Unilateral generalized morphea: A case report and literature review

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Morphea is an autoimmune connective tissue disease of the dermis and subcutaneous fat characterized by sclerosis due to excessive collagen deposition. Morphea is classified into different subtypes based on clinical and histological characteristics of cutaneous lesions. An extremely rare subtype of morphea is termed unilateral generalized morphea (UGM). UGM is characterized by indurated plaques on dermatomes of the trunk or back with a sharp midline demarcation line. Ipsilateral limbs may also be affected. There is still debate on whether this type of morphea should be considered as a distinct type or a presentation of linear morphea. Here we report the case of an 8-year-old boy with this type of morphea.

Keywords: morphea, dermatomal, unilateral generalized morphea

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INTRODUCTION

Morphea, also known as localized scleroderma, is an autoimmune connective tissue disease of the dermis and subcutaneous fat characterized by sclerosis due to excessive collagen deposition. Morphea is classified into circumscribed, generalized, linear, and pansclerotic or deep subtypes. This classification is based on clinical presentation and depth of tissue collagen deposition [1,2]. Unilateral generalized morphea (UGM) is a rare subtype of morphea reported in the literature. Here we report a case of this subtype of morphea.

CASE REPORT

An 8-year-old boy was admitted to our clinic with a 3-month history of progressive skin stiffness on the left side of his trunk which had spread to his limbs. On physical examination, sharply demarcated diffuse sclerosis and hyperpigmentation was found on the anterior left side of his trunk along with the posterior of his left leg (Figures 1, 2). The lesions on the trunk had a sharp midline demarcation. There were no similar cutaneous lesions detected on the opposite side of his trunk or limbs. He had no complaints of gastroesophageal reflux, dyspnea, or vascular complaints as seen in Raynaud's phenomenon. On examination, there was no evidence of periungual telangiectasia or cutaneous calcinosis. There was no history of similar lesions in any first or second degree family members of the patient. An incisional biopsy was taken from the indurated plaques on the trunk. Histological examination revealed hyalinization of thick and closely packed collagen bundles in the reticular dermis and subcutis. Eccrine glands

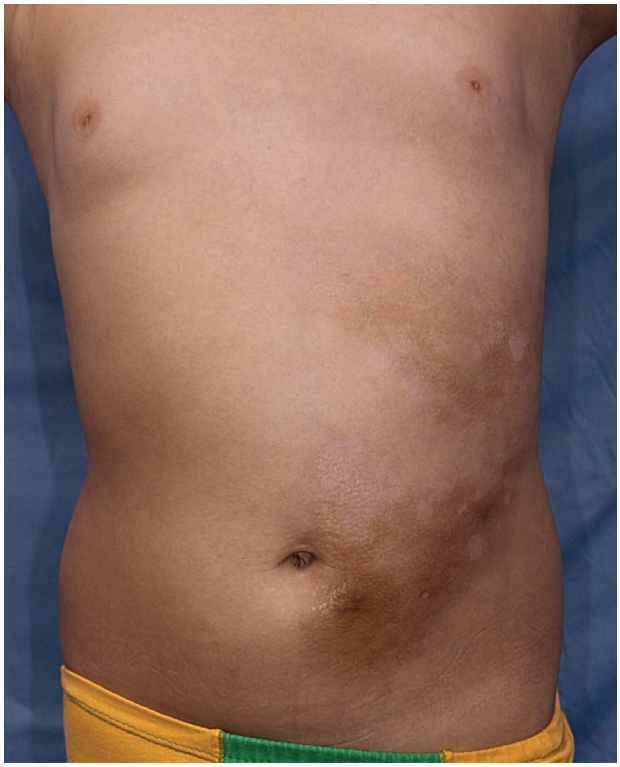

Figure 1. Brown indurated plaques on the left side of the trunk with sharp midline demarcation.

