A CASE REPORT: DERMATOMYOSITIS IN YOUNG MALE

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Abstract

A rare condition called dermatomyositis results in skin rashes and muscle inflammation. These are a collection of illnesses affecting the muscles that result in swelling and inflammation.It's different from other muscle diseases because it also causes skin problems. Dermatoyositis is the term used to describe involvement of both muscle and skin symptoms.

Mostly seen in the age group of 50 to 70 years but can present at any age. The disease is twice as likely to strike women as it does men. Some people with the disease also have a connective tissue disorder, such as lupus or rheumatoid arthritis. Diagnosis at time and administration of steroid holds better prognosis of the patient.

Keywords: dermatomyositis, myopathy, creatinine phosphokinase

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Introduction

Dermatomyositis is the idiopathic inflammatory myopathy along with skin manifestations. The disease has more female preponderance. There are juvenile and adult versions of it. Women are more likely than males to be affected by dermatomyositis, which has an incidence of 1 in 100,000 cases.[1, 2] Skin rashes, gradual muscular weakening, increased blood muscle enzymes, abnormal electromyogram results, and abnormal muscle biopsy findings are the basis for the diagnosis of dermatomyositis.[3]

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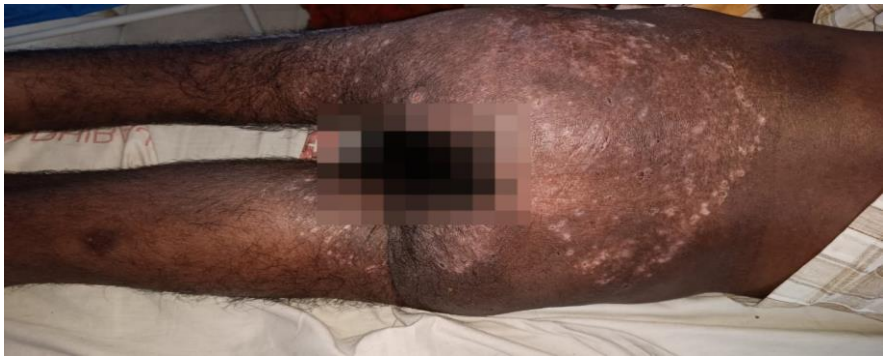
Case Report

A 38-year-old male patient, occasional drinker living in Khargone, Madhya Pradesh, India, known case of Polio since childhood. Atrophy of Left lower limb muscles involved since then. Presented with complaints of bilateral Upper limb weakness (Right > Left) for 15-20 days followed by bilateral Lower limb weakness (right > left) followed by truncal and neck muscle weakness with bilateral lower limb swelling since 1 year with blackish discoloration of skin over face, over buttocks and back, associated with itching and lips swelling. Also associated with difficulty in swallowing of food particle (solid>liquid)since15 days. There was no history of fever, dyspnoea and use of any medication. On general physical examination patient was afebrile, pulse was 88 per minute, Blood pressure was 116/70mmHg. Other systemic examination was normal. Neurological examination revealed normal higher mental functions and cranial nerves. Pupils were bilateral reactive to light and plantars were flexors bilaterally. Tone was reduced in left upper and lower limb and markedly reduced in right upper and lower limb.

REFLEXES	AJ	KJ	BJ	TJ	SJ
RIGHT LIMB	+1	+1	+2	+1	+2
LEFT LIMB	+1	+1	+2	+1	+2

POWER	UPPER LIMB	LOWER LIMB
RIGHT	2/5	2/5
LEFT	3/5	3/5

The sensory system was within normal limits. The rest of neurological examination was normal. In respiratory system, chest was clear and bilateral air entry was present. In cardiovascular system, S1 and S2 sound was noted. The murmur was absent. Investigation revealed Hb- 11gm%, WBC- 5500/cumm, PLATELETS- 3.13 lakh/cumm, CRP- 6.95, UREA- 26mg% (reference value- 11-45mg%), CREATININE-0.5mg%, SGPT- 24IU/L (reference value- up to 40IU/L), SGOT- 68(reference value- up to 40IU/L). Urine analysis and blood sugars were normal. Patient is Euthyroid. Creatinine phosphokinase- 2765 IU/L (reference value: 24-190 IU/l) and ANA was positive. Chest Xray, ECG, ultrasound abdomen and pelvis were normal.



