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Urosepsis-triggered acute polymyositis with Inflammatory Subcutaneous Edema-a rare combo

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Abstract
Polymyositis usually has a gradual progression, no cutaneous involvement, and sometimes a viral infection as the trigger. Here, we present a case of severe Polymyositis presenting acutely with inflammatory subcutaneous edema triggered by bacterial sepsis. Polymyositis is a chronic autoimmune inflammatory myopathy typically characterized by symmetrical proximal muscle weakness that progresses over time. A 28-year-old woman came in with a history of low-grade fever and proximal weakness in all four limbs for one month. In emergency investigations, neutrophilic leucocytosis, deranged renal function tests, elevated serum creatine phosphokinase levels (7387 U/L), elevated serum lactate dehydrogenase levels (3378 U/L), and a full field of pus cells with positive nitrite test on urinalysis were revealed. Ultrasound of the soft-tissue neck and arms showed inflammation in the subcutaneous plane/ inflammatory subcutaneous edema. Empirical antibiotics (IV ceftriaxone) gradually lead to the resolution of fever, leucocytosis, sepsis, and serum creatinine. Further testing revealed a negative Antinuclear Antibody, Rheumatoid factor, Antineutrophil Cytoplasmic Antibodies, and Extractable Nuclear Antigen Antibodies (ENA) Panel. Muscle atrophy, muscle fiber necrosis, and endomysial lymphocytic infiltration were seen in the muscle biopsy, indicating Polymyositis. The patient began to recover symptomatically, with oropharyngeal dysphagia, muscular weakness, and subcutaneous edema resolving quickly within a week after starting steroids. A gradual fall in serum creatine phosphokinase levels was also noted. During the outpatient follow-up, prednisolone was decreased to a low dose, and oral Azathioprine was begun as a steroid-sparing medication. Prednisolone was discontinued at subsequent outpatient visits, and the patient continued to be asymptomatic on oral Azathioprine.

Keywords: polymyositis, sepsis, subcutaneous edema

Introduction
Polymyositis is a chronic autoimmune inflammatory myopathy typically characterized by symmetrical proximal muscle weakness that progresses over time. The overall incidence is 1 in 100,000, with a female to male ratio of 2:1 affecting persons over 20 [1]. Coxsackie B virus, parvovirus, enterovirus, human T-cell lymphotropic virus (HTLV-1), and human immunodeficiency virus have all been suggested to set off myositis [2]. Inflammatory myopathy patients had a higher frequency of positive antibodies to Coxsackie B virus and HTLV-1 than healthy controls [3]. Immune dysregulation and a greater infection risk from the underlying inflammatory process and immunosuppression might explain the increased frequency of viral antibodies in myositis. Skin involvement is typically absent in Polymyositis. However, there are very few cases reports which showed subcutaneous edema in patients with Polymyositis [4]. Here we report a case of Polymyositis with an unusual presentation.

Case Report
A 28-year-old woman came in with a history of low-grade fever and proximal weakness in all four limbs for one month. She had difficulties walking, combing her hair, and sipping a glass of water at first, but these abilities rapidly deteriorated, and she became bedridden. She also developed swelling in her bilateral upper limbs, neck, and upper back at the same time. For the past five days, she reported difficulties swallowing and coughing while eating and noticed reduced urine output for the past two days. She denied any long-term drug use or addiction. There is no history of a similar illness in the family.

On examination, the patient was awake, alert and oriented, and vitally stable. Non-pitting soft tissue swelling in bilateral upper limbs, neck and upper back was noted with moderate erythema and tenderness. The central nervous system evaluation indicated decreased tone in all four limbs, with proximal muscles in both the upper and lower limbs having a power of 2/5 and distal muscles in both the upper and lower limbs having a power of 3/5. Plantar reflex showed bilateral flexor response, and all deep tendon responses were absent. Higher mental functions were normal, and there was no evidence of cranial nerve, sensory, or cerebellar involvement. The rest of the systemic examination was unremarkable.

