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Keloidal Morphea: A Unique Variant of Morphea Presenting in a Black Female

Nicole C. Syder MD,[a] Autumn L. Saizan MD,[b] Shanice McKenzie MD,[c] Brittney De Clerck MD,[c] Arielle Carolina Mora Hurtado BS,[d] Nada Elbuluk MD MSc[c]

[a]Department of Dermatology, University of California San Francisco School of Medicine, San Francisco, CA
[b]Department of Dermatology, Johns Hopkins University School of Medicine, Baltimore, MD
[c]Department of Dermatology, Keck School of Medicine of the University of Southern California, Los Angeles, CA
[d]University of Wisconsin School of Medicine and Public Health, Madison, WI

ABSTRACT

Keloidal morphea is a rare variant of cutaneous scleroderma that presents with keloid-like plaques and nodules that occur most commonly on the neck, trunk, and proximal extremities. They occur in the absence of trauma or injury. Keloidal morphea most often manifests in patients with skin of color and may be mistaken for keloids and/or hypertrophic scars. This report presents a case of keloidal morphea in a young Black female. While effective therapies are limited, timely diagnosis can initiate the evaluation for systemic sclerosis and other systemic organ involvement. Increased awareness and recognition of keloidal morphea can help prevent dermatologic health disparities, which disproportionately occur in patients of color.

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CASE REPORT

A 26-year-old Black female with a history of Raynaud's syndrome presented with complaints of a pruritic dermatitis on the right abdomen, right thigh, and chest. She noted that it first began on her right thigh 1.5 years prior and, more recently spread to her chest and neck. She denied any pain or tenderness or any prior dermatitis or trauma. She endorsed a family history significant for systemic lupus erythematous in her aunt.

Physical exam findings revealed dark brown to violaceous, smooth, well-demarcated, indurated, and hypertrophic plaques and dark brown patches on her upper chest, right neck, abdomen, right thigh, bilateral flanks, and suprapubic region.

A punch biopsy revealed marked cellular fibroplasia with a proliferation of capillaries and fibroblasts. CD34 staining showed a decrease in dermal dendritic cells. Histopathologic findings showed dermal fibroplasia, consistent with scar possibly related to keloid formation. Repeat biopsy revealed a diffuse fibrosing process with features of a hypertrophic scar and loss of the CD34 dermal dendritic cell network, as well as sclerosis into the subcutaneous septae. Through her combined clinical presentation and histopathology, she was diagnosed with keloidal morphea. Topical treatment was initiated with a combination of topical clobetasol and calcipotriene cream with a plan to add intralesional steroid injections. The patient was also advised to follow up with rheumatology for further

FIGURE 1A. Indurated violaceous brown nodules and plaques on left chest.

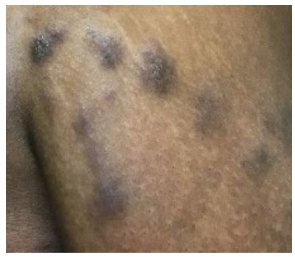


FIGURE 1B. Diffuse dark brown patches and plaques along the right flank.

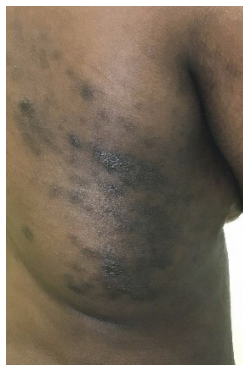
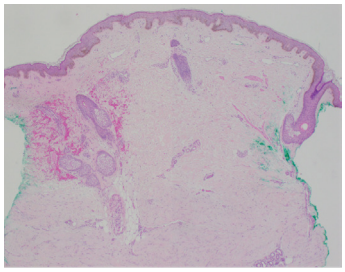


FIGURE 1C. Indurated brown patches and plaques on the mid-abdomen.

FIGURE 2. Marked cellular fibroplasia with a proliferation of capillaries and fibroblasts with variable lymphohistiocytic infiltrate. 10x magnification H&E.



workup to evaluate for systemic scleroderma. The patient was unfortunately lost to follow up.

DISCUSSION

Scleroderma is a chronic autoimmune connective tissue disease that can be systemic, referred to as systemic sclerosis, or purely cutaneous, also known as morphea. Keloidal morphea and nodular scleroderma are considered rare variants of cutaneous scleroderma. Of note, Raynaud's phenomenon can precede this disease by many years, as seen in this case.[1]

Keloidal morphea can sometimes be mistaken for keloids and/or hypertrophic scars. This condition presents mainly in Black women and can occur at any age.[2] Some studies have suggested that the mechanism behind this disorder involves a dermal inflammatory process of sclerosis forming keloidal lesions.[3] Another proposed mechanism involves microtrauma and inflammation, leading to excessive fibrosis in individuals predisposed to keloid formation.[2]

Keloidal morphea typically presents as multiple keloid-like plaques and nodules that occur in the absence of trauma or injury. They can be skin colored, dark brown, or violaceous plaques with pseudopod extensions. The most commonly affected areas are the neck, trunk, and proximal extremities.[2] On histology, high levels of tenascin, connective tissue growth factor, and transforming growth factor-beta (TGF-beta) cytokines can be seen, all factors known to contribute to fibrosis and scar formation.[3]

Due to its rarity, there is limited data in the literature regarding effective treatment options. Topical or intralesional corticosteroids, topical vitamin D analogs, topical tacrolimus, methotrexate, azathioprine, ultraviolet (UV) light therapy, and systemic steroids are current options for therapy.[4] Few cases have been able to achieve full resolution with surgical removal. Recently, a study found Dipeptidyl peptidase-4 (DPP-4) to be a marker of active fibroblast activity and, thus, a potential target for anti-fibrotic therapy in patients with systemic sclerosis.[5] More studies are needed to investigate this further.

Physicians need to have a high index of suspicion for this condition when patients present with extensive keloidal-appearing plaques and nodules without prior history of trauma or injury to affected areas. Patients with confirmed keloidal morphea should subsequently be evaluated for systemic sclerosis and other systemic organ involvement. Knowledge of this condition can help lead to an appropriate and timely diagnosis as well as help decrease dermatologic health disparities amongst skin of color populations.

DISCLOSURES

Dr. Elbuluk has served as a consultant, advisory board member, and/or speaker for Avita, Incyte, VisualDx, La Roche Posay, Beiersdorf, Allergan, Eli Lilly, Galderma, Pfizer, Takeda, Abbvie, Janssen, Sanofi, L'Oreal, McGraw Hill, Dior, Medscape. She has grant funding from Pfizer, has received royalties from McGraw-Hill and has stock options in VisualDx. The other authors have no conflicts to disclose.

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AUTHOR CORRESPONDENCE

Nada Elbuluk MD MSc
E-mail:..... elbuluk@usc.edu