Dermatomyositis in an SCI Patient Complicated by Hemorrhagic Myositis: A Case Report

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INTRODUCTION

Dermatomyositis (DM) is an idiopathic inflammatory myopathy. Diagnosis is based on the presence of characteristic skin changes, muscle weakness, elevated serum muscle enzymes, abnormal EMG, and muscle biopsy [1]. Treatment with corticosteroids and immunotherapy can be curative, but in most patients DM is persistent or cyclical and can lead to severe disability [2]. Studies have shown that early exercise in DM can improve functional outcomes [3]. In a patient with a pre-existing spinal cord injury, development of this disease can be profoundly debilitating. We will present a case of dermatomyositis in a paraplegic patient complicated by acute spontaneous intramuscular hemorrhage.

CASE DESCRIPTION

A 59-year-old male with chronic paraplegia presented with a 4-week history of bilateral upper extremity (BUE) weakness and diffuse pruritic rash that made him unable to perform ADLs. He had previously been functionally independent. Physical exam was remarkable for decreased muscle strength in BUE's and diffuse maculopapular rash on the upper chest, face, and extremities (see Image 1). Labs were notable for mildly elevated inflammatory markers and CPK (see Table 1). MRI of the BUE's was concerning for diffuse inflammatory myositis (see Image 2). Skin and muscle biopsies were consistent with dermatomyositis. Malignancy workup was grossly negative.

The patient was evaluated by Rheumatology and started on high-dose steroids and immunomodulatory therapy. Following initiation of treatment, his rash resolved and muscle strength returned to near-baseline. With these improvements he was able to begin physical and occupational therapy. A few days after staring therapy he developed left forearm pain and swelling. Imaging revealed a hematoma in his forearm musculature (see Image 3). The patient had been on prophylactic low-molecular weight heparin (LMWH) while inpatient which was then discontinued. The hematoma was treated conservatively while PT and OT regimens were modified. Although progress was delayed, he is improving and is expected to return to his baseline level of independence.

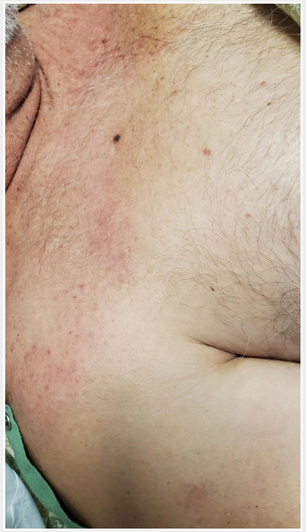

IMAGE 1: Erythematous rash seen on patient's face, chest, and upper arms

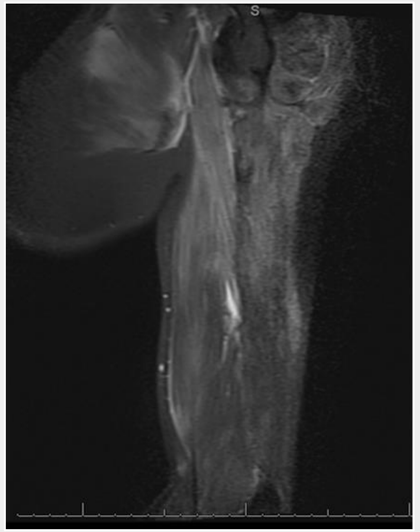

IMAGE 2: MRI of left upper extremity on admission showing diffuse myositis

LAB TEST	ADMISSION	2 WEEKS	REFERENCE
WBC	4.39 x 10^9/L	9.5 x 10^9/L	3.9-9.9 x 10^9/L
Hgb	13.9 g/dL	14.9 g/dL	13-17 g/dL
Platelets	**140 x 10^9/L**	177 x 10^9/L	167-378 x 10^9/L
ESR	**24 mm/hr**	17 mm/hr	<1-20 mm/hr
CRP	**1.5 mg/dL**	0.7 mg/dL	0.0-1.0 mg/dL
CPK	**393 U/L**	50 U/L	51-296 U/L
LDH	193 U/L	--	133-226 U/L
Myoglobin	**150 ng/dL**	--	24-121 ng/mL

TABLE 1: Patient's labs on admission and two weeks after initiation of treatment

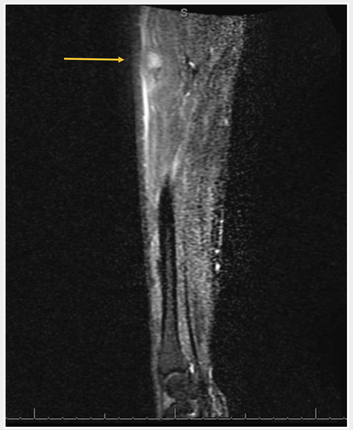

IMAGE 3: MRI of left arm showing brachioradialis intramuscular hematoma (yellow arrow)

DISCUSSION

This is the first case, to our knowledge, of a diagnosis of dermatomyositis in a paraplegic patient. The differential diagnosis on presentation was broad, including myelopathy, neuromuscular junction disorder, autoimmune disease, and infection. Along with Rheumatology, Neurology was consulted as well and a thorough workup with labs and imaging was completed before the final diagnosis was made. Muscle enzymes were less elevated than expected for DM likely due to the decreased muscle mass in his lower extremities. With regards to the hematoma, it is unclear whether therapy caused or accelerated its development. There are multiple case reports that detail the risk of hemorrhagic myositis with the use of LMWH in DM patients [4,5]; however, as far as we are aware, these cases were not in the rehab setting. Mild cases, such as this one, can delay rehabilitation. More research must be done in order to establish guidelines for DM management and therapy in a SCI rehab patient.

CONCLUSIONS

Early interdisciplinary management between PM&R and Rheumatology was essential to establish the diagnosis and begin treatment. While physical and occupational therapy are vital in maintaining and improving functionality and strength gained from pharmacological treatment, it is unclear whether premature initiation of therapy can trigger development of hemorrhagic myositis in a DM patient while on LMWH.

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