SINAM: A Rare Case Leading to Respiratory Failure

D. Christine Hermiz

Patrick Bradley

Follow this and additional works at: https://scholarlycommons.henryford.com/merf2020caserpt
SINAM: A Rare Case Leading to Respiratory Failure

D. Christine Hermiz, DO; Patrick Bradley, MD
Henry Ford Health System, Detroit, Michigan

Introduction

Statins are commonly prescribed in primary care offices daily; however, they are not exempt from adverse effects. Statin-induced myopathies are best described on a continuum, as patients can range widely in presentation. The most extreme of cases displaying respiratory distress, quadriaparesis, dysphagia, and rhabdomyolysis. Cases on the severe end of the spectrum with respiratory failure are exceedingly rare and should be shared with the medical community in order to gain more insight into the types of statins most at risk for causing complications, most effective treatments based on disease severity, as well as characteristics and objective findings among affected patients in order to diagnose and treat in a time-effective manner.

Case Description

The patient was a healthy, physically active 79-year-old male with past medical history of CVA in 2018 without residual deficits, controlled type 2 diabetes, HTN, and sleep apnea on CPAP presenting in February of 2020 with worsening quadriaparesis, dystarsis, and dysphagia over the past 3 months. Physical exam revealed extreme muscle weakness of 3/5 in upper extremities, 4/5 in lower extremities, unable to raise arms above a 45 degree angle, normal cranial nerve exam. CT of the head/neck was negative, moderate stenosis in carotid US of 50-60%, and MRI of head showing no acute changes. CPK elevated over 4,000 with elevated AST ALT in the 300s. Autoimmune work up was nonrevealing. EMG showed possible myopathy vs motor axonal polyradiculoneuropathy vs demyelinating polyneuropathy. Treatment was conservative and discharged to subacute rehab (SAR).

Over the course of 2 weeks at SAR, his weakness progressed with CPK >7,000 with continued elevated liver function prompting a transfer to Henry Ford for escalation of care in March of 2020. An exhaustive work up was completed with negative autoimmune panel and EMG showing chronic myopathy. History revealed he had been maintained on Azathioprine 20 mg after his CVA in October of 2018 and began to manifest weakness in December 2019 with discontinuation in February 2020. MRI revealed diffuse muscular edema (Figure A). He was initially treated conservatively with intravenous fluids and over the course of 7 days had progressive dyspnea and dysphagia leading to respiratory failure and intubation. Muscle biopsy (Figure B) with anti-HMGCR IgG confirming statin-induced necrotizing myopathy. He was promptly treated with IV dexamethasone for 5 days with transition to 60 mg prednisone daily with IVIG 2g/kg x 5 days. Despite treatment, the patient continued to decline with increasing CPK and inability to liberate from the ventilator. Given his prior wishes was terminally weaned per family request and passed soon after.

Figure A. MRI L. glutus minimus edema. Profound muscular edema displayed as increased signal of the L. glutus minimus (red arrow) characteristic of myositis. A common finding in those with statin induced necrotic myopathy.

Figure B. Muscle necrosis on H&E stain. Areas of patchy white are necrotizing muscle cells (black arrow) surrounded by unaffected areas of normal muscle.

Discussion

The rapid decline of an otherwise healthy patient should prompt further inquiry as to the most efficacious treatment regimens; however, making the diagnosis can be challenging as it is a rare condition with a prevalence in less than 1% of statin-compliant patients. SINAM typically present with progressively worsening symmetrical proximal muscle weakness with CK levels in the thousands despite discontinuation of statins [1, 7]. Other findings include MRI showing muscle edema, atrophy, and/or fatty infiltration, positive anti-HMGCR, EMG positive for myopathy, and muscle biopsy displaying active necrosis [2]. Several case reports show widely varying time courses of 2 months to 10 years after the initiation of statin therapy to symptomatic necrotizing myopathies [2, 7]. Therefore, SINAM should be considered as a differential in the patient with new onset myopathies with past or current use of statins. The type of statins most likely to cause this condition were Atorvastatin, Simvastatin, Lovastatin, and Fluvastatin due to their lipophilic nature and ability to enter muscle cells [2]. Many patients are diagnosed when their most debilitating symptom is proximal muscle weakness; however, retrospective studies have shown that up to one-third of autoimmune myopathy cases had respiratory complications with 5 of these patients requiring intubation [4]. Our patient was unique in that despite the use of aggressive immunosuppression progressed to respiratory failure. To date there are no reported cases of death due to SINAM.

Conclusion

Our patient presented with statin-induced necrotizing autoimmune myopathy that progressed to respiratory failure. SINAM is a rare complication of statin therapy that has high morbidity with rare mortality. SINAM should be on the differential in patients presenting with new onset progressive weakness who have been exposed to statin therapy. The mainstay of treatment is immunosuppressive therapy; however, the optimal regimen and duration is unknown and further studies are required to discern optimal treatment.

Bibliography