Alemu and Ambaye
Journal of Medical Case Reports (2025) 19:478
https://doi.org/10.1186/s13256-025-05446-1

Journal of
Medical Case Reports

CASE REPORT

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Diagnostic challenges of dermatomyositis in a resource-limited setting: a case report

Getachew Sahile Alemu[1*] and Mekdes Tarekegn Ambaye[1]

Abstract

Background Dermatomyositis is a rare idiopathic inflammatory myopathy characterized by distinct cutaneous manifestations and progressive proximal muscle weakness. However, in areas with limited diagnostic tools without a high index of suspicion, early identification is often challenging, increasing the risk of complications.

Case presentation We report the case of a 50-year-old Ethiopian woman who presented with progressive difficulty swallowing, proximal muscle weakness, and characteristic dermatologic manifestations. Despite multiple emergency visits and extensive workups, such as chest computed tomography and barium swallow, the diagnosis was delayed. Key findings included elevated muscle enzymes and low serum albumin, and a muscle biopsy revealed muscle fiber atrophy. The diagnosis of dermatomyositis was confirmed by the American College of Rheumatology–European League Against Rheumatism classification criteria.

Management We treated the patient with high-dose prednisolone and methotrexate as a steroid-sparing agent, along with nutritional support. Her muscle strength and dysphagia significantly improved within weeks of therapy initiation.

Conclusion Our goal is to emphasize the importance of early recognition of dermatomyositis, especially in resource-limited settings, to avoid unnecessary, financially debilitating workups and prevent complications such as malnutrition, which are completely avoidable with early detection.

Keywords Dermatomyositis, Inflammatory myopathy, ACR–EULAR classification criteria

Introduction

Dermatomyositis (DM) is a type of idiopathic inflammatory myopathy (IIM) characterized by distinctive rashes such as heliotrope rash, facial erythema, V-sign, and Gottron's papules over the knuckles, along with inflammation of the musculature, symmetrical and progressive proximal muscle weakness, and elevated serum levels of muscle-associated enzymes. Although DM is considered a rare autoimmune disease with an incidence of 9.63 per 1 million individuals, its associated complications can be detrimental to the patient's well-being [1].

*Correspondence:
Getachew Sahile Alemu
sahlegetachew30@gmail.com
[1] Menelik II Comprehensive Specialized Hospital, Addis Ababa, Ethiopia

Case presentation

A 50-year-old Ethiopian woman presented to the emergency department with a 3-month history of progressive dysphagia. She initially experienced gradual difficulty swallowing both solid and liquid diets, with more difficulty for solid diets.

At 5 months before dysphagia onset, she noticed bilateral skin discoloration and hardening of her hands. This discoloration gradually appeared in her periorbital area, neck, and knees. At 3 months into the illness, she experienced significant muscle weakness, including difficulty with standing up from a seated position, lifting objects, and holding up her head. Over the following month, her weakness worsened, eventually leading to difficulty moving all of her extremities, resulting in her becoming bedridden. During this period, she also

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Fig. 1 Erythematous violaceous eruptions over both upper eyelids (heliotrope rash)



Fig. 2 Macular, hyperpigmented, and several depigmented skin eruptions over the back of the neck, upper back, and shoulders (shawl sign)

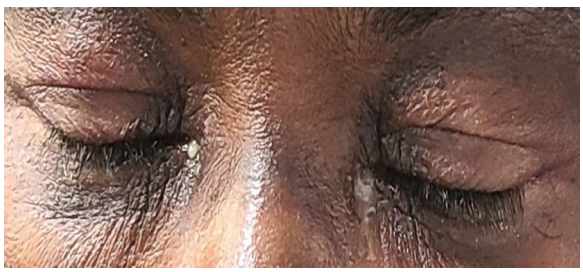


Fig. 3 Macular, depigmented, and hyperpigmented skin eruptions over both knees (Gottron's sign)



Fig. 4 Cracked and rough hands and digits (mechanic's hand)

reported fatigue, malaise, muscle pain, and bilateral leg swelling, which gradually progressed to facial swelling and unquantified significant weight loss. In addition, she had a 1-week history of reddish urine color discoloration.

There was no history of coughing or sputum production, previous joint pain or swelling, dyspnea, orthopnea, or palpitations. She also had no history of visual disturbances, diabetes mellitus, confirmed malignancy, exposure to toxic drugs or chemicals, or a family history of similar illness.

