Superficial morphea: case report, look-alikes, pathogenesis, and treatment

Jameson Loyal[1] BA, Ivy I. Norris[1,2] MD, Elizabeth B. Lester[1,2] MD, Joseph C. Pierson[1,2] MD

Affiliations: [1]University of Vermont College of Medicine, Burlington, Vermont, [2]University of Vermont Medical Center, Burlington, Vermont

Corresponding Author: Elizabeth Lester, University of Vermont Medical Center, Division of Dermatology, 111 Colchester Avenue, Burlington, VT 05401, Email: Elizabeth.Lester@uvmhealth.org

Abstract

Superficial morphea, a rare variant of morphea, is characterized by hypopigmented to hyperpigmented skin lesions located predominantly in a symmetric fashion at intertriginous sites. These patches and plaques typically lack the significant induration, contractures, and atrophy seen in other subtypes of morphea. Histologic examination is key for accurate diagnosis considering the number of similar conditions which may clinically mimic superficial morphea. Herein, we present a case of a 25-year-old woman who re-presented for consultation in our clinic after gradual progression of her skin lesions. In addition, we review dermatologic look-alikes, as well as the pathophysiology and treatment options for superficial morphea.

Keywords: superficial morphea, morphea, localized scleroderma, scleroderma

Introduction

Superficial morphea, one of the many variants of morphea, is characterized by cutaneous hypopigmented to hyperpigmented patches and plaques in a symmetric fashion located predominantly at intertriginous sites that lack significant induration, contractures, and atrophy [1-3, 6]. Histologic examination of the lesions along with a detailed history and physical examination is critical as there are many look-alike conditions that clinically present similarly.

Case Description

A 25-year-old woman previously diagnosed with morphea localized to the left hand was recently referred to the dermatology service owing to progression of her morphea to include the bilateral forearms, flanks, abdomen, chest, axilla, thighs, and posterior knee (**Figures 1, 2**). Although she initially experienced no change in the sensation of involved skin, she also now noted the development of a feeling of "tightness" in and around some of the affected areas.

The patient had numerous brown, polycyclic patches and plaques located in a symmetric distribution on the forearms crossing the elbow joint, left hand, bilateral inner thighs, bilateral flanks, and central chest (**Figures 1, 2**). There were several notable plaques on the flanks with an inflamed appearance. The plaques on the bilateral forearms were firmer than those appreciated on the patient's abdomen and chest.

A punch biopsy of the left upper torso was submitted for pathologic examination and revealed superficial dermal fibrosis with associated patchy inflammation and reactive changes (**Figures 3, 4**). The histologic features of the deeper levels of the specimen were relatively non-specific. Superficial morphea was diagnosed based on the patient history, clinical examination, and biopsy results.

Initial treatment of the patient's left hand consisted of topical clobetasol 0.05% and triamcinolone 0.1% cream, which were relatively ineffective. She subsequently underwent a three month trial of narrow-band (nbUVB) without improvement. Topical

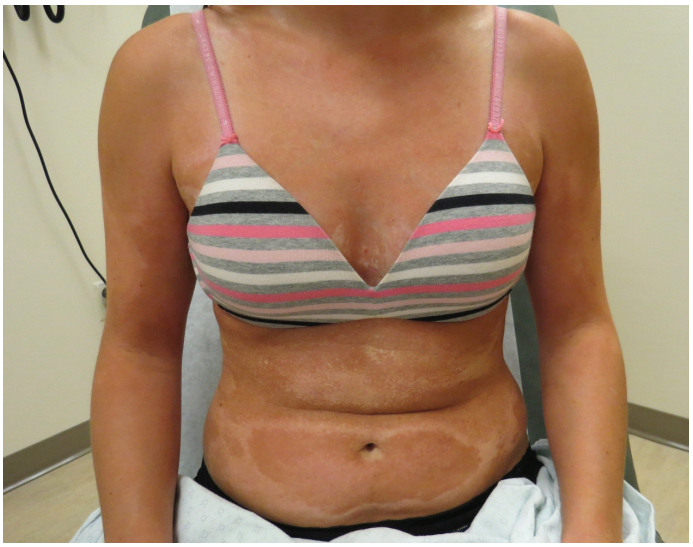

Figure 1. *Hypopigmented and hyperpigmented, minimally indurated plaques are seen on the bilateral inferior neck, upper arms, chest, and abdomen.*