In emergency investigations, neutrophilic leucocytosis, deranged renal function tests, elevated serum creatine phosphokinase levels (7387 U/L), elevated serum lactate dehydrogenase levels (3378 U/L), and a full field of pus cells with positive nitrite test on urinalysis were revealed. Ultrasound of the soft-tissue neck and arms showed inflammation in the subcutaneous plane/ inflammatory subcutaneous edema. Cerebrospinal fluid analysis and a Magnetic Resonance Imaging of the spine were normal. Further testing revealed a negative Antinuclear Antibody, Rheumatoid factor, Antineutrophil Cytoplasmic Antibodies, and Extractable Nuclear Antigen Antibodies (ENA) Panel. Thyroid profile and serum electrolytes were also normal.

Empirical antibiotics (IV ceftriaxone) gradually lead to the resolution of fever, leucocytosis, sepsis, and serum creatinine. Urine culture grew Escherichia Coli which was sensitive to ceftriaxone, and blood culture was sterile. Serologies for possible infective etiologies of myositis, such as Dengue fever, Chikungunya, Lymes disease, Rickettsiae, HIV, were all negative. As per the initial impression of acute infectious myositis or autoimmune myopathy, a muscle biopsy was performed. The patient was put on steroids (Pulse high-dose methylprednisolone followed by oral prednisolone) after fever and leukocytosis resolved. The patient began to recover symptomatically, with oropharyngeal dysphagia, muscular weakness, and subcutaneous edema resolving quickly within a week after starting steroids. A gradual fall in serum creatine phosphokinase levels was also noted. Muscle atrophy, muscle fiber necrosis, and endomysial lymphocytic infiltration were seen in the muscle biopsy, indicating Polymyositis.

On discharge, the patient had 4+/5 strength in all four limbs, could walk without assistance, and dysphagia and subcutaneous edema were entirely resolved. During the outpatient follow-up, prednisolone was decreased to a low dose, and oral Azathioprine was begun as a steroid-sparing medication. Prednisolone was discontinued at subse quent outpatient visits, and the patient continued to be asymptomatic on oral Azathioprine.

Discussion

Polymyositis is an idiopathic inflammatory myopathy typically characterized by insidious onset and gradual progression of weakness, with an uncommon acute presentation. Polymyositis patients often have symmetrical proximal muscular weakness in both the upper and lower limbs. Neck flexors weakness is also common. Polymyositis patients may have muscular soreness and tenderness, which can be mistaken for Polymyalgia Rheumatica symptoms. The condition can last for months before the patient seeks medical help, and it generally affects all of the muscles in the legs, trunk, shoulders, hips, and upper arms [5]. The patient in our case had an acute onset, rapidly progressing weakness that resulted in quadriparesis and dysphagia secondary to bulbar muscle involvement.

Cutaneous involvement is classically not seen in Polymyositis. Polymyositis does not have a rash like dermatomyositis. However, "mechanic's hands," or hyperkeratotic eruptions across the finger pads and lateral parts of the fingers, can be found. Antisynthetase antibodies have been linked to the Raynaud phenomenon. Periorbital edema is a rare complication. Calcinosis is a condition that affects around 5% of Polymyositis patients. Telangiectasias are a rare occurrence. However, in our case, the patient has inflammatory subcutaneous edema, which is an extremely rare presentation of Polymyositis. Only a few cases of Polymyositis presenting with inflammatory subcutaneous edema have been reported [6].

Myositis has been linked to the human leukocyte antigen (HLA) haplotypes A1, B8, and DR3, which are also associated with an elevated risk of autoimmune disorders. Viruses such as Coxsackievirus B1, HIV, HTLV-1, Hepatitis B virus, Influenza, Echovirus, and Adenovirus have all been reported to trigger Polymyositis [5]. However, when it comes to bacterial infections, this phenomenon has not been well documented. In our case, Urosepsis secondary to Escherichia Coli precipitated acute and severe Polymyositis in a previously undiagnosed patient.

Conclusion: Polymyositis is a rare autoimmune disorder. Polymyositis can present acutely with inflammatory subcutaneous edema and can be triggered by bacterial sepsis, despite its rarity as a trigger.

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