Seronegative Immune Mediated Necrotizing Myopathy Presenting with Rapidly Progressive Muscle Weakness and Acute Kidney Injury

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Introduction

- Immune mediated necrotizing myopathy (IMNM) belongs to a group of rare inflammatory myopathies characterized by significant necrosis and minimal lymphocytic infiltrates. IMNM has been associated with anti-signal recognition particle (SRP) and anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (HMGCR).
- Two-thirds of patients with immune-mediated necrotizing myopathy have an identifiable myositis-specific antibody, of which anti-SRP and anti-HMGCR are the most common.
- A small subset of patients will have seronegative IMNM.

Case description

A 78-year-old woman with a presented to the emergency department with bilateral lower extremity weakness for the last three days. She reported sudden inability to ambulate after sustaining a fall from ground level.

Past medical history: Hypertension, hyperlipidemia on statin therapy, prior DVTs and PAD.

Physical examination: Her physical exam revealed decreased muscle strength in the lower limbs of 0/5 proximally, 4/5 distally and 2/5 strength in proximal upper limbs and 5/5 distally. Sensation was normal bilaterally.

Significant laboratory results: Creatinine of 5 (normal 0.6-1.1) from normal baseline and creatinine kinase level (CK) of 20,000 (normal 29-168). CK continued to increase despite IV fluids.

Myositis panel: Extended myositis panel was negative for myositis specific antibodies seen in IMNM including HMGCR and SRP.

Electromyogram: Myopathic pattern of injury which was likely inflammatory.

MRI lower extremity: A magnetic resonance imaging (MRI) of the lower extremities demonstrated edema to thigh muscles bilaterally. See figure 2

Muscle biopsy: Extensive necrosis with few inflammatory cells consistent with a diagnosis of IMNM. See figure 3

Figures

Figure 1. Graph showing sharp decline in CK levels after initiation of pulse dose steroids

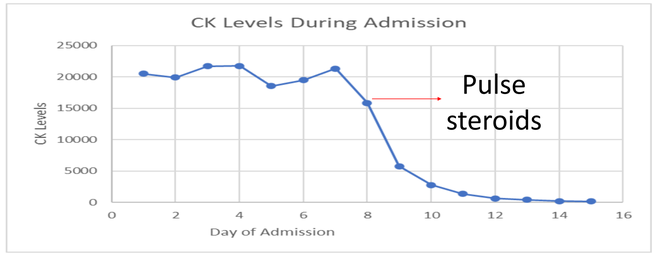
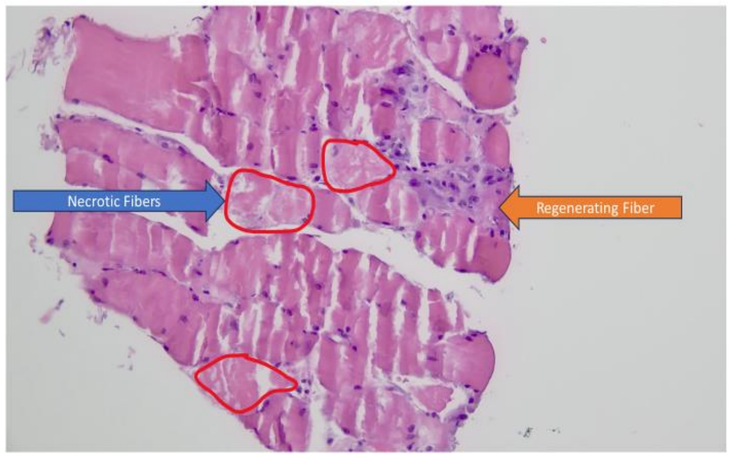


Figure 2. T2 weighted MRI images of the thigh muscles showing hyperintense signals in keeping with intra and intermuscular and subcutaneous edema

Figure 3. Left quadriceps muscle biopsy showing necrotic and regenerating fibers



Outcome

- She received pulse dose steroids for 5 days which resulted in a rapid decrease in her CK levels.
- Treatment was continued with three days of IVIG followed by rehabilitation.
- She has since had improvement in muscle weakness and is able to ambulate.
- Unfortunately, her renal function did not improve, and she remains on hemodialysis.

Discussion

- Inflammatory myopathies can present a diagnostic challenge.
- The atypical presentation of rapidly progressive sudden onset muscle weakness and negative myositis antibodies resulted in a delay in definitive diagnosis and treatment.
- It is paramount to have a high index of suspicion for IMNM in patients with muscle weakness and elevated CK levels even in the absence of positive myositis as early treatment has been shown to have favorable outcomes.

References

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