Distinctive cutaneous features of dermatomyositis in Black adults: A case series

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INTRODUCTION

Dermatomyositis (DM) is a rare inflammatory myopathy shown to disproportionately affect Black individuals, with estimated incidence 3-fold greater than that in White individuals.[1] As with multiple cutaneous diseases in skin of color, specific manifestations of DM in Black patients remain underreported in educational sources and scientific literature, leading to potential misdiagnoses or diagnostic delays.[2]

CASE SERIES

Black patients diagnosed with DM from 2000 to 2021 at Mass General Brigham were identified using the Research clinical database. Demographics and clinical presentation data were extracted from records. This study was approved by the Mass General Brigham Institutional Review Board.

Fourteen adult patients (12 women and 2 men) with an average age of 52 years (range, 27-78 years) were identified (Table 1). Dyschromia was the most prominent cutaneous feature noted, with 8 of 14 patients demonstrating hypopigmentation, hyperpigmentation, and/or poikiloderma. Gottron papules were observed in 5 patients. Six patients demonstrated proximal nailfold changes (dilated capillary loops, ragged cuticles, cuticular hypertrophy, capillary darkening). Facial erythema was noted in 3 patients, 4 had V-neck erythema, and 5 had a positive shawl sign. Two patients demonstrated a classic heliotrope rash, whereas another was found to have periorbital changes described as "hypopigmented patches with telangiectasias, and erythema."

Abbreviations used:

DM: dermatomyositis

Systemic manifestations were also common. Thirteen patients had confirmed evidence of proximal muscle weakness (positive muscle biopsy, elevated muscle enzymes, magnetic resonance imaging, electromyography findings). Interstitial lung disease, one of the strongest prognostic indicators of DM, was found in 43% of the patients compared with a prevalence of 41% in a global meta-analysis of polymyositis/DM cases.[3] Five patients had associated neoplasms: multiple myeloma (n = 1), metastatic breast cancer (n = 1), pituitary adenoma (n = 2), and meningioma (n = 1). Calcinosis was present in 4 of 14 patients.

DISCUSSION

There is a dearth of literature examining the clinical presentation of DM in Black patients, with the potential to contribute to diagnostic delays and poor outcomes. In juvenile DM —a disease often with a distinct clinical course relative to adult DM—calcinosis is more common among Black individuals than in other groups.[4] In our analysis, calcinosis was present in 29% of Black adults compared with that in prior literature, which

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Consent for the publication of recognizable patient photographs or other identifiable material was obtained by the authors and included at the time of article submission to the journal stating that all patients gave consent with the understanding that this information may be publicly available.

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Table I. Clinical characteristics of 14 Black patients with dermatomyositis

Age	Sex (M/F)	Muscle involvement*	Gottron papules	Nailfold involvement	Heliotrope rash	Facial erythema	V-neck erythema	Shawl sign	Dyschromia	Scalp findings	Other findings
38	F	Yes	Yes	Dilated capillary loops	No	No	No	No	Poikiloderma	Yes	Nonsecreting pituitary adenoma
67	F	Yes	No	Ragged cuticles; dilated capillaries	No	Yes	Yes	Yes	Dyschromic xerotic poikilodermatous plaques	Yes	ILD, esophageal dysfunction
27	F	Yes	Yes	Dilated capillaries and dropout	Yes	No	Yes	No	Poikiloderma	Yes	Calcinosis cutis
37	M	Yes	Yes	No	No	No	No	No	No	No	ILD, calcinosis cutis; Raynaud; esophageal dysmotility
41	F	Yes	No	Dilated capillary loops, dropout; ragged cuticles; cuticular hypertrophy	Hypopigmented patches with telangiectasias and erythema	Yes	Yes	Yes	Atrophic plaques + poikiloderma	Yes	Calcinosis cutis; Raynaud; dysphagia
73	F	No	No	No	No	No	No	No	Hyperpigmentation	No	Meningioma; calcinosis cutis; Raynaud
38	F	Yes	Yes	No	No	No	No	Yes	Hypopigmentation; hyperpigmentation w/ thickening; poikiloderma	No	Dysphagia, metastatic breast cancer
27	M	Yes	No	Capillary darkening	No	Yes	No	No	Hypopigmentation and erythema in a malar distribution	No	Dysphagia
76	F	Yes	No	No	Yes	No	No	Yes	No	No	ILD; esophageal dysmotility
54	F	Yes	No	No	No	No	No	No	No	No	ILD, adrenal insufficiency
55	F	Yes	No	No	No	No	No	No	No	No	No
78	F	Yes	No	No	No	No	No	No	No	No	Multiple myeloma
70	F	Yes	No	No	No	No	No	No	No	No	ILD; intestinal dysmotility; pituitary microadenoma
66	F	Yes	Yes	Dilated capillary loops, ragged cuticles	No	No	Yes	Yes	Dyschromia; atrophy; pigment dropout	No	ILD

CK, creatine kinase; ILD, Interstitial lung disease.
*Muscle involvement as proven by magnetic resonance imaging, electromyography, muscle biopsy, muscle enzymes (CK and aldolase).

