Fatal Respiratory Failure from Polymyositis

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INTRODUCTION

•Polymyositis is a fairly uncommon inflammatory myopathy that presents with symmetrical proximal muscle weakness.

•This case is presented to emphasize that polymyositis can also involve respiratory muscles and acutely result in fatal outcomes.

CASE DESCRIPTION

Initial Presentation:

A 43-year-old male with no significant past medical history presented to the hospital with progressively worsening upper and lower extremity weakness for three weeks.

•Physical examination was notable for severe tenderness in all his extremities.

•Strength in bilateral hip flexors and shoulder abductors and adductors preserved at 5/5.

Pertinent Lab Values	
CK	14,803
Aldolase	139.5
C3	207 (Reference range 82-185)

•An MRI of his thighs showed diffuse myositis (Figure 1).

•Muscle biopsy was performed, and prednisone was started with rapid improvement in his pain and CK level. He was discharged on steroids; on outpatient follow up, he was started on azathioprine as his muscle biopsy was consistent with polymyositis.

•Further extensive rheumatological work up was otherwise unremarkable. On his own, he stopped taking azathioprine due to eventual improvement in his symptoms.

Hospital Readmission:

•Patient was subsequently readmitted to the hospital within one year of initial hospitalization with similar symptoms.

•His CK level was elevated to about 2000, and he was started on prednisone.

•However, a new diffuse reticular rash developed within 12 hours of admission along with worsening myalgias and further elevation of his CK to about 12,000 (Figure 2).

FIGURES

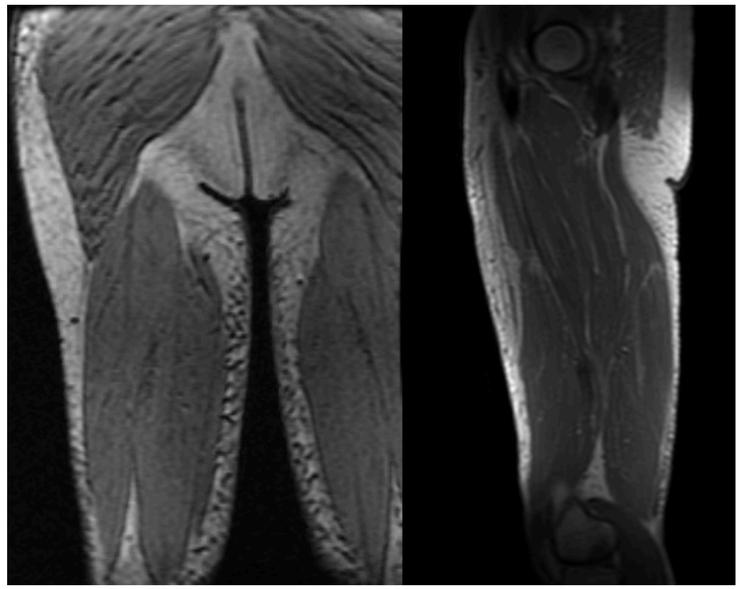

Figure 1. MRI with contrast of right mid-thigh displaying mild diffuse patchy and streaky edema in almost all muscles of the thigh.

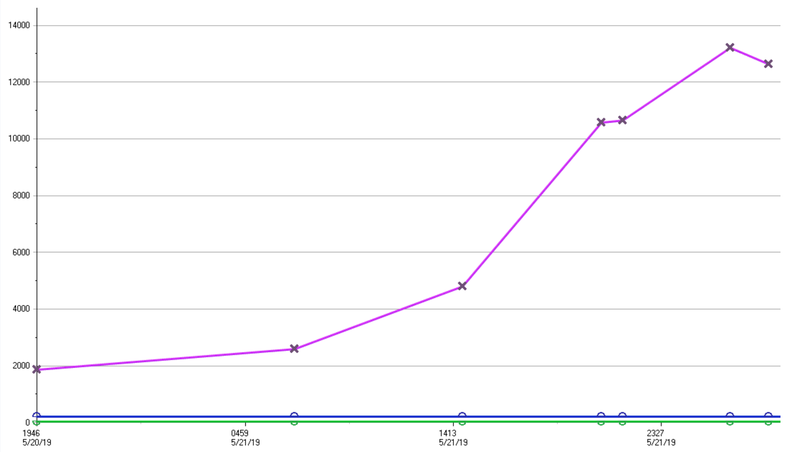

Figure 2. Graph displaying trend of marked rise in CK levels upon readmission to hospital one year following initial presentation.

CASE DESCRIPTION

- Subsequently, the patient then started to develop respiratory distress and was significantly tachypneic. He unfortunately experienced a PEA cardiac arrest with successful return of spontaneous circulation after 20 minutes of CPR.
- The ensuing ICU course was complicated by development of cardiogenic shock and two additional episodes of PEA arrest, and he unfortunately passed away.
- An autopsy revealed evidence of inflammatory interstitial infiltrates in skeletal muscles, and the cause of death was deemed to be secondary to complications of polymyositis.

DISCUSSION

- Polymyositis is a connective tissue disease characterized by inflammation of the muscles.
- Most commonly, this disease is characterized by symmetrical proximal muscle involvements.
- However, sometimes, extra-muscular manifestations affect other organ systems and can manifest with symptoms of dysphagia and rash.
- The diagnosis of this disease is established primarily by electromyography and muscle biopsy.
- Respiratory muscle involvement is very rare in comparison to limb muscle disease and usually occurs later in the disease course.
- However, there have been rare cases reporting fatal respiratory failure from early involvement of respiratory muscles. Pulmonary involvement leads to worse outcomes and increased mortality.
- This case highlights that clinicians should be aware that polymyositis can also involve respiratory muscles and result in catastrophic outcomes due to possible respiratory failure. Closer monitoring of respiratory status may be necessary in patients with rapidly worsening respiratory symptoms despite treatment.