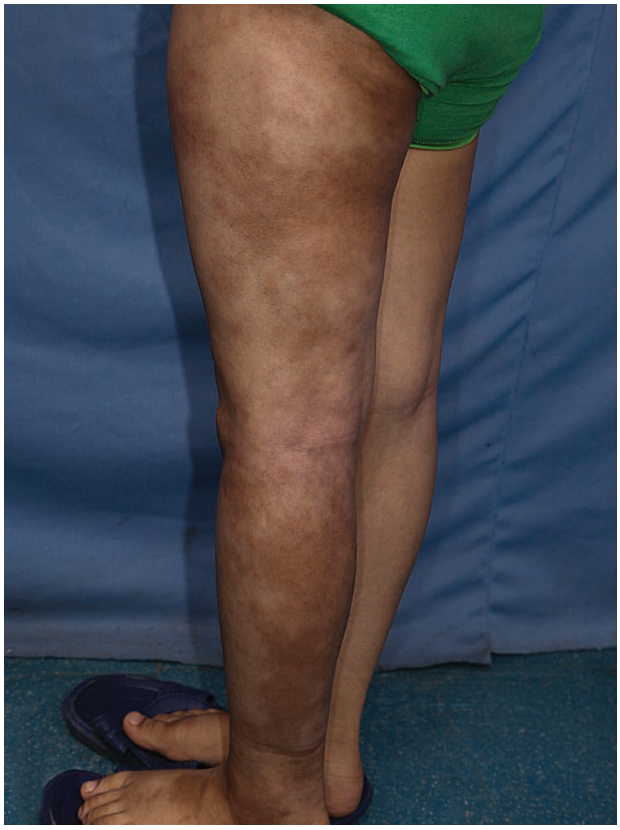

Figure 2. Linear indurated morphea band along the left lower extremity.

were atrophic and entrapped within thickened collagen bundles. Sparse inflammatory lymphocytes infiltrated the junction between the reticular dermis and subcutaneous fat (Figure 3). These histopathological findings pointed towards a diagnosis of morphea.

Laboratory studies showed normal complete blood count (CBC). Anti-nuclear antibody (ANA) was negative at 0.5 IU (cut-off: 10 IU). Anti-double strand DNA antibody was also negative at 11 IU (cut-off: 35 IU). Anti-histone antibody was 5.9 IU, which was also negative. Other laboratory tests such as viral markers, ESR, CRP, and a PPD test were within their normal ranges.

Considering the diagnosis of morphea, the patient received prednisolone 10 mg/day and methotrexate 15 mg/week. Sclerosis of the plaques resolved by the end of the second month of treatment. The prednisolone was tapered and discontinued by the end of the second month and after two months the dose of methotrexate was reduced and the patient has remained on methotrexate 7.5 mg/week at the time of this report.

DISCUSSION

Linear morphea is a rare subtype of morphea. Linear morphea presents as discrete, indurated linear bands that are most often unilateral and single. The most common site of involvement is the lower extremities. The lesions usually follow the Blaschko lines [3]. Blaschko lines represent a pathway of embryonic cell migration during development

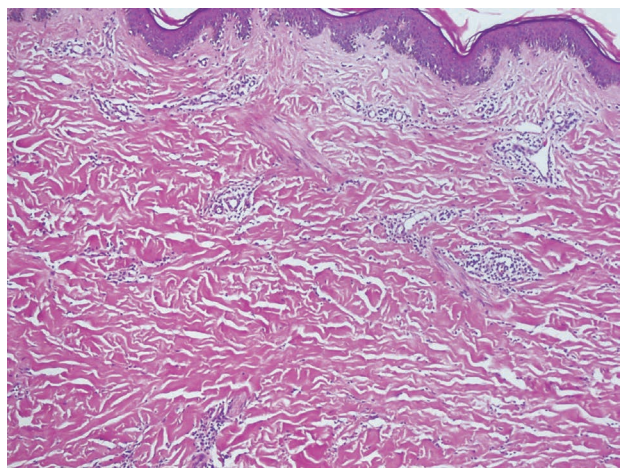

Figure 3. Dermis is thickened by dense collagen bundles and sparse perivascular lymphocytic infiltration. H&E staining, magnification: 10×.

of the skin. Many acquired and congenital skin disorders such as epidermal nevi, incontinentia pigmenti, and linear lichen planus are distributed along these lines and may be a consequence of a genetic mosaic [4,5].

