Unilateral Multisegmental Morphea

Kristy F. Fleming, MD; Jashin J. Wu, MD; Senait W. Dyson, MD; Arline M. Tsuchiya, MD

There are 5 subtypes of morphea that are based on disease distribution and presentation, including plaque, localized, generalized, linear, and deep morphea. We report a case of a young patient with morphea lesions in scattered locations confined to 1 side of the body, which we have termed unilateral multisegmental morphea.

Cutis. 2011;88:41-42.

Morphea, also known as localized scleroderma, is a rare disease of unknown etiology diagnosed by clinical findings and histopathologic features.[1] Morphea is a type of cutaneous sclerosis characterized by early erythematous or violaceous lesions that later become ivory-colored, hardened skin. Early lesions typically contain an inflammatory infiltrate, either perivascular or diffuse, and later fibrotic lesions are characterized by dense hyalinized collagen bundles extending from the superficial structures into the deep dermis.[1] Morphea can present in a number of different forms. We report a case of unilateral multisegmental morphea.

Case Report

A 7-year-old boy presented with reticulated whorled hyperpigmentation and indurated plaques in scattered locations (ie, arm, back, chest, abdomen, leg) on the left side of his body of 2 years' duration (Figure 1). No lesions or cutaneous abnormalities were noted on the right side of his body. He reported no systemic symptoms and had no family history of skin disorders. Biopsy results of the lesions showed moderately dense, hyalinized collagen encasing the skin adnexa and focally extending into the subcutaneous fat as well as a sparse perivascular inflammatory infiltrate containing numerous plasma cells, consistent with morphea (Figure 2). Laboratory workup was unremarkable.

Comment

Morphea is classified into the following 5 subgroups based on clinical morphologic findings: plaque, localized, generalized, linear, and deep.[2] Plaque morphea

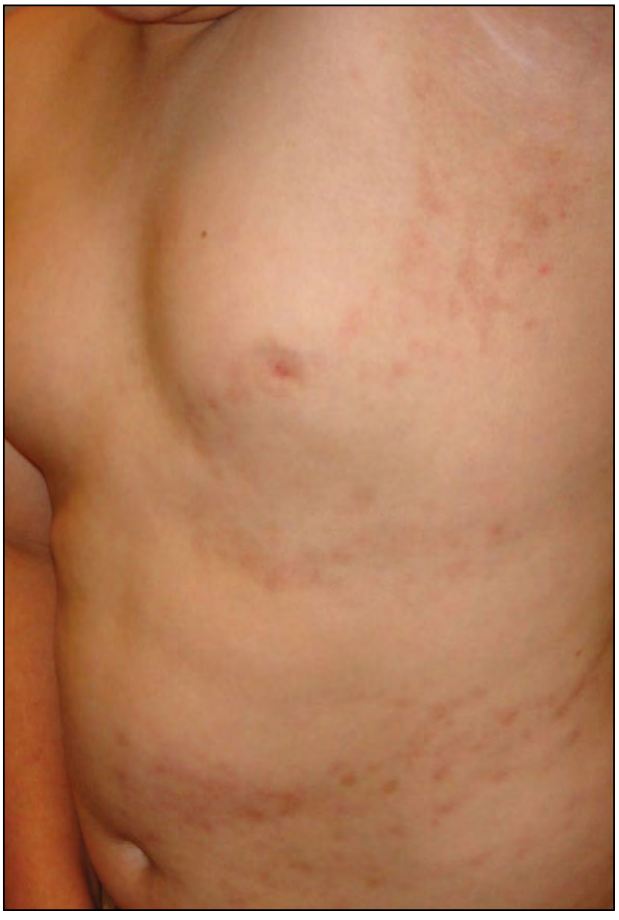

Figure 1. Scattered, indurated, hyperpigmented plaques on the left side of the abdomen.

Dr. Fleming is from the Department of Dermatology, Baylor College of Medicine, Houston, Texas. Dr. Wu is from the Department of Dermatology, Kaiser Permanente Los Angeles Medical Center, California. Dr. Dyson is from Specialists in Dermatology, PLLC, Tucson, Arizona. Dr. Tsuchiya is from the Department of Dermatology, Kaiser Permanente Orange County Medical Center, Anaheim, California.

The authors report no conflict of interest.

Correspondence: Jashin J. Wu, MD, Kaiser Permanente Los Angeles Medical Center, 1515 N Vermont Ave, 5th Floor, Los Angeles, CA 90027 (jashinwu@hotmail.com).

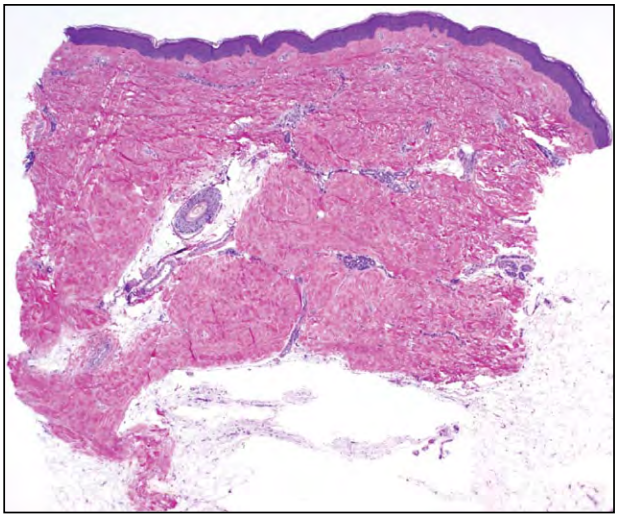

Figure 2. Hyalinized collagen encasing the skin adnexa with extension into the subcutaneous fat as well as a perivascular inflammatory infiltrate with numerous plasma cells (H&E, original magnification ×2).

is the most common form and typically presents with 1 to a few localized lesions. A number of variants of plaque morphea have been described, including guttate, keloidal (nodular), and atrophoderma of Pasini and Pierini.[2]

Plaque morphea may evolve into generalized morphea, which usually is insidious in onset and is defined by the coalescence of previously distinct plaques or the occurrence of multiple lesions on more than 2 anatomical sites.[2] Generalized morphea is a potentially severe form that may involve underlying muscles, resulting in contractures and atrophy.

Linear morphea is the most common form in children and adolescents.[3] It is characterized by linear areas of induration that may extend from the dermis and subcutaneous tissue all the way into the muscle and underlying bone, sometimes resulting in limb contractures. Lesions in linear morphea typically are unilateral.[3] En coup de sabre and progressive facial hemiatrophy (Parry-Romberg syndrome) are recognized variants of linear morphea. Two case reports have been published describing 5 cases of unilateral generalized morphea (UGM) in children.[4,5] These cases identify UGM as a new variant of the linear form of morphea. Our patient presented with a variation of UGM, which we have termed *unilateral multisegmental morphea*. Unlike the typical characteristics of UGM, our patient presented with areas of obvious sparing on the affected side. Recognition of this unique presentation will undoubtedly be beneficial for clinical diagnosis of this subtype of linear morphea.

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