Autoimmune Statin-Associated Myopathy: An Unusual Case

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Objective
To present an unusual case of autoimmune HMGCR-antibody positive statin-mediated myopathy

Introduction
• Statin drugs have been widely used due to their cardio-protective and lipid lowering qualities (1). The side effect of myalgia is well known and managed by discontinuing the statin (2).
• Autoimmune statin-associated necrotizing myopathy is less familiar to physicians. This condition typically presents with symmetrical, proximal muscle weakness (3).
• However, our patient had an unusual presentation with asymmetric weakness and dysphagia resulting in significant weight loss.

Case Description
We describe a case of a 57-year-old male who presented with 5 months of worsening pain and weakness of the proximal thigh and shoulder muscles, with more severe weakness on the left side. He also related significant dysphagia, resulting in a 10kg weight loss. He had a history hyperlipidemia, treated with atorvastatin 40mg for the last 4 years. The patient initially presented to his family physician, who drew labs and instructed him to discontinue atorvastatin, suspecting myalgia, but this did not relieve his symptoms. The patients creatine phosphokinase was elevated at 7085 u/L. He was instructed to go to the ER due to concern of rhabdomyolysis.

Neurological exam was significant for reduced motor strength: bilateral elbow extension 4/5, hip flexion 3/5 on the left and 4/5 on the right; otherwise motor exam was 5/5 throughout. The patient was unable to rise from a chair without using his arms. His gait was slow and had a waddling quality. The patient underwent muscle biopsy which revealed mild necrotizing myopathy. A standard myositis panel was negative. However, the HMGCR antibody returned as positive, which allowed diagnosis of autoimmune statin-associated necrotizing myopathy. The patient started on 30mg prednisone and 150mg azathioprine and discharged but his symptoms continued to worsen. He was then admitted and started on IVIG, which resulted in significant improvement. He was discharged home after 3 days treatment.

References

Discussion and Conclusion
• Due to the autoimmune nature of this disease, it is managed very differently than its non-autoimmune counterpart. Discontinuing the statin will not result in cessation of the symptoms, and often multiple disease-modifying immunomodulatory agents must be used to gain control of the myositis and accompanying symptoms (3).
• Additionally, patients who present with the autoimmune myopathy do not tolerate re-challenge with statins favorably and usually begin exhibiting some form of myopathy upon restarting the medication (4).
• Although rare, it is important that physicians are able to recognize the various types of statin-associated muscle adverse events as they demand distinct management and produce variable prognoses.

Myopathy: muscle weakness +/- pain with elevated CK level

Treatment Algorithm

- Confirmed Statin-Associated Myopathy (CK>10x normal and muscle weakness)
  - Discontinue statin
  - Prednisone
  - Immunosuppressant Medications (Azathioprine, Methotrexate, Rituximab)
  - IVIG