

Superficial morphea: A rare condition and report of three unique cases



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INTRODUCTION

Classic morphea, or localized scleroderma, is an inflammatory connective tissue disorder involving sclerosis and fibrosis of the skin and underlying tissues. There are many subtypes, including plaque, linear, generalized, pansclerotic, mixed, and superficial. Morphea and its subtypes predominantly affect light-skin women. We present 3 unique cases of superficial morphea: a 35-year-old male (Fitzpatrick type III), a 45-year-old male (Fitzpatrick type V), and a 28-year-old female (Fitzpatrick type IV).

CASE SERIES

Case 1—A 28-year-old female presented to clinic for a 1-year history of worsening “stretch marks” in her bilateral axilla and medial aspects of the thighs. The patient reported mild discomfort with movement of her upper extremities. Past medical history was significant for a 1-year history of dysuria and vaginal itching. Sexually transmitted infection workup was negative. Physical examination revealed numerous light-pink, atrophic, shiny patches in the bilateral axillae, medial aspects of the upper arms, antecubital fossa, and medial aspects of the thighs (symmetric intertriginous). Some areas had superficial ulceration (Fig 1).

Case 2—A 45-year-old male presented with a 1-year history of hyperpigmentation around the neck and lower face, which worsened with sweating and during summer. He was treated with hydroquinone cream for 6 months with no improvement. He also tried and failed ketoconazole cream and shampoo prescribed by his primary care provider for possible tinea versicolor. The patient was also treated with betamethasone-clotrimazole cream with no improvement. Physical examination revealed hyperpigmented patches on the anterior and posterior

Abbreviations used:

IAPP: idiopathic atrophoderma of Pasini and Pierini
 LS: lichen sclerosis

aspects of the neck, lower face, and back (Fig 2). The axillae and arms were clear.

Case 3—A 35-year-old male presented to the clinic with a 6-month history of hyperpigmented, itchy, patches on the chest, inguinal folds, and back. Physical examination revealed hyperpigmented, shiny, atrophic patches in the inguinal folds, chest, and back along with a hyperpigmented patch on the central aspect of the back (Fig 3).

DISCUSSION

Superficial morphea was first reported by McNiff et al in 1999.¹ Since then, 7 cases have been published. The major defining characteristics of superficial morphea include bilateral atrophic plaques or patches in the intertriginous areas and/or trunk and histologically thickened collagen in the superficial dermis with presence of elastic fibers.² Differential diagnosis includes classic morphea, lichen sclerosis (LS), and idiopathic atrophoderma of Pasini and Pierini (IAPP). IAPP commonly coexists with morphea and is believed to be an abortive scleroderma in which sclerosis fails to form. IAPP is histologically characterized with variable epidermal atrophy: decreased thickness with flattening of rete ridges.³

Case 1 presented with atrophic patches and few areas of superficial ulceration. Case 2 presented with hyperpigmented patches around the neck. Case 3 presented with hyperpigmented atrophic patches in

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Fig 1. Case 1: Superficial morphea: A 28-year old female with hyper- and hypopigmented atrophic, painful, shiny patches in the bilateral intertriginous areas.

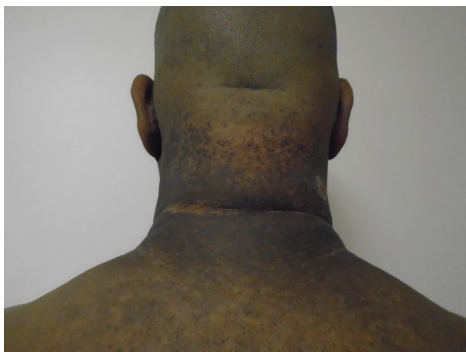


Fig 2. Case 2: Superficial morphea: A 45-year old male with hyperpigmented patches on the neck and lower aspect of the face.

the intertriginous areas. Our cases reflect the variability in presentation of superficial morphea, which can be a result of disease progression or differences in demographics.