During the physical examination, she appeared acutely ill on a chronic basis. Her vital signs were as follows: blood pressure (BP) 120/80 mmHg, pulse rate (PR) 80 beats per minute, respiratory rate (RR) 22 breaths per minute, temperature (T°) 36.4 °C, and oxygen saturation (SaO_2) 94% while breathing room air. On the head, eyes, ears, nose, and throat (HEENT), she had erythematous violaceous eruptions over both upper eyelids (heliotrope rash) see Fig. 1. She has pink conjunctivae and nonicteric sclera. She had swollen lips. Lymphoglandular, respiratory, and cardiovascular system examinations were normal.

On locomotor examination, she had swollen and tender muscles over her biceps, triceps, brachioradialis, quadriceps, and calf muscles. Furthermore, she had grade 2 pitting edema bilaterally. On integumentary examination, there were scaly, depigmented skin lesions over the knuckles (Gottron's sign) and V-shaped, distributed, depigmented skin lesions over the anterior neck and chest (V-sign) (see Figs. 2 and 3) as well as on her back and shoulders. Her hands and digits showed roughening and cracking, resembling "manual laborer" hands (mechanic's hands) (see Fig. 4). Typical erythematous lesions were not noticed in this patient, which might have been due to her darker skin color.

Upon neurological examination, she was alert and oriented to time, place, and person. There was no cranial nerve palsy. On motor examination, she had tender and swollen muscle groups bilaterally, but her muscle tone was normal. Muscle power was 1/5 over hip flexion, abduction, and adduction, as well as shoulder adduction and abduction. Knee flexion and extension were graded

2/5, while dorsiflexion, plantar flexion, ankle, and finger movements were 4/5. Sensory function was intact, and there were no signs of meningeal irritation.

Laboratory investigation

On laboratory tests, white blood cell (WBC) count was 9×10^3/μL ($N=72\%$, $L=21\%$) (normal range: 4.5–11.0×10^3/μL), hemoglobin (Hgb) was 10.3 g/dL (normal range: 12–18 g/dL), and platelets were 206×10^3/μL (normal range: $150–450\times10^3$/μL). Erythrocyte sedimentation rate (ESR) was 75 mm/h (normal range: 1–20 mm/h). Urine analysis showed hematuria of +2. Liver biochemical tests showed aspartate aminotransferase (AST) at 464.1 μ/L (11.5 × elevated) and alanine aminotransferase (ALT) at 114.3 μ/L (2.8 × elevated) (normal range: 0–40 μ/L); moderate hypoalbuminemia was at 2.25 g/dL (normal 3.5–5.0 g/dL). Renal function was normal. The creatine kinase level in the serum was 1649 μ/L (normal range: 29–168 μ/L). Serum tests for antinuclear antibodies and rheumatoid factor were negative. Other immunoassay panels were not performed owing to the limited clinical setup (see Table 1).

The chest X-ray showed no abnormalities. Echocardiography (ECG) and abdominal ultrasound findings were

Table 1 Laboratory profiles of the patient with dermatomyositis at Menelik II Comprehensive Specialized Hospital, Addis Ababa, Ethiopia

Variables	Laboratory values at admission	Reference range
White blood cells ($\times10^3$/μL)	9.0	4.5–11.0
Hemoglobin (g/dL)	10.3	12.0–18.0
Platelets ($\times10^3$/μL)	206	150–450
MCV (FL)	83.3	
ESR (mm/hr)	75	0–20
Urine analysis	**Results**	
Protein	Negative	
Glucose	Negative	
Blood	+2	
Liver biochemical tests	**Laboratory values at admission**	**Reference range**
ALT (IU/l)	114.3	0–40
AST (IU/l)	464.1	0–40
ALP (IU/l)	131.33	0–270
Bilirubin (total) (mg/dl)	0.8	0–0.1.0
Bilirubin (direct) (mg/dL)	0.2	0–0.2
Albumin (g/dL)	2.25	3.2–5.4
Renal function tests		
Serum Cr (mg/dL)	0.66	
BUN (mg/dL)	30.28	0–20
Other chemical tests		
Serum creatine kinase (IU/l)	1649	29–168
FBS (mg/dL)	95	70–110
Serum K^+ (mmol/L)	3.79	3.5–5.5
Serum Na^+ (mmol/L)	136.98	135–145
Serum calcium (mmol/L)	2.01	1.75–2.65
Serological tests	**Result**	
ANA	Nonreactive	
RF	Nonreactive	
HBSAg	Negative	
Anti-HCV antibody	Negative	
Anti-HIV antibody	Nonreactive	

ALT alanine aminotransferase, *ANA* antinuclear antibody, *AST* aspartate aminotransferase, *BUN* blood urea nitrogen, *Cr* creatinine, *ESR* erythrocyte sedimentation rate, *HBSAg* hepatitis B surface antigen, *RF* rheumatoid factor

normal. Upper gastrointestinal endoscopy revealed mild duodenitis; otherwise, the stomach and esophagus were normal. A muscle biopsy revealed muscle fiber atrophy, leading to the diagnosis of dermatomyositis on the basis of the American College of Rheumatology–European League Against Rheumatism (ACR–EULAR) classification criteria for dermatomyositis.