In 1976, it was suggested that linear morphea follows Blaschko lines [6]. Although, in 1984, the same author has reported linear morphea to be dermatomal [7]. Dermatomes are skin segments determined by sensory innervations of the skin. Distribution of skin dermatomes differ from Blaschko lines [6]. However, since the arrangement of Blaschko lines can be similar to dermatomal distributions, especially in the extremities which are the most common site for linear morphea, the distinction between the two distributions has been controversial [4,8]. The first case of frontoparietal scleroderma (en coup de sabre) was described in 1998 [9] as a form of linear morphea; since then, lesions with linear or dermatomal distribution have been classified as the linear subtype of morphea.

In 2002, a 6-year-old boy presented with large brown indurated plaques with sharp midline demarcation on the right side of his chest and back. The boy also suffered from sclerosis in his right leg. The authors termed this subtype of morphea as UGM [10]. To best of our knowledge 8 patients with similar distributions of skin sclerosis have been reported and classified as UGM (Table 1).

In our case, the morphea lesions have shown the same distribution. In UGM, sclerotic plaques on the trunk often have a sharp midline demarcation, which is more in favor of a dermatomal distribution. This type of morphea is considered to be a rare variant of linear morphea.

Our case had a negative ANA test, which was similar to 2 previously reported cases [11-14]. Similar to linear morphea, most reported cases of UGM had a positive ANA test.

In conclusion, it seems necessary to better recognize the demographic, clinical, and histopathologic characteristics of this type of morphoeic lesion. It is somewhat debatable as to which subtype of morphea UGM should be classified or whether it should be considered a distinct subtype of morphea.

Table 1. Characteristics of unilateral generalized morphea (UGM) cases in the literature.

Author (year)	Age (years)/ sex	Family history of sclerosing disease	Location	Laboratory studies with positive results	Treatment
Nagai et al. [10] (2002)	6/m*	None	Right side of the trunk, right leg and arm	ANA† (1/320), anti-ssDNA+ (99 IU/ml §)	Topical steroid
Appelhans et al.[11] (2006)	14/f*	Systemic sclerosis in first degree relative	Right side of the trunk, right leg and arm	ANA (1/1250), Anti-histone H1,H3 antibody, RF ¥ (800 IU/ml), anti-fibrillin antibody	Prednisolone 5 mg/day, methotrexate 15 mg/week, low dose UVA1
	23/f	None	Right cheek, right extremities, right side of the trunk	ANA (1/1250), anti-smooth muscle antibody (ASMA)	Chloroquine, prednisolone, UVA1 therapy, PCMT
	38/f	None	Right side of the trunk, right extremities	ANA (1/2560), AMA ‡ (1/80), RF (800 IU/ml), anti-histone H2A,H2B, H3, H4, human type III procollagen	PCMT, low dose UVA1
	20/f	None	Left side of trunk, left limbs	ANA (1/1280), ASMA, antibody against extractable nuclear antigens	PCMT
Gerceker Turk et al. [12] (2009)	25/m	None	Left side of face, neck, trunk, left limbs	ANA (1/320)	Prednisolone, methotrexate
Ozturk [13] (2011)	14/m	None	Left side of posterior trunk, left shoulder, left arm left leg, left foot	None	Topical steroid, intralesional steroid (3 mg/ml)
Rodriguez et al. [15] (2011)	7/m	None	Right side of trunk, left limbs	None	—
Lestari et al. [16] (2016)	12/f	None	Right side of trunk, right limbs	ANA (1/320), polyclonal increase in gamma globulins, anti-histone	Methyl prednisolone pulse therapy (750 mg for 3 days), methotrexate 15 mg/week, physiotherapy, prednisolone 10 mg/day

*F: Female; M: Male; †ANA: Anti-nuclear antibody; +anti-ssDNA: Anti-single strand DNA; §IU/ml: International unit per milliliter, ¥RF: Rheumatoid factor; ‡AMA: Anti-mitochondrial antibody

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