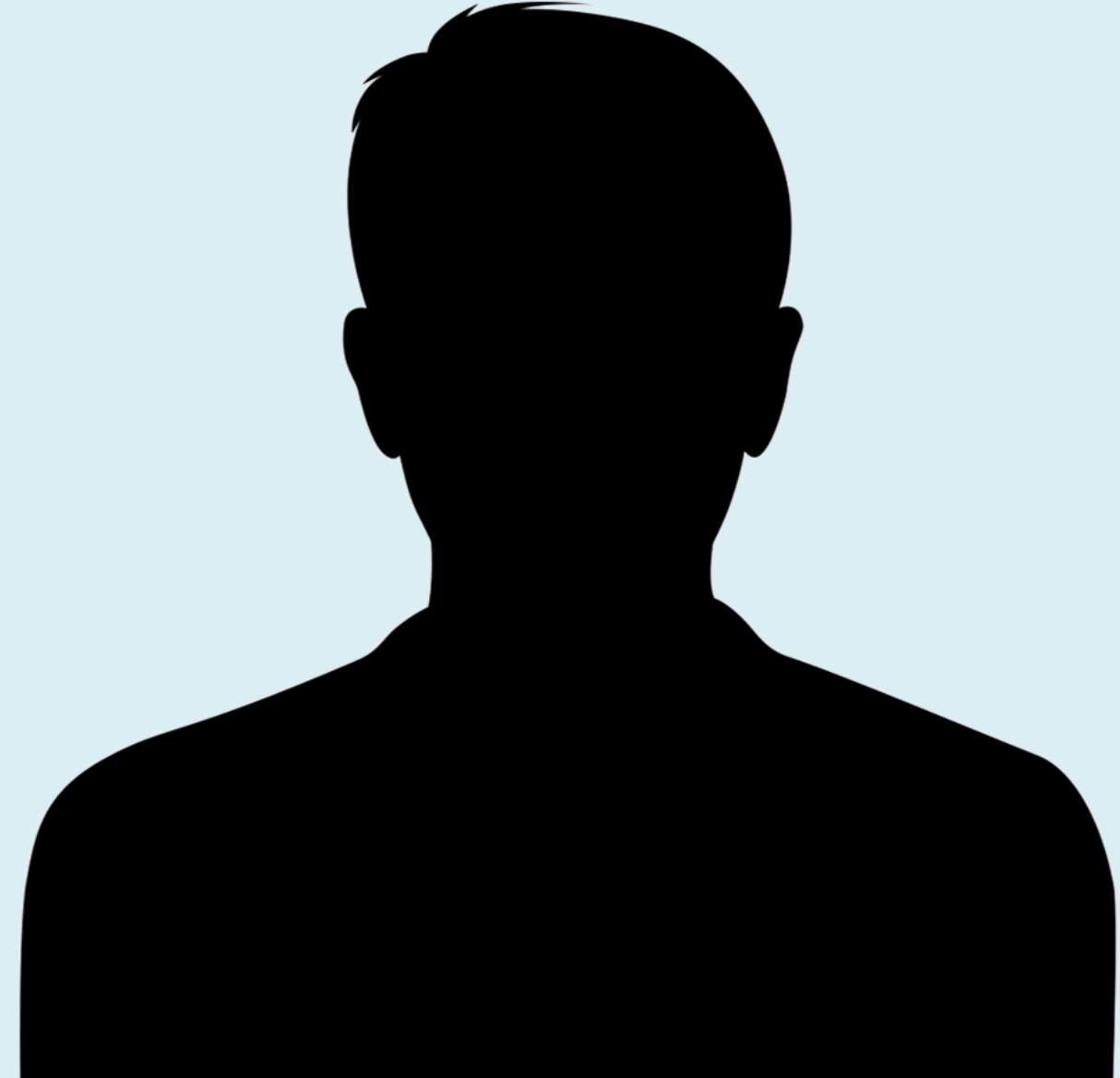


Statin Induced Necrotising Autoimmune Myositis

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Presenting Complaint



An 82 year old male presented to the emergency department after being found on the floor for 36 hours. He had attempted to transfer from his bed and fell, without loss of consciousness or head trauma.

History

Prior to falling, he reported 4 months of gradual weakness affecting his proximal lower limbs and spine, causing kyphosis. He was previously fully mobile and independent. This weakness was preceded by 17 months of atorvastatin use (10mg daily) for primary prevention of ischemic heart disease. The statin was discontinued 4 months prior his fall due to an elevated ALT and muscle weakness. He had atrial fibrillation and was taking aspirin.

Examination

- Bilateral medial quadriceps wasting
- Proximal lower limb weakness
- Power in the upper limbs and ankles were preserved
- Cranial nerves were normal
- Neck extensor weakness with associated head drop
- Normal cardiovascular examination
- Increased respiratory effort
- He was haemodynamically stable and cognitively intact

Investigations

- CK was elevated at 4000 (upper limit 150 UI/L)
- Normal renal function
- Needle electromyogram of the biceps, first dorsal interosseous and vastus medialis showed positive sharp waves and myotonic discharges with spontaneous fibrillations, consistent with myopathy
- Quadriceps muscle biopsy showed evidence of necrotising myopathy, with necrotic fibres, rimmed p62 positive vacuoles and the absence of inflammation
- Autoantibody screen was positive for HMG-CoA reductase, consistent with statin induced necrotising autoimmune myositis (SINAM)
- Screening for other autoimmune myopathies including dermatomyositis and polymyositis was negative

Management

- The patient required intubation and ventilator support in intensive care for type 2 respiratory failure
- He required PEG feeding for severe oropharyngeal dysphagia which was shown on video fluoroscopy
- He was commenced on high dose IV methylprednisolone before switching to 60mg of oral prednisolone
- After 2 months, he was extubated and weaned off tracheostomy
- After 3 months, he was transferred to a tertiary neurology centre and received 2g/kg of IV immunoglobulin (IVIg) in divided doses over 4 days
- After initial improvement in lower leg strength, the patient was commenced on six cycles of cyclophosphamide at 2 weekly intervals, with a tapering steroid course
- After the second cycle he was able to stand independently and was transferred to neurorehabilitation
- After the fourth cycle, he was mobilising independently with a walking stick and discharge home with follow up

Discussion

Cases of SINAM have been reported as early as 2007¹. Patients present with subacute symmetrical proximal limb weakness. Other presentations include mild joint pains, rash and in severe cases, respiratory failure^{2,3}. A similar but fatal case of SINAM was recently reported in which a 71 year old male also required intubation and PEG feeding. However, the patient had minimal response to high dose steroids and subsequently died from sepsis secondary to fungal infection³. Our case is the first to describe effective use of high dose steroids, IVIg and cyclophosphamide in combination for the treatment of near fatal SINAM.

National or International guidelines are not currently available for management of this condition and treatment relies on clinical experience. An algorithm for the diagnosis of SINAM has been suggested². If SINAM is suspected, statins must be discontinued immediately. Once a diagnosis is established, immunosuppressants are initiated as first-line treatment, typically high dose prednisolone (1 mg/kg)². Other immunosuppressants may be used in combination, such as methotrexate, azathioprine or mycophenolate, to optimise response². If severe weakness persists after 8–12 weeks, IVIg or biologics such as rituximab may also be considered².

Morbidity and mortality from SINAM remains uncertain. This case highlights it is important that clinicians are aware of this significant adverse effect from statins, which can develop even after long-term use and discontinuation.

References

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