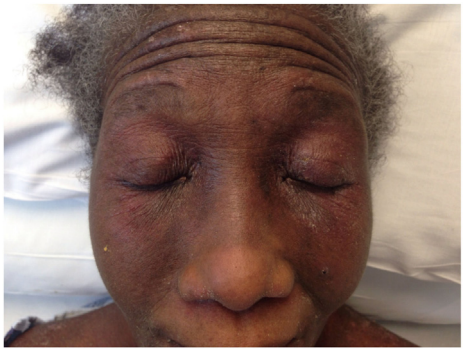

Fig 1. Subtle periorbital erythema involving the upper and lower eyelids and upper portion of the cheeks ("Heliotrope rash") in a Black patient with dermatomyositis.

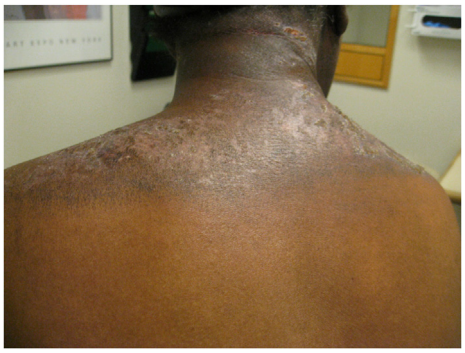

Fig 2. Subtle erythema with a background of dyschromia on the upper portion of the back, shoulders, and back of neck (Shawl sign) in a Black patient with dermatomyositis.

estimated a lower prevalence of calcinosis in up to 20% of adults with DM.[5] Our analysis represents one of few to examine the cutaneous manifestations of DM specifically in Black patients (Figs 1 to 3). Although these differences may be in line with estimates based on larger data sets, more subpopulation analyses are warranted to better understand specific comorbidities of DM in Black patients by assessing greater numbers of patients.

In this study, skin dyschromia, rather than erythematous/violaceous patches and plaques, was more pronounced in pathognomonic locations compared with that in individuals with lighter skin tones. These findings suggest that clinicians should consider skin tone when evaluating classic anatomic locations for DM in Black patients. Nearly all patients with DM present with evidence of proximal nailfold changes (eg, capillary changes, ragged cuticles). However, less than half of patients herein demonstrated proximal nailfold changes, suggesting that these findings present more subtly in Black patients. Thorough nailfold examinations for capillary and cuticular changes may be of particular diagnostic value.

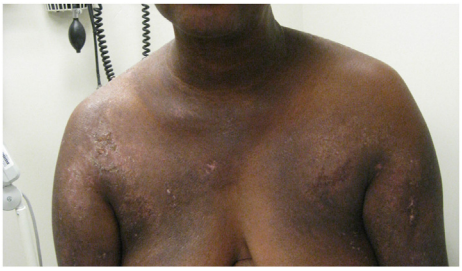

Fig 3. Dyschromia on the lower portion of the anterior aspect of the neck and upper portion of the anterior aspect of the chest ("V sign") in a Black patient with dermatomyositis.

Although this study is the first to detail the clinical manifestations of DM in Black patients, it is not without limitations. Because these patients were seen in routine clinical practice, variability in documentation in medical records is a limitation; some patients were assessed using the Cutaneous Dermatomyositis Disease Area and Severity Index, whereas others were not.

Further, this study is based on retrospective data, and the sample size is limited. It is possible that the database undercaptures Black individuals because of a variety of factors, including lack of standardized race/ethnicity capture across sites and possible disproportionate opt-out of the system by individuals from backgrounds historically marginalized by systemic forces within academic medicine. Despite these limitations, this study aimed to raise awareness regarding distinct features that may aid in the diagnosis of DM in Black patients. Future studies examining larger cohorts and additional racial groups are warranted.

Conflicts of interest

Dr LaChance is the principal investigator for a research grant from Pfizer for a project exploring the role of the JAK/STAT pathway in cutaneous connective tissue disease. Dr Vleugels is a principal investigator for Pfizer. Drs Ezeofor, O'Connell, Cobos, and Nambudiri have no conflicts to declare.

REFERENCES

1. Medsger TA Jr, Dawson WN Jr, Masi AT. The epidemiology of polymyositis. *Am J Med*. 1970;48(6):715-723. https://doi.org/10.1016/s0002-9343(70)80006-7
2. Adelekun A, Onyekaba G, Lipoff JB. Skin color in dermatology textbooks: an updated evaluation and analysis. *J Am Acad Dermatol*. 2021;84(1):194-196. https://doi.org/10.1016/j.jaad.2020.04.084
3. Sun KY, Fan Y, Wang YX, Zhong YJ, Wang GF. Prevalence of interstitial lung disease in polymyositis and dermatomyositis: a meta-analysis from 2000 to 2020. *Semin Arthritis Rheum*. 2021;51(1):175-191. https://doi.org/10.1016/j.semarthrit.2020.11.009

4. Phillippi K, Hoeltzel M, Byun Robinson A, Kim S; Childhood Arthritis and Rheumatology Research Alliance (CARRA) legacy registry investigators. Race, income, and disease outcomes in juvenile dermatomyositis. *J Pediatr*. 2017;184:38-44. https://doi.org/10.1016/j.jpeds.2017.01.046

5. Valenzuela A, Chung L, Casciola-Rosen L, Fiorentino D. Identification of clinical features and autoantibodies associated with calcinosis in dermatomyositis. *JAMA Dermatol*. 2014;150(7):724-729. https://doi.org/10.1001/jamadermatol.2013.10416