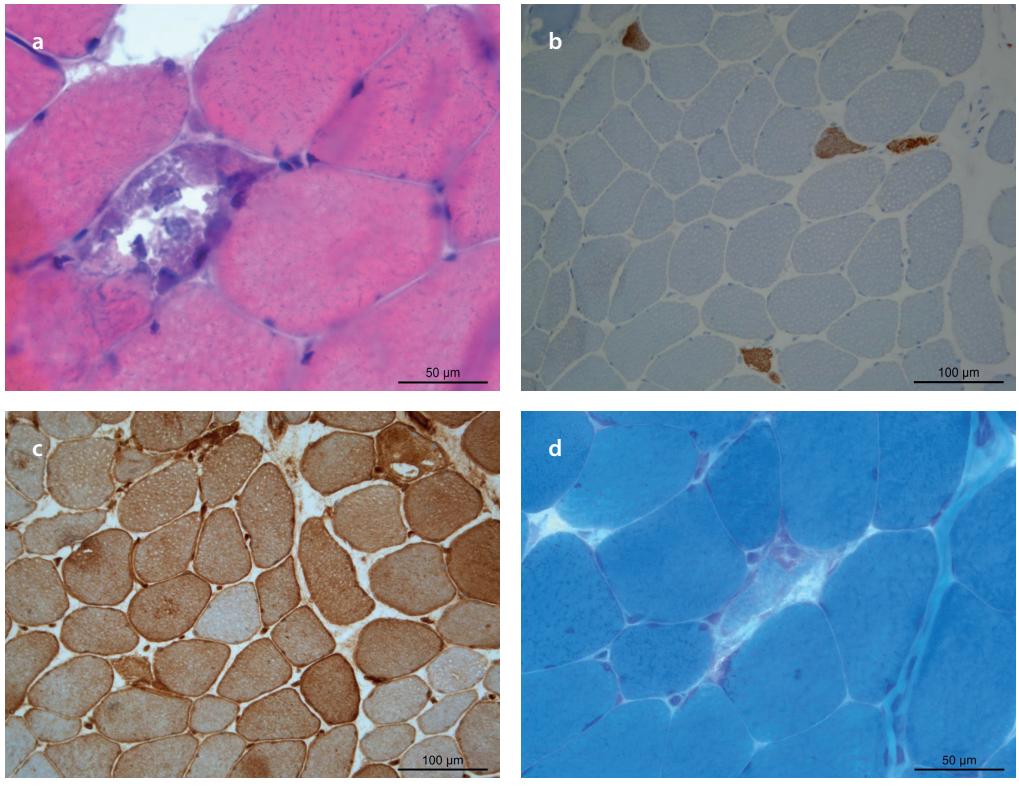

Figure 1. a-d. Left deltoid muscle biopsy showed scattered necrotic fibers without associated inflammation (a, b). There was regenerating fibers (c) and upregulation of sarcolemmal MHC class I (d).
a) H&E, b) Gomori trichrome, c) Fetal myosin; d) MHC class I

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and autoantibody-negative IMNM (1). However, only 10% of the cases have positive antibodies (1). Unlike the other subtypes, anti-SRP myopathy is not associated with cancer (2, 3) and has characteristically poor responsiveness to steroid monotherapy and others immunosuppressive therapies (4). In conclusion, we present a case of a patient with an anti-SRP antibodies-associated inflammatory myopathy, a rare diagnosis that should be considered in cases of elevated CK levels that do not resolve with appropriate management and exclusion of other causes of rhabdomyolysis.

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