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# Atypical Myositis Presenting with Peripheral Neuropathy

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**Abstract** Necrotizing Autoimmune Myositis (NAM) is an Acute autoimmune disease of the proximal skeletal muscle. The increased rate of morbidity and mortality in NAM is related to the severety and extant of muscle weakness. Patients with NAM develop antibodies to a specific signal recognition particle (anti-SRP ab). Peripheral neuropathy is extremely rare during the course of the disease. We report the case of a 35 y old male, hospitalized for severe proximal and distal muscle weakness with positive anti-SRB ab and concomitant peripheral neuropathy. The outcome was severe respiratory failure and death.

***Keywords:*** *necrotizing myopathy, anti-signal recognition particle (SRP) autoantibody, peripheral neuropathy*

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## 1. Introduction

NAM rarely causes peripheral neuropathy [1], therefore it is rarely considered as diagnosis in the setting of an acute myositis with concomitent peripheral neuropathy. [2]. Typically, patients presenting with NAM develop proximal muscle weakness which occurs suddenly and becomes rapidly progressive and could ultimately be life threatning. These patients also are positive for anti-SRB ab. Most of these cases seem to be resistant to treatment with corticosteroids [3].

This following case report is of an atypical presentation of NAM with positive serology for anti-SRB ab and concomittent peripheral neuropathy. This association causing a more severe outcome.

## 2. Case Report

A 35 y old male presented with a rapid onset of proximal and distal muscle weakeness with subsequent inabilty to stand or walk. He denied any alcohol or drug abuse. He had no past medical history and denied any similar symptoms in the past, he had no recent illness or fever. His family history was negative for muscle disease.

On physical exam, the patient was afebrile and his vital signs were stable. He had no rash or other skin lesions. His examination revealed generalized weakness and muscle pain that was more accentuated in the proximal and distal lower extremities.

Neurological examination revealed symmetrical upper and lower extremity weakness with greater proximal involvement compared to distal muscle groups. Reflexes were present but diminished in the upper extemities and absent in the lower extremities. Both Hoffmann and Babinski reflexes were absent and there were no pyramidal signs. The sensory exam was within normal limits. Cranial nerves were intact and there was no cerebellar ataxia. Cardiovascular as well as pulmonary exam were within normal limits. Both the abdominal and genitu-urinary exams were unremarakble.

Laboratory tests were performed. A CBC showed no leukocytosis. The platelet count was normal. There was no anemia. The CMP revealed elevated liver enzymes: AST 909 IU/L ( Nl 10-40 IU/L) and ALT of 301 IU/L ( Nl 10-55 IU/L). Both the GGT and the Alkaline Phosphatase were within normal limits, 33 IU/L and 40 IU/L respectively. There was a significant elevation of the CPK's at 7003 IU/L as well as the LDH at 15556 IU/L. There was no elevation of the uric acid and the 24 hour total urine protein level was within normal limits. There was a significant elevation of both the ESR (70 mm/h) and CRP ( 20 mg/dL).

Serologies for Lyme disease, Rickettsia, Brucella, Salmonella, TPHA-VDRL, HIV, HTLV1, Hepatitis B, Hepatitis C, CMV and EBV were all obtained and negative. Plasma Glucose, Thyroid function tests, B12 levels and Folic acid levels were also all within normal limits.

Serum Serum ANA were elevated at 1/2560 with elevated anti-SRP levels. Anti-Jo-1, anti-PL-12 were negative.

Arterial blood gases were also abnormal with a non compensated metabolic alkalosis (pH 7.52; PCO2 37 mmHg; pO2 73 mmHg; HCO3 32 Eq/L; O2 Sat 95%).

A lumbar puncture was negative for leukocytes, glucose and proteins as well as for immunogloblulins and oligoclonal bands.

EKG was normal.

Imaging tests, including abdominal ultrasound, a CT with contrast of the brain and abdomen showed no lesions.

The thoracic CT was positive for a Left basal lung collapse and hypoventilation.

EMG showed small amplitude and short duration with low potential fibrillation in the muscles of the lower and upper extremities and the nerve conduction revealed decreasing velocities.

A quadriceps muscle biopsy demonstrated scattered necrotic and regenerating fibers, with endomysial inflammation (Figure 1).

After careful review of the patient's clinical symptoms of sudden and severe proximal muscle weakness combined with the serological findings of elevated CPK and positive anti-SRP as well as necrotizing fibers on the muscle biopsy the diagnosis of inflammatory myositis with peripheral neuropathy was made, more specifically NAM [2].

