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Clinical Image

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# Necrotizing Autoimmune Myopathy: A Rare Subset of Inflammatory Myopathy

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## Abstract

A 19-year-old Hispanic male with past medical history of hypothyroidism presented for progressive muscle weakness. This muscle weakness was present throughout the entire body including upper and lower extremities. Upon further evaluation was found to have antibody positive for anti-HMG CoA reductase antibody along with muscle biopsy patient showing active necrotizing myopathy, consistent with diagnosis of necrotizing autoimmune myopathy.

## Keywords

Necrotizing autoimmune myopathy, Muscle weakness, Myopathy, Muscle biopsy

## List of Abbreviations

**NAM:** Necrotizing autoimmune myopathy

## Case

A 19-year-old Hispanic male with history of hypothyroidism was seen in rheumatology clinic with complaint of worsening muscle weakness. Muscle weakness had started two years ago and progressively worsened leading to difficulty with activities of daily living, including getting up from chair or bed without assistance. He denied any skin rash, photosensitivity, cough, dyspnea, joint pain, fevers, night sweats or skin changes suggestive of raynaud phenomenon. Patient reported that he was diagnosed with polymyositis by his previous rheumatologist in Mexico and was treated with high dose corticosteroids without significant improvement. Physical examination showed markedly decreased strength of proximal muscles of upper and lower extremities as well as muscle atrophy.

Initial labs showed creatinine kinase of 13604 units/liter, aldolase 83.5 units/liter, and negative ANA. Treatment was initiated with high dose prednisone and mycophenolate mofetil. Patient did not show any significant improvement over the next 3 months. Due to lack of improvement EMG, myositis antibody panel and muscle biopsy were requested for further evaluation. EMG was consistent with myopathic process and myositis antibody panel showed positive HMG-CoA reductase antibody.

Muscle biopsy showed a moderately active necrotizing myopathy myofiber necrosis, regeneration, and myophagocytosis. There was a focal minimal inflammation present in the endomysium surrounding a degenerating myofiber in a perivascular distribution. Prominent widening of the perimysium as well

as endomysial fibrosis with significant rounded atrophy throughout all fascicles (Figure 1). There was no prior drug exposure to account for the pathology findings.

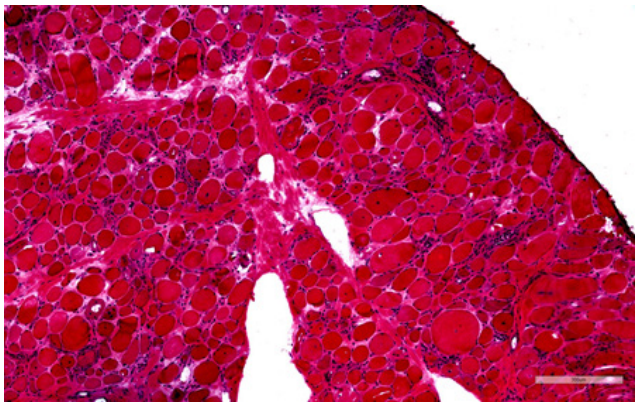

**Figure 1:** Muscle biopsy with findings of moderately active necrotizing myopathy myofiber necrosis, regeneration, and myophagocytosis.

Patient was diagnosed with necrotizing autoimmune myopathy. No paraneoplastic workup was done as patient did not have other signs and symptoms pointing to a paraneoplastic process. Therapy was switched to IVIG and methotrexate with significant improvement in muscle strength within 12 weeks. NAM is a rare form of inflammatory myopathy. Muscle biopsy in NAM typically shows necrotizing myopathy with minimal inflammation [1]. NAM patients present with progressive proximal muscle weakness of upper and lower extremities [2, 3]. Labs usually show markedly elevated creatinine kinase with levels in tens of thousands. Muscle biopsy and myositis antibody panel help establish the diagnosis [3]. Positive anti-HMGCR antibody can be seen in NAM patients even without prior statin exposure [4].

## Disclosure statement

The authors have no conflict of interest.

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