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Anti-HMGCoA necrotizing autoimmune myopathy: a case report

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CASE REPORT

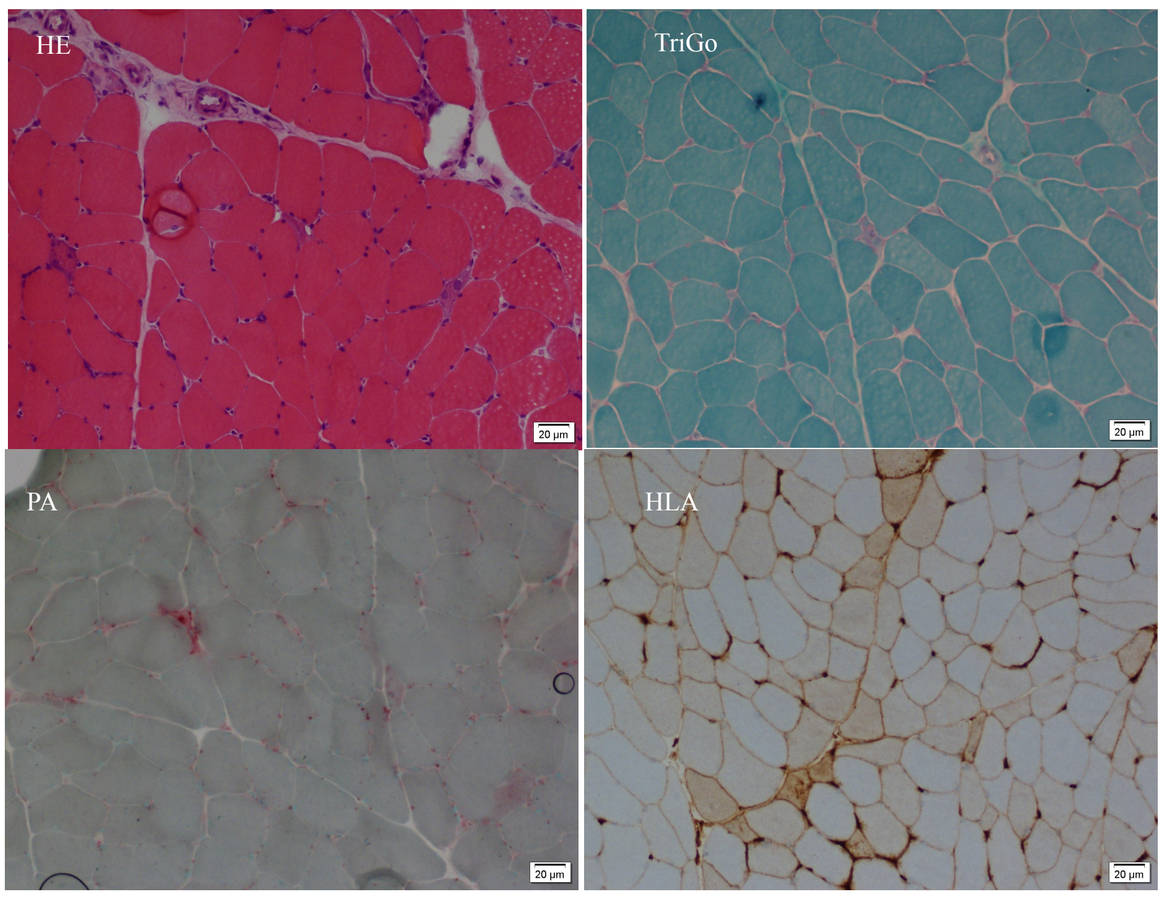
A 70 year old woman arrived to our attention for six-months history of progressive upper and lower limb muscle weakness and elevated levels of serum creatine kinase (CK) activity (4000 UI/L). In anamnesis, she reported a positive history for type 2 diabetes mellitus, treated with metformin, and hypercholesterolemia, treated with atorvastatin (in daily doses of 10 mg) for last twelve months.

Neurological examination revealed a moderate limb-girdle hyposthenia (score 4 at MRC scale). The electromyography investigations confirmed a myopathic pattern.

Quadriceps muscle biopsy showed several necrotic fibers, without an inflammatory infiltrate;

at immunohistochemical analysis several muscle fibers resulted MHC-I and HLA positive.

Necrotizing myopathy



The patient was then tested for the presence of novel **anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (anti- HMGCR) antibodies**, that resulted positive, and the diagnosis of statin-associated necrotizing autoimmune myopathy was performed.

The patient started on 50 mg of prednisone daily for 10 days and subsequent decrement 12,5 mg daily, with mild improvement; therapy with methotrexate 15 mg/ week was then added. After three months of follow up, the clinical picture further improved (last blood CK 750 UI/L).

DISCUSSION

Statins are some of the most prescribed medications. It is well-known that statins can lead to a self-limited myopathy in a minority of patients. Recently, these drugs have been also associated with a necrotizing autoimmune myopathy.
Statins are thought to represent the environmental trigger of immune-medicated necrotizing myopathy by upregulating the expression of HMG-CoA reductase and subsequently triggering an autoimmune response in the context of an appropriate genetic background.
Statin-associated necrotizing autoimmune myopathy is a relatively rare entity, but it is an important consideration in patients who develop CK elevation and weakness after statin therapy. Further research is necessary to better define statin-specific and dose-dependent risk, as well as prognosis and optimal treatments for this condition.