Prior to her final hospitalization and diagnosis, the patient had made several trips to the emergency room due to chronic dysphagia and weakness. Despite these visits, the underlying reason of her problem was not identified, which resulted in a number of needless and costly tests, such as a chest computed tomography (CT) scan and a barium swallow, which did not help determine that she had dermatomyositis. The prolonged course of undiagnosed dysphagia resulted in the development of adult malnutrition, as evidenced by her hypoalbuminemia (serum albumin 2.25 g/dL).

We started her on prednisolone 1 mg/kg (by mouth, po) daily (50 mg po daily), methotrexate 7.5 mg po weekly, folic acid 5 mg po daily, cotrimoxazole 960 mg three times weekly, and omeprazole 20 mg po twice daily. Mometasone body cream and liquid paraffin (moisturizer) were applied to the skin on a daily basis. Physical therapy was initiated as scheduled. She showed significant improvement in muscle strength and dysphagia within 3 weeks of therapy. She was referred to the chronic illness clinics of the hospital for close follow-up 3 weeks after admission to the medical ward at Menelik II Comprehensive Specialized Hospital.

Discussion

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by symmetric proximal muscle weakness and photosensitivity, with erythematous skin eruptions over the fingers, face, neck, and trunk. Most patients usually have elevated skeletal muscle enzymes during active myositis, and a muscle biopsy of perimysial and/or perivascular inflammatory infiltrates with muscle fiber necrosis suggests inflammatory myopathy. Electromyography (EMG) helps confirm the myopathic DM-related changes [1, 2].

The diagnosis of the patient was classified under "definite" idiopathic inflammatory myopathy [score 14.4 points (> 8.7 points with biopsy)] as per the "ACR–EULAR Classification Criteria for Idiopathic Inflammatory Myopathies," evidenced by an adult woman who had progressive, symmetric, proximal muscle weakness of both upper and lower extremities, neck flexor weakness, Gottron's and heliotrope rash, esophageal dysmotility, and elevated creatine kinase (CK) levels. In addition, a "probable" diagnosis of dermatomyositis was met using "Bohan and Peter's diagnostic criteria for dermatomyositis," evidenced by the detection of symmetric proximal muscle weakness, elevated serum skeletal muscle enzyme levels, and the typical skin rash of DM [3, 4].

The annual incidence of DM is approximately 1 per 100,000 with two primary forms: juvenile and adult, each having overlapping yet distinct presentations. There is a female-to-male ratio of 2:1, with the peak incidence in adults occurring between the ages of 40 and 50 years, although individuals of any age may be affected [4, 5]. Histopathologically, DM is characterized by muscle fiber necrosis, degeneration, and regeneration, with an inflammatory cell infiltrate. It is considered a humorally mediated disorder, with cellular infiltration predominantly in the perifascicular regions, leading to vascular destruction and tissue ischemia [2, 6]. The terminal complement C5b-9 membrane attack complex can be detected in vessel walls before inflammatory cells infiltrate, suggesting early activation of the complement cascade in DM [2, 6, 7]. Even through the advances in nonspecific immunotherapy, which have significantly improved outcomes, the absence of clearly defined target autoantigens continues to present challenges in developing specific immunotherapeutic approaches [7, 8].

To systematically evaluate potential differential diagnoses, this table highlights both the supporting and excluding features for each diagnosis.

Differential diagnoses	Rule-in findings (patient's findings)	Rule-out findings (patient's findings)
Dermatomyositis (DM)	• Heliotrope rash and Gottron's papules • Symmetric proximal muscle weakness • Elevated CK levels • Dysphagia due to esophageal dysmotility	• Hematuria (+2) is atypical for DM but can possibly be explained myoglobinouria
Pellagra	• Photosensitive hyperpigmented dermatitis (resembles some DM skin features) • Dysphagia potentially owing to malnutrition • Hypoalbuminemia owing to malnutrition	• Muscle-specific symptoms such as proximal muscle weakness and elevated CK is absent—hematuria is uncommon in pellagra

Differential diagnoses	Rule-in findings (patient's findings)	Rule-out findings (patient's findings)
Overlap syndrome scleroderma	• Esophageal dysmotility similar to patient's dysphagia • Possible renal involvement leading to hematuria in severe cases (renal crisis) • Elevated CK or significant muscle weakness • Typical DM skin findings (heliotrope rash and Gottron's papules)	• Absence of sclerodactyly (skin tightening/thickening) and Raynaud's phenomenon • Lack of systemic findings such as joint pain or pulmonary involvement, which are often seen in connective tissue overlap syndromes • No positive ANA • Normal renal function test