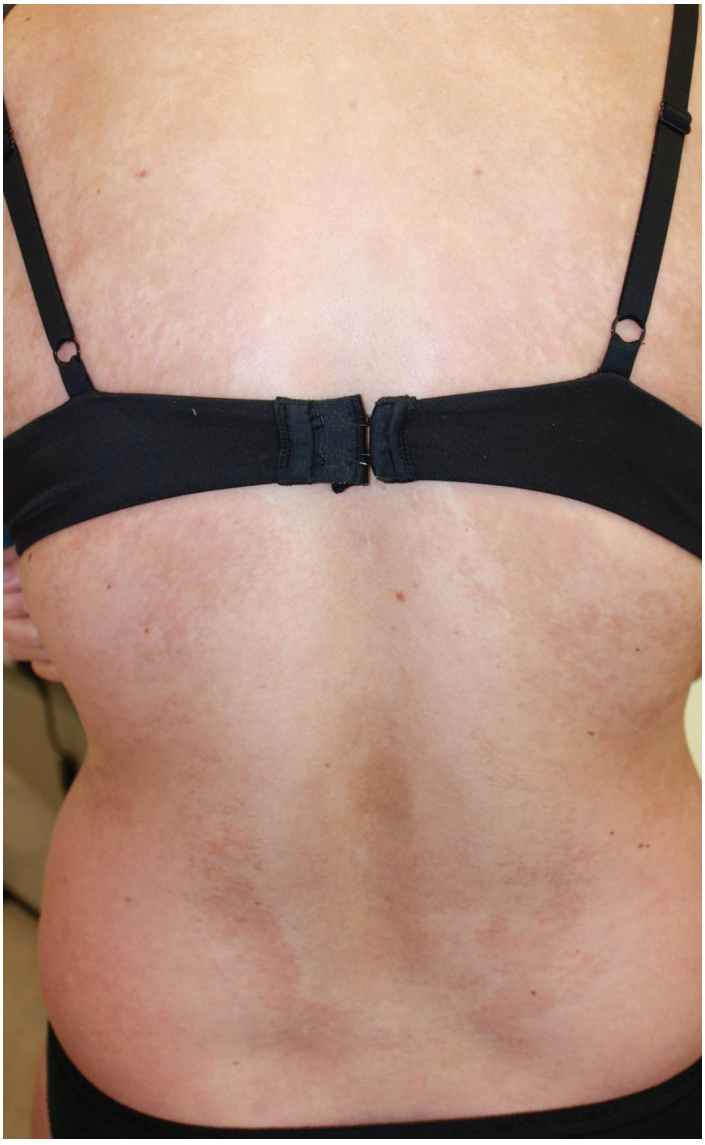

Figure 2. *A broad plaque with mottled hyperpigmentation and hypopigmentation involves the lower back.*

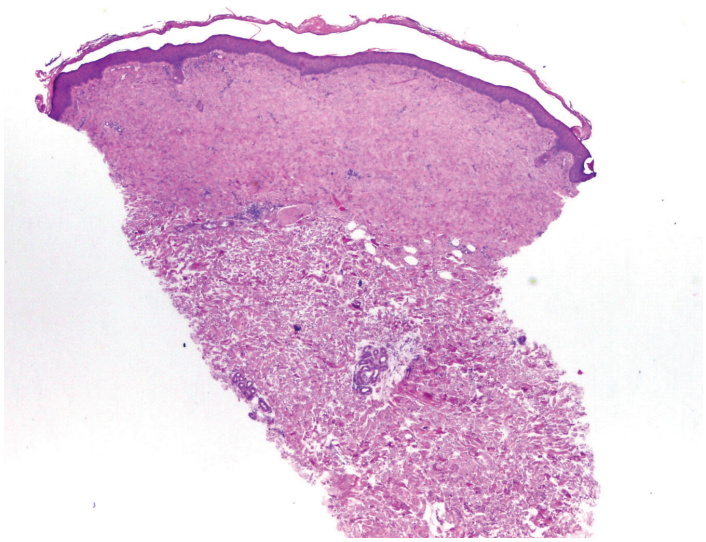

Figure 3. *Superficial dermal sclerosis with associated patchy, lymphomononuclear inflammation. H&E, 4x.*

