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Lichen sclerosus et atrophicus, bullous morphea, and systemic lupus erythematosