Medicine & Clinical Science

Case Report

Dark Lesions in Dark-Skinned People - Dermatomyositis Diagnostic Challenge: First Report In The Gambia

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Abstract

Dermatomyositis (DM) is an autoimmune condition characterized by progressive muscle weakness and distinctive skin rashes. Diagnosing DM in black patients can be challenging, as they often experience disease manifestations that differ, in their clinical expression, from traditional descriptions. Classic skin signs, such as erythema and Gottron papules, may not be as easily visible on darker skin. In contrast, chronic changes such as dyschromia and poikiloderma are more prominent. These more subtle manifestations can lead to delays in diagnosis and misdiagnosis. It is essential that the doctor be able to include dermatological manifestations as part of the systemic picture, in our case expressed by respiratory manifestations, arthritis, Raynaud's phenomenon and muscle weakness. Facilitating the diagnosis of classic forms. Knowledge of these distinctive features is crucial for physicians to ensure a timely and accurate diagnosis.

Introduction

A 45-year-old black patient came to the clinic complaining of hyperpigmented skin lesions in areas of increased sun exposure. The changes began three years ago, followed by progressive fatigue, lasting about a year. Two months later, a progressive and marked reduction in strength in the upper limbs began. Recurrent attacks of pain and inflammation in the left wrist. He works in the port and is continually exposed to the sun and physical activity, being interpreted by medical personnel who have previously assessed it as the cause of the change in skin color and muscle weakness. He has no recorded history of previous illnesses, blood transfusions, or toxic habits. He denies difficulty swallowing. Dry cough and dyspnea after mild physical activity of 5, on a scale of to 10.

On physical examination, he appears to be chronically ill, with marked weight loss. Skin examination revealed melanodermic patches, in the regions most exposed to solar radiation, on the face, neck and arms (Figure 1), changes in the scalp with formation of scaly plaques, sometimes itchy (Figure 1.1). Flat, darker lesions on the upper back, shoulders, and nape (Figure 2). On the hands, multiple, flat, red, hyperkeratotic papules with central atrophy were seen on the posterior parts of the patient's metacarpophalangeal and interphalangeal joints, as well as mechanic's hands (Figure 3), cutis calcinosis of the auricle and cuticular hypertrophy with periungual capillary dilation were also observed (3.1). Arthritis of the left wrist is noted (Figure 4), with no evidence or history of another affected joint. Muscle strength assessment showed 4/5 lower and 3/5 upper limbs (MRC modified grading system). Globally decreased chest expansion and breath sounds. Raynaud phenomenon was observed (Figure 5).

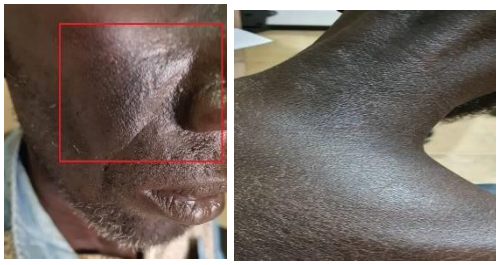

Figure 1. *Melanodermic patches exposed areas*

Figure 1.1. *Alopecia and scaly plaques*

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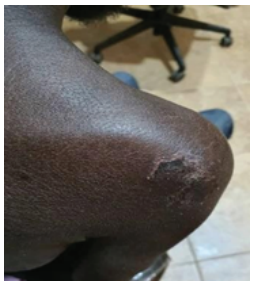

Figure 2. *Sclerosis and ulceration*

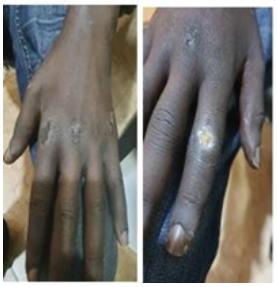

Figure 3. *Mechanic hands and Gottron papules*

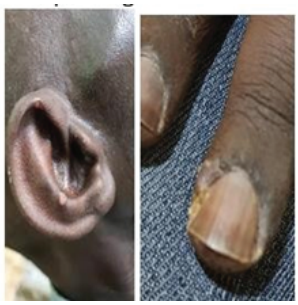

Figure 3.1. *Chondrocalcinosis And periungual infarcts*

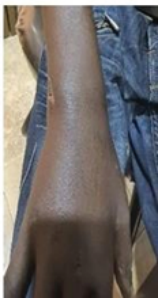

Figure 4. *wrist arthritis*

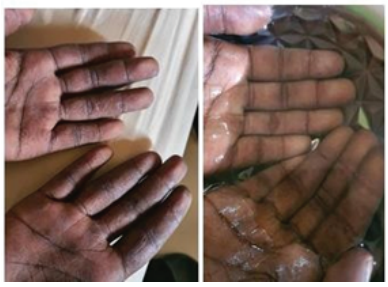

Figure 5. *Raynaud phenomenon*

Vital signs: HR 110beats/min BP 100/65mmhg
RR 21cycles/min; O2 sat: 94%
Temp: 36.80 C.