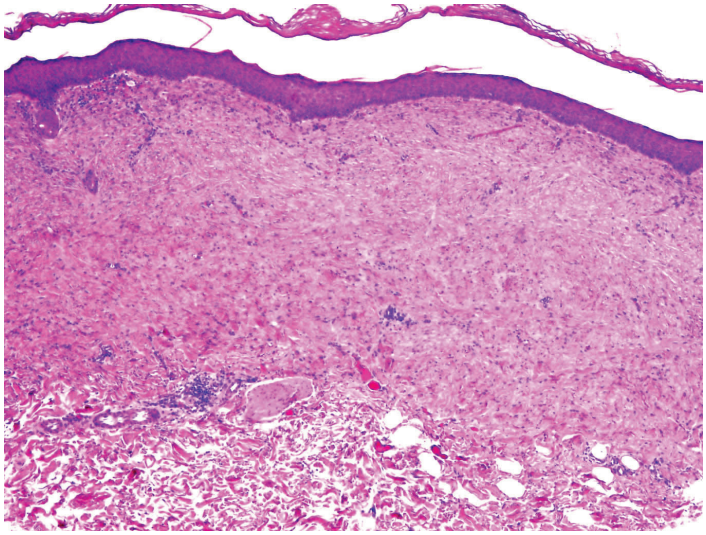

Figure 4. *Higher power view of the superficial dermis. H&E, 10x.*

tacrolimus 0.1% ointment was then utilized, yielding mild improvement.

Case Discussion

Morphea is a localized form of scleroderma that presents with skin pathology alone - there is an absence of involvement of internal organs, nailfold capillary changes, Raynaud phenomenon, or sclerodactyly, which are typically seen in systemic sceroloderma [1]. Morphea manifests clinically with indurated plaques. These lesions typically have a violaceous periphery [6] and occasionally a central white coloration [3]. On histologic examination, classic morphea will manifest with sclerosis and thickening of collagen bundles of predominately the deep reticular dermis but involvement of the upper portions of the reticular dermis as well as the papillary

dermis has also been described [5]. Evidence of linear elastic fibers and loss of CD34-positive dendrocytes is also seen throughout the majority of the dermis with the deeper dermis more heavily involved [6]. Morphea typically has a female predominance with 3:1 female:male ratio with an incidence of approximately 0.4 to 2.7 per 100,000 individuals [1, 3]. There are various subtypes of morphea which include bullous, generalized, guttate, keloidal, linear, plaque, segmental, and the lesser-known superficial form.

Superficial morphea, typically has a clinical presentation consisting of hypopigmented to hyperpigmented patches and plaques that characteristically involve intertriginous sites symmetrically and lack significant induration, contractures, or atrophy [3, 6]. To date there has been no report of any systemic signs and/or symptoms resulting from this morphea sub-type [6]. There has been only one case report of superficial morphea occurring in men [8] with the remaining cases occurring in women [3]. On microscopic examination, skin lesions indicative of superficial morphea will demonstrate sclerosis in, and around the superficial reticular dermis [6]. Other characteristic findings on histology include thickened collagen bundles, decreased fibroblasts and skin appendages, infiltration of lymphocytes in and around vasculature and glands, and reduced or absent staining for CD34 in dermal dendrocytes in the upper portions of the reticular dermis [3, 6]. Parallel elastic fibers seen in the superficial dermis (which may be demonstrated by positive-elastin stains) are distinctive for superficial morphea as these elastic fibers typically organize in a haphazard and disordered fashion throughout the entire dermis [3, 6]. Normal skin structure should be intact in the deeper portions of the reticular dermis and in the hypodermis.

