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A rare overlap of statin-induced anti-3-hydroxy-3-methyl-glutaryl-coenzyme A necrotizing autoimmune myositis and dermatomyositis

Key message

- This case portrays a rare overlap of statin-induced Anti-HMG-CoA necrotizing autoimmune myositis and dermatomyositis.

Dear Editor, A 73-year-old woman with a history of hyperlipidaemia had been treated with a statin for approximately 2 years. She suddenly developed persistent bilateral leg and arm weakness as well as occasional morning stiffness. Her initial blood work showed a markedly elevated creatine kinase (CK) level of 13 300 U/L and she had persistent liver enzyme elevation. Her ANA was positive and elevated at 1:1280 in a speckled pattern. MRI of her lower extremities showed a small amount of oedema and enhancement in her hamstrings. Subsequent muscle biopsy of the lower hamstrings showed multiple degenerating myofibres; multifocal perivascular and endomysial infiltrates; CD3, CD4 and CD8 lymphocytes and CD68 macrophages. Her serology was positive for anti-3-hydroxy-3-methyl-glutaryl-coenzyme A (HMG-CoA) reductase (HMGCR) antibodies > 200 and weakly positive for Ku antibodies, which were detected using ELISA. Her statin was discontinued and she was initially started on intermittent corticosteroid bursts and tapers, as well as IVIG infusions. She was also started on weekly rituximab and MTX. Due to persistent symptoms, prior medications were stopped and she was switched to MMF. During the course of this treatment she developed erythematous, pruritic plaques on her chest, which is classic of DM, these lesions occurred almost a full year after she began to experience muscle weakness. Cutaneous manifestations of DM often precede weakness, however, a minority of patients can have skin disease with no appreciable muscle symptoms or vice versa. Thus the temporal presentation of her symptoms alone cannot confirm an overlap of DM and NAM, and it becomes essential to investigate serologic and histologic findings.

Our patient had multiple findings indicative of an overlap of DM and NAM. While she did exhibit erythematous plaques on her chest, which is classic of DM, these lesions occurred almost a full year after she began to experience muscle weakness. Cutaneous manifestations of DM often precede weakness, however, a minority of patients can have skin disease with no appreciable muscle symptoms or vice versa. Thus the temporal presentation of her symptoms alone cannot confirm an overlap of DM and NAM, and it becomes essential to investigate serologic and histologic findings. While our patient had high levels of anti-HMGCR antibodies associated with a history of statin use and suggestive of NAM, her muscle biopsy did not exhibit classic features of NAM. Her histology indicated no necrosis and instead she had significant inflammation suggesting an autoimmune inflammatory myopathy. Subsequently she was referred to dermatology and received an incisional biopsy that was very typical of DM, including overexpression of MHC class I and inflammatory cells around fascicles and atrophy of muscle fibres near the border of fascicles. Thus these results are strongly suggestive of a rare overlap of DM and NAM, as the patient’s serology pointed towards NAM but her biopsy was indicative of DM.
There are no formal recommendations or specific guidelines on the treatment of NAM. However, based on prior reports, first-line treatment remains oral high-dose corticosteroids or pulse i.v. methylprednisolone. Some experts recommend IVIG as a first-line therapy, especially in anti-HMGCR myopathy [6]. AZA is the preferred agent in patients where steroids are contraindicated.

Overall, idiopathic inflammatory myopathy is a group of autoimmune disorders that typically has various distinctions in presentation, serology and histology. However, an overlapping of these disorders can exist, as outlined in this case. Thus it becomes essential to fully evaluate patients for all forms of autoimmune myopathies when they present with progressive muscle weakness and rash, as there could be a rare overlapping as evidenced in our patient. Early diagnosis can lead to fewer complications and decreased mortality.

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Data availability statement

Data are available upon reasonable request by any qualified researchers who engage in rigorous, independent scientific research, and will be provided following review and approval of a research proposal and Statistical Analysis Plan (SAP) and execution of a Data Sharing Agreement (DSA). All data relevant to the study are included in the article.

Supplementary data

Supplementary data are available at Rheumatology Advances in Practice online.

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