All 3 biopsies showed superficial dermal sclerosis, pauci-cellular subepithelial sclerosis, and presence of elastic fibers. The deeper dermis was unchanged (Figs 4, 5, and 6). The dermis in case 2 had fewer inflammatory cells and more extensive sclerosis (Fig 5), which could indicate a late post-reactive phase, since he was presented to our clinic more than a year after his initial symptoms began. The dermis in case 3 had an extensive inflammatory infiltrate and thinner layer of sclerosis, consistent with recent active disease (Fig 6). He presented to our clinic within 6 months of his initial symptoms, and his patches responded well to class 1 topical steroids. Histopathology of case 3 does not rule out LS. Morphea and its subtypes can exist as a spectrum within LS; however, we were not inclined to diagnose LS because of the distribution of the lesions and the clinical findings. Similarly, these patients could have concomitant deeper morphea with



Fig 3. Case 3: Superficial morphea: A 35-year old male with hyperpigmented slightly atrophic, pruritic, shiny patches on the waistline, inguinal folds, chest, and back.

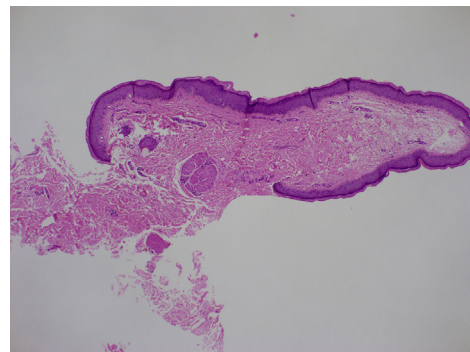


Fig 4. Case 1: Superficial morphea. Biopsy specimen shows a narrow area of sclerosis in the superficial reticular dermis with intact elastic tissue. (Hematoxylin-eosin-stain; original magnification, $\times 40$.)

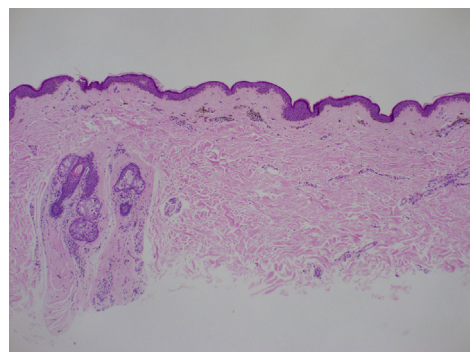


Fig 5. Case 2: Superficial morphea. Biopsy specimen shows a narrow area of sclerosis in the superficial reticular dermis with intact elastic tissue. (Hematoxylin-eosin-stain; original magnification, $\times 40$.)

superficial morphea. However, in each case, the biopsy site was an active and representative lesion, and there was no fibrosis in the deeper dermis or subcutaneous tissue.

All 3 cases showed significant improvement with 0.05% clobetasol propionate ointment applied twice

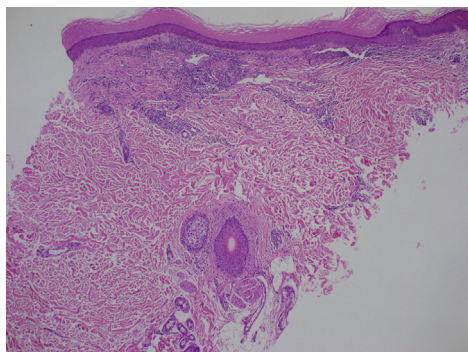


Fig 6. Case 3: Superficial morphea. Biopsy specimen shows compact orthokeratosis overlying epidermal atrophy. There is a layer of pauci-cellular subepithelial sclerosis with a vacuolar interface at its leading edge. There is a thin layer of superficial papillary dermal sclerosis. Even deeper is interspersed inflammatory infiltrate with rope-like collagen bundles. Elastin fibers are intact. (Hematoxylin-eosin-stain; original magnification, $\times 40$.)

daily to affected areas. All patients reported softening of the skin and resolution of the atrophy/thinning noted in the skin of those areas. Case 1 reported significantly softer, less atrophic, and less itchy patches (Fig 7). She reported less pain and improved mobility of her upper extremities as well. There were areas of continued involvement and new lesions in her antecubital fossa. She did not present to our clinic until 1 year after her first symptoms, and her generalized involvement with ulceration could give insight into disease progression of untreated superficial morphea. Case 2 reported softer, less atrophic



Fig 7. Case 1: Superficial morphea: A 28-year old female after 0.05% clobetasol propionate ointment applied twice daily to affected areas.

patches on his neck and face. Case 3 reported that all the patches became softer and were no longer shiny.

Conflicts of interest

Dr. Swapna Reddy is a speaker for Ilumya (tildrakizumab) with Sun Pharmaceuticals.

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