Diagnostic delays and early recognition

This case exemplifies diagnostic delays in resource-limited settings, with the patient undergoing multiple emergency visits and unnecessary tests before diagnosis, including a chest CT scan and a barium swallow. Early recognition of key clinical features, including heliotrope rash, Gottron's papules, and elevated CK levels, could have shortened the diagnostic timeline and reduced the need for expensive and potentially irrelevant investigations. As evidenced in this patient, who had malnutrition as a result of dysphagia, which led to hypoalbuminemia (2.25 g/dL) and mild anemia, extended diagnostic delays can further raise the risk of ramifications. Immunosuppressive treatment combined with early dietary management is essential to avoiding serious consequences [8, 9]. DM is relatively rare, with a global incidence of 1 to 10 cases per million people annually. While African Americans have a higher predisposition to developing DM and tend to experience more severe disease presentations, there are limited data regarding the prevalence of DM in Ethiopian and other African populations. African Americans are more likely to present with severe pulmonary involvement and myositis-associated malignancies, both of which increase morbidity and mortality [6, 10, 11].

Therapeutic approach

The first-line management of DM remains high-dose systemic corticosteroids. In our patient, she was started on high-dose prednisolone along with methotrexate, an immunosuppressive agent that was added as a steroid-sparing agent. It is an important addition to avoid the long-term use of high-dose steroids to avoid their associated side effects. She also received proton pump inhibitors for gastroesophageal reflux disease, a common complication in patients with DM, and also for protection from corticosteroid-induced gastric ulcers. Cotrimoxazole was administered for opportunistic infection prevention, and finally, topical steroids and moisturizers were prescribed for the dermatologic manifestations [12, 13]. After discharge, she was scheduled for outpatient follow-up to monitor the disease activity and adjust therapy accordingly.

Recommendations for clinical practice

In places similar to our country, where resources are limited, there is a need for early clinical recognition to avoid unnecessary diagnostic workups and complications that are highly preventable with early detection. Clinicians should have a high index of suspicion for identifying hallmark features such as skin rashes, symmetrical proximal muscle weakness, and elevated CK levels. In addition to this, clinicians should be aware that DM presents with more clinical severity in Africans, where it is highly underdiagnosed in this same population. Finally, further research and collaboration may be helpful in further clarification of racial disparity and disease severity for the high-risk populations.

Conclusion

Dermatomyositis is a rare disease entity. Prognosis depends on the severity of myositis, the presence of malignancy, and cardiopulmonary system involvement. Patients severely affected at presentation or treated after long delays, those with severe dysphagia or respiratory difficulties, older patients, and those with associated cancer tend to have a worse prognosis. The treatment of dermatomyositis includes high-dose glucocorticoids in addition to disease-modifying agents or cytotoxic agents such as methotrexate. Most patients improve with therapy, and many make a full functional recovery, often sustained with maintenance therapy. However, up to 30% of patients may be left with some residual muscle weakness. Relapses can occur at any time. Early management of the disease, identification of any internal malignancy, and monitoring for treatment toxicity are crucial during follow-up.

In conclusion, the patient described had dermatomyositis based on ACR–EULAR classification criteria. The low expectations and diagnostic challenges of this rare disease in limited clinical settings might contribute to delayed diagnosis and treatment. A high index of clinical suspicion is required to diagnose dermatomyositis in resource-limited settings to prevent adverse outcomes.

Abbreviations

ACR–EULAR	American College of Rheumatology–European League Against Rheumatism
ANA	Antinuclear antibody
ASS	Antisynthetase syndrome
DM	Dermatomyositis
EMG	Electromyography
MCTD	Mixed connective tissue disease

PM	Polymyositis
RA	Rheumatoid arthritis
RF	Rheumatoid factor
SSc	Systemic sclerosis
SLE	Systemic lupus erythematosus

Acknowledgements
We are grateful to the medical personnel who were caring for the patient.

Author contributions
All authors contributed to data analysis, drafting, or revising the article; have agreed on the journal to which the article will be submitted; gave final approval of the version to be published; and agree to be accountable for all aspects of the work.

Funding
The authors received no financial support from any source.

Data availability
The data set is available with authors and can be obtained from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
Our hospital does not require ethical approval for reporting individual case reports.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare no potential conflict of interest with respect to the research, authorship, or publication of the article.

Received: 26 February 2025 Accepted: 2 July 2025
Published online: 01 October 2025

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