Hb: 9g/l	**Hep B/C: Negatives**	**CK total: 1750 u/L**
WCC: 12.5 x10^9/L	Urine analysis: Protein+, Red cels traces	LDH: 500 u/l
Plt: 75000 mcl	Rheumatoid Factor: Negative	Albumin: 21 g/l
CRP:77 mg/l	VDRL: Negative	
ESR: 96 mm/h	HIV(Negative)	
ASAT: 55 u/	ANA: Positive (qualitative)	
AlAT: 67 u/L	RBS: 8.5 mmol/l	

Wrist X ray: No erosive arthritis

Chest X ray: A bilateral pulmonary infiltrative pattern was observed with volume loss of the lower lobes.

Spirometry: Reduction in FEV1 and FVC with a restrictive pattern.

Skin Biopsy: increased lymphocytic infiltrate, and mucin deposition in the dermis with hyperpigmentation secondary to chronic inflammatory process. Degeneration, regeneration, necrosis, phagocytosis, and mononuclear perivascular infiltrates.

The patient's clinical presentation was characterized by typical skin lesions, Raynaud phenomenon, monoarthritis and respiratory symptoms. This picture was consistent with the diagnostic criteria (EULAR/ACR) for Dermatomyositis (DM) and Interstitial pulmonary disease. Initial management involved the administration of oral and topical steroids, along with hydroxychloroquine, azathioprine, and photo protection. Although there was a brief period of clinical improvement, the patient opted to discontinue treatment and did not attend the scheduled follow-up. Upon returning three months later with exacerbated skin lesions and new respiratory symptoms. Re-admitted with previously prescribed medications, plus antibiotics, and cyclophosphamide. Despite these interventions, his condition deteriorated, and passed in the ICU one week later, due to complications of acute interstitial pneumonia.

Discussion

The historical context of DM traces back to the 19th century, with significant contributions from E. Wagner in 1863 and P. Potain in 1875, which laid essential groundwork for the understanding of inflammatory muscle diseases [1,2]. However, the prevailing diagnostic criteria and medical literature have largely been shaped by experiences from light-skinned populations, leading to considerable diagnostic disparities for individuals with darker skin tones. This lack of representation may contribute to the underdiagnosis and delayed recognition of DM, particularly in sub-Saharan Africa, where patients often present only after the disease has advanced significantly [3-5]. An initial evaluation for suspected DM should encompass a comprehensive history, physical examination, total skin assessment, and muscle strength testing of the extremities and neck flexors. Our case coincides with the classic descriptions of DM. Highlighting the importance of identifying typical patterns in dark-skinned patients.

DM variants

Classic DM	It affects both the skin and muscles. In up to half of patients, skin changes appear before muscle weakness.
Amyopathic DM	Only involves the skin, without muscle weakness for at least six months.
Hypomyopathic DM	The skin and some evidence of muscle inflammation, but no obvious muscle weakness
Clinically amyopathic DM	skin but doesn't cause obvious muscle symptoms.
Juvenile DM	Affects patients under 18 years old. It can cause muscle weakness, calcinosis cutis, and other symptoms.
Post-myopathic DM	A type of DM that affects patients who previously had classic DM but have recovered from myositis while still experiencing skin rashes.
DM sine dermatitis	It doesn't cause a rash, but a muscular biopsy shows signs of DM.

The differential diagnosis can be long, depending on the existence or not of muscular and respiratory manifestations. It can include primary or secondary muscular diseases, as well as autoimmune disorders, vitamin deficiencies, paraneoplastic syndromes, endocrine conditions, or sicknesses induced by toxic and drugs:

1. Polymyositis (PM): Unlike DM, PM does not have characteristic skin manifestations.
2. Inclusion Body Myositis (IBM): This condition often affects older adults and has slower progression with distinctive biopsy findings.
3. Systemic Lupus Erythematosus (SLE): SLE can present with muscle weakness and skin rashes, but it has other systemic features and specific autoantibodies.
4. Conditions like mixed connective tissue disease and scleroderma can have overlapping features with DM, however biopsy and specifics ANA could be helpful in the diagnosis.
5. Muscular Dystrophies: These genetic disorders can present muscle weakness but have different clinical and genetic profiles and are unlikely to start in an adult patient.
6. Drug-Induced Myopathies: Certain medications can cause muscle inflammation and weakness, mimicking DM.
7. Endocrine myopathy usually presents as a late complication of the disease; however, it can begin even before endocrine manifestations [6]. Therefore, it is recommended to request studies and rule out endocrine axis syndromes (Pituitary, Thyroid and adrenal cortex).