Dermatologic Look-Alikes

Classic morphea described above should certainly be ruled-out. Two other diagnoses that should be considered in the differential diagnosis include idiopathic atrophoderma of Pasini-Pierini (IAPP) and lichen sclerosus et atrophicus (LSA). IAPP, typically presents in adolescence and early adulthood [4]. Clinical examination can distinguish these from superficial morphea by noting gray-colored plaques with a cliff-drop border predominantly on the trunk [6, 8]. Histologically IAPP demonstrates hyalinization of collagen bundles in the dermis [8]. LSA clinically shows hypo- and de-pigmentation of skin with atrophy but minimal to no induration and often involves the genitals (which is less common in superficial morphea). Histologically, LSA typically is characterized by hyperkeratosis, follicular plugging, epidermal atrophy, thick bundles of collagen, loss of elastic fibers in the upper dermis, and vacuolation of basal keratinocytes in the epidermis [3, 5, 6].

Pathogenesis

The underlying cause of superficial morphea remains unclear. It has been noted that the CD34-positive dendritic cells having potential immunologic properties could play an important role in the pathogenesis of morphea [3, 7]. CD34-positive dendrocytes help regulate collagen synthesis; specifically, with the loss of CD34-positive dendrocytes there is a concordant increase in collagen production [7]. As noted earlier there are decreased numbers of CD34-positive dendrocytes in skin specimens from patients diagnosed with morphea. It has also been reported that an inflammatory response related to microvascular injury contributes to the up-regulation of transforming growth factor beta that subsequently increases collagen production and decreases matrix metalloproteinases resulting in a reduction in collagen destruction [1].

Treatment

Treatments that have been used successfully in classic morphea can be implemented in patients exhibiting the superficial sub-type. These include topical regimens such as potent topical steroids, tacrolimus, imiquimod, and vitamin D analogs. Narrow-band UVB (nbUVB), broad-band UVA (bbUVA), UVA1, and psoralen plus UVA (PUVA) have all been used successfully in the treatment of classic morphea. It should be noted that although UVA1 phototherapy has been shown to be a beneficial treatment, patients require frequent and long exposure times for effectiveness. More importantly, UVA1 is not widely available. Systemic therapies include methotrexate in conjunction with high dose steroids. Mycophenolate mofetil can be added to other systemic therapies for recalcitrant cases [2].

Conclusion

This case serves to raise awareness of superficial morphea, a lesser-known variant of morphea. Appropriate diagnosis requires astute clinical and histopathologic diagnosis. There is currently no uniformly effective and easily accessible treatment available for this frustrating condition

References

1. Fett N, Werth VP. Update on morphea: part I. epidemiology, clinical presentation, and pathogenesis. J Am Acad Dermatol 2011; 64: 217-228. [PMID: 21238823]
2. Fett N, Werth VP. Update on morphea: part II. Outcome measures and treatment. J Am Acad Dermatol 2011; 64: 231-242. [PMID: 21238824]
3. Jacobson L, Palazij R, Jaworsky C. Superficial morphea. J Am Acad Dermatol 2003; 49 (2); 323-325. [PMID: 24048434]
4. Jaworsky C. Is superficial morphea synonymous with atrophoderma Pasini-Pierini?: Reply. J Am Acad Dermatol 2004; 50(6): 980. [PMID: 12894089]
5. McNiff JM, Glusac EJ, Lazova RZ, Carroll CB. Morphea limited to the superficial reticular dermis: an underrecognized histologic phenomenon. Am J Dermatopathol 1999; 21(4): 315-319. [PMID: 10446770]
6. Saleh Z, Arayssi T, Saleh Z, Ghosen S. Superficial morphea: 20-year follow up in a patient with concomitant psoriasis vulgaris. J Cutan Pathol 2009; 36: 1105-1108.[PMID: 19602063]
7. Skobieranda K, Helm KF. Decreased expression of the human progenitor cell antigen (CD34) in morphea. Am J Dermatopathol 1995; 17(5): 471-475. [PMID: 8599452]
8. Srinivasan SK, DiMaio D. Superficial morphea in a man. J Am Acad Dermatol 2004; 51: S156-S158. [PMID: 15577758]