NEW TEMPL MUSCLE BIOPSY(FRESH)- ROUTINE WITH ENZYME HISTOCHEMISTRY
(EHC) - Date: 01/07/2023 04:38 PM

Name / Age / Sex /Lab No	**Mr. MAHESH VERMA**	38/M	X-3344/2023
Biopsy (Fixed)	**EHC (N)**	**Tissue preserved (N)**	**Unstained (N)**
Clinical diagnosis: Dermatomyositis		**Biopsy site: NA**	
Gross details	Received muscle bit measuring 0.4cm in diameter each. All processed-A1. Grossed By : Dr. Rima S, Dt. 28/06/2023		
PARAFFIN SECTIONS			
Fixation	Optimal		
H & E	Section show muscle biopsy with preserved fascicular architecture. There is significant variation in fibre size. There is prominent perifascicular atrophy. Myonuclear clumps are noted. Regenerating fibres and myophagocytosis are seen in the perifascicular zone. There is increased vascularity and interstitial inflammation in the perifascicular zone with lympho-histiocytic infiltrates around the blood vessels. There are no rimmed vacuoles or fibrosis		
MAT	There is no increase in endomysial connective tissue		

Impression:

Dermatomyositis, muscle biopsy

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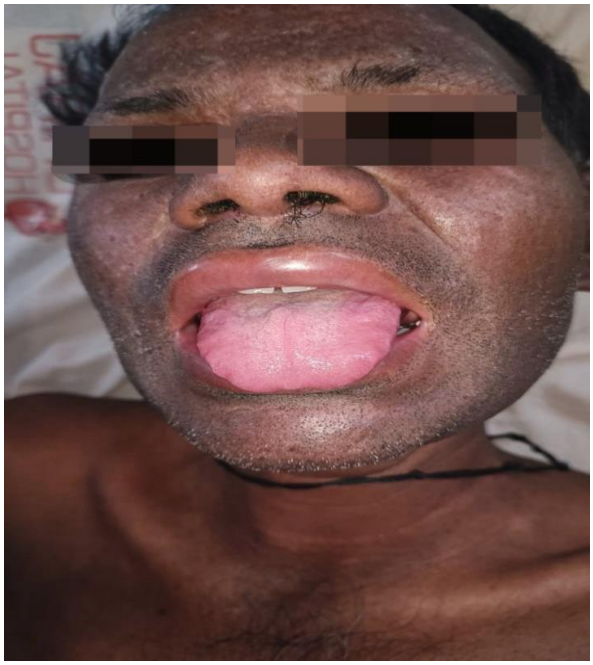
END OF REPORT



Discussion

The erythematous rash on the face, neck, and anterior chest (V neck sign), or the back and shoulders (shawl sign), on the knees, elbows, and malleoli, is one of the typical cutaneous observations. The rash might aggravate after sun exposure and become itchy in specific circumstances. Blue-purple discoloration of the upper eyelids known as heliotrope rash, which is often associated with edema. Elevated violaceous rashes or papules near the knuckles, particularly in the interphalangeal and metacarpophalangeal joints, are the characteristics of Gottron rash.

The patient showed typical features for diagnosis of dermatomyositis such as progressive muscle weakness, elevated muscle enzyme, dysphagia, and erythematous rashes all over the body with positive anti-Mi-2 antibody, specific to dermatomyositis.

It is determined that this patient has classic dermatomyositis.

High-dose prednisone, azathioprine, mycophenolate mofetil, or methotrexate for steroid sparing effect are treatment modalities, IVIg. After using prednisolone 60 mg once daily, our patient's muscle power was not improved. Newer biological agent Rituximab (anti CD20 antibody) was used and patient improved after that. With a better prognosis and a more favourable response to treatment, dermatomyositis can fully recover functionally and is frequently maintained with maintenance therapy. Relapses might happen at any moment. Patients who have advanced stage of disease upon presentation, those whose initial therapy is delayed, and those who have significant dysphagia or breathing difficulties have a worse prognosis.

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