Cutaneous manifestations are essential clinical features of DM and can be classified as pathognomonic, characteristic, compatible, or rare [7]. Notably, pathognomonic signs include Gottron papules (observed in 86% of cases) and heliotrope rash. In fact, 92.6% of patients lacking Gottron's sign exhibit alternative findings such as the V sign, shawl sign, holster sign, mechanic's hands, telangiectasia, erythema, or periungual dystrophy. Additionally, Raynaud's phenomenon occurs in up to 40% of DM patients, while calcinosis cutis is reported in approximately 4% of adults with DM. Rarely, tumoral calcinosis of the auricle may present, classified as dystrophic, metastatic, idiopathic, or iatrogenic. The overlap of cutaneous manifestations with other connective tissue diseases can complicate diagnosis, leading to potential misdiagnosis as lupus erythematosus (LE) due to similar photo-distributed lesions. Creatine kinase serves as the primary muscle enzyme for diagnosis and monitoring therapeutic response in polymyositis/dermatomyositis, although some patients may present with normal levels despite the disease [8,9]. Electromyographic studies have historically been a support in the diagnosis, despite their positive or negative nature by themselves neither confirming nor excluding the diagnosis.

The diagnosis of DM remains complex, with delays between initial presentation and conclusive diagnosis varying from 2.2 weeks to several years, with an average of 15.5 months. Historically, DM was defined using the Bohan and Peter criteria, which include five key elements: symmetrical weakness of the neck and girdle flexor muscles, muscle biopsy confirming myositis, elevated serum skeletal muscle enzymes, characteristic findings in needle EMG and specific dermatological manifestations such as heliotrope rash and Gottron sign. A diagnosis is considered "definite" with three criteria and the characteristic rash, "probable" with two criteria and rash, and "possible" with one criterion and rash. Since then, additional classification systems have emerged, including those of Tanimoto, Targoff, Dalakas, and the European Neuromuscular Center [10-13]. The most recent criteria established by the European League Against Rheumatism and the American College of Rheumatology in 2017 reflect significant advances in this evolving field. In our case, the patient satisfied the past and recent diagnostic criteria, including the EULAR/ACR, which we accessed through the link (http://www.imm.ki.se/biostatistics/calculators/iim.

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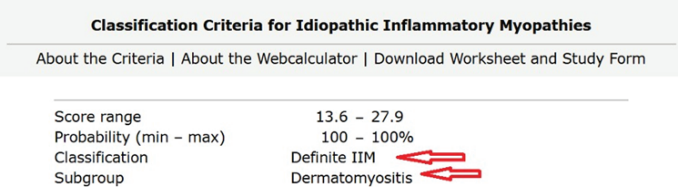

The etiology of the disease is multifactorial, incorporating genetic, environmental, and immunological factors, although its full understanding remains elusive. Environmental triggers—such as ultraviolet radiation, viral infections, medications, and pollutants—may induce immune dysregulation in individuals with genetic predispositions. Incidence rates exhibit significant geographic variability, with a higher prevalence observed in women, particularly in middle-aged populations. North America and Europe report incidences ranging from 0.5 to 7.6 cases per 100,000 individuals annually, whereas East Asia shows much lower rates, around 0.4 cases per 100,000. In Africa, the reported incidence is approximately 7.5 per million person-years, with a prevalence of 11.49 per 100,000; however, limited documentation suggests underdiagnosis due to inadequate healthcare resources and a scarcity of specialists. This delayed diagnosis poses serious risks, particularly as the likelihood of malignancy increases significantly within the first two years following symptom onset [14-16].

To effectively address the diagnostic challenges associated with DM, particularly in individuals with hyperpigmented skin,

it is imperative to enhance awareness and education within healthcare regarding the varied presentations of skin conditions across diverse populations. Recognizing the correlation between typical dermatological manifestations, symmetric proximal muscle weakness, and interstitial lung disease can streamline the diagnostic process. Delayed recognition may result in severe complications, including respiratory impairment and an increased risk of malignancy.

Therefore, early intervention is critical for improving patient outcomes. As a limitation in our report, it is valid to point out the absence of specific ANA studies for dermatomyositis, they are not available in the country.

Conclusion

Diagnosing Dermatomyositis in patients with hyperpigmented skin presents a considerable challenge, often resulting in delayed recognition and treatment of this rare yet potentially serious condition. It is essential to address this knowledge gap within clinical practice to ensure equitable access to timely diagnosis and appropriate management. By doing so, we can enhance outcomes for all individuals affected by Dermatomyositis, irrespective of their skin tone.

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