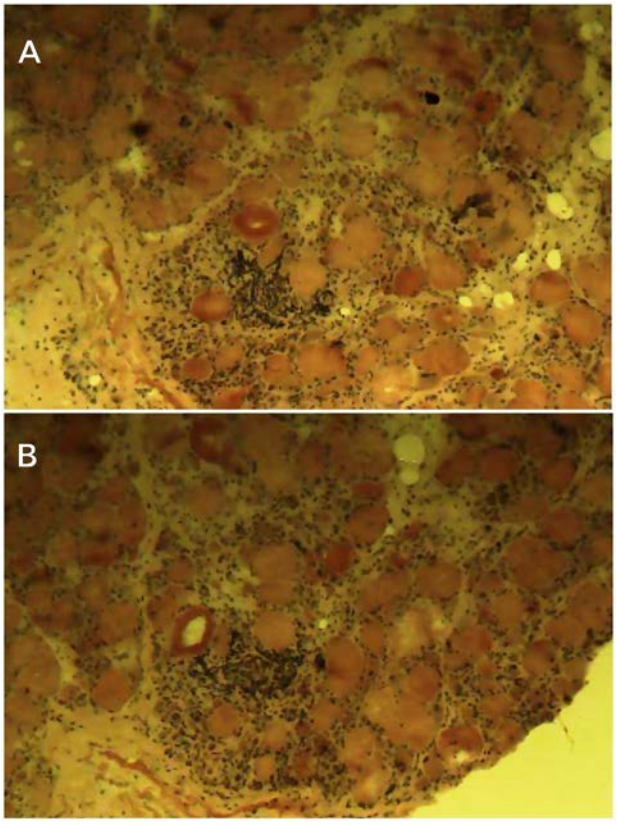


**Figure 1.** Panel A & B: Biopsy of the femoral muscle showing necrotic and regenerating fibers with endomysial inflammation.

The patient was started on Methylprednisolone at the dose of 1gm/ QD for 3 days combined with IV Immunoglobulin at the dose of 2g/kg and careful monitoring of his CPK levels was initiated.

Unfortunately the patient didn't regain any of his muscle strength and suffered swallowing difficulties and well as respiratory failure ultimately leading to his demise.

## 3. Discussion

Peripheral neuropathy rarely occurs during the course of an inflammatory myopathy [1]. In fact, it's presence usually leads to the exclusion of the diagnosis of myositis [2]. The frequency of neurological involvement in myositis is unknown. There have been a few isolated cases reported in the medical literature notably an article published in 1996 which reviewed six known cases of NAM that had been already documented and introduced four new cases of NAM associated with peripheral neuropathy [1].

In 2003, Seinturier and al. did a retrospective study of 31 patients with inflammatory myositis and searched for any data of neurological involvement. They were able to identify 3 patients with neuro-myositis, suggesting that this feature could be underestimated [4].

Some authors remain skeptical and consider these cases of neuro-myositis to be a form of vasculitis or other sub-type of inflammatory disease that can be associated with symptoms of peripheral neuropathy [5]. Others have embraced the diagnosis and even proposed criteria for the diagnosis of neuro-myositis which combine the typical inflammatory myositis criteria and the presence of neurological impairment which can't be explained by any other underlying cause [1].

The patient in our case report met all the criteria for acute myositis and peripheral neuropathy, including sudden loss of tendon reflexes, absence of muscle atrophy, minimal myalgias, acute distal muscle weakness and decreased nerve conduction velocities. Causes of peripheral neuropathy were also ruled out in this case, mainly Guillan-Barre syndrome. Last but not least, this patient was positive for anti-SRP ab. which are only present in 4 to 6% of patients with confirmed myositis [6]. This association of elevated anti-SRP [7], elevated CPK, rapidly progressing proximal muscle weakness and peripheral neuropathy confirmed the diagnosis of NAM.

Most patients with NAM are resistant to corticosteroids [8,9] although a few seem to benefit from IV immunoglobulin [10,11]. Unfortunately our patient's clinical presentation worsened rapidly and he died of respiratory failure.

## 4. Conclusion

The presence of peripheral neuropathy in the presentation of inflammatory myopathies must be considered despite it's rare presentation. As reported in other cases of necrotizing myositis, the presence of anti-SRP antibodies is a predictor of severe and negative outcome for most patients.

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