CORRESPONDENCE

Coexistence of morphea and lichen sclerosus et atrophicus in a zosteriform pattern

Dear Editor,

A 23-year-old female presented with a 3-month history of an asymptomatic, slowly expanding area of dyschromia with textural changes over the left side of her face and neck. Physical examination showed a band of hyperpigmented, indurated plaque that ran from the left anterior to the posterior aspect of her neck (Figure 1). A slightly hypopigmented patch was aligned, in parallel to the plaque, on the adjacent upper portion of her neck. A small part of her lower left cheek and postauricular area were also involved. A skin biopsy was obtained from the junction of the hyper- and hypopigmented areas on her neck. Under low-power magnification, both areas showed the presence of thickened collagen bundles with entrapped, infiltrated eccrine glands (Figure 2A). There was epidermal hyperkeratosis with follicular plugging. The papillary dermis showed rather homogeneous changes with pigment incontinence, suggesting a previous interface inflammatory process (Figure 2B). An orcein stain indicated the loss of elastic fibers in the superficial dermis (Figure 2C and D). These findings were compatible with morphea showing superficial changes reminiscent of lichen sclerosus et atrophicus (LSA).

Laboratory tests scored positive for speckled-type antinuclear antibodies at a 1:80 dilution, and anti-Ro antibodies at a 1:240 dilution. Her genital area was normal in appearance. There was no evidence of sicca syndrome, Raynaud's phenomenon, or any symptoms suggestive of systemic scleroderma. A complete blood cell count, biochemistry profile, and urinalysis were all unremarkable for the patient. On the basis of the clinical and laboratory examination, we inferred that the patient had coexistence of morphea and extragenital LSA. She received treatment with hydroxychloroquine (200 mg/day) and topical steroid of clobetasol propionate. Follow-up, the color and induration of the lesions gradually improved without involving any other organs.

Morphea and LSA are two cutaneous disorders with different clinical and pathologic presentations. The relationship between the two diseases has been extensively debated for decades, and frequent reports of an association between the two conditions have led some clinicians to hypothesize the existence of a common pathologic link. Wallace1 studied 380 LSA patients and reported that histologically-confirmed morphea was present in 13 of these patients. Uitto et al2 were the first to describe a series of 10 patients in which the two entities coexisted. In seven of the 10 cases, biopsies indicated that features of both morphea and LSA presented with equal intensity. Subsequently, sporadic cases of these two coexisting diseases have been published. Most recently, Peterson et al3 proposed a revised classification of morphea to include LSA as a subtype owing to the histologic similarities between the two conditions. However, many investigators have argued that coexistence of morphea and LSA is coincidental and insisted that there is sufficient clinical and histologic differences between them. Rahbari4 developed a discriminating method using elastic-tissue staining and reported that elastic fibers in the upper dermis are lost in LSA, but not in morphea. With the use of laser scanning confocal microscopy, Kowalewski et al5 proposed that the alteration of the basement membrane zone in morphea was different from that in LSA. The former preserved continuity of structures in basement membrane zone, which was lost in the latter.

Our case presented simultaneous occurrence of both types of clinical manifestation, which proved to be deep morphea and superficial LSA by histopathology. In the study by Uitto et al,2 repeated biopsies were taken from patients at the same location at varying time points. In one case, a transition from LSA into morphea was found over a period of 2 years, with complete disappearance of LSA eventually. Schaffer et al6 described six patients with both LSA and morphea as manifestations of sclerodermoid chronic graft-versus-host disease. In one of the six patients, the morphea-like plaques underwent a transition to whitish, superficial LSA-like lesions before resolving. Taken together, we suggest that an evolutionary relationship exists between morphea and LSA.

Another interesting observation is that the pattern of the patient’s skin lesion corresponded to dermatomal distribution. Linear morphea is a well-known subtype of localized scleroderma. Linear extragenital LSA has been reported in one case with a bilateral zosteriform pattern7 and in six cases following Blaschko’s lines.8 Our patient did not have any history of wounds, trauma, or herpes zoster on her neck. Therefore, Koebner phenomenon and an isotopic response were excluded. Furthermore, cutaneous mosaicism in different cell types could result in the characteristic dermatomal distribution and additional studies will be required to elucidate whether our case represents mosaicism.

In conclusion, we report a patient with coexisting morphea and LSA in a zosteriform pattern exhibiting positive treatment response to plaquenil and topical steroid. As unilateral dermatomally-distributed LSA is clinically seldom seen, cases of dermatomal LSA with simultaneous morphea are extremely rare. Whether the unilateral zosteriform distribution pattern of the two conditions is related to mosaicism will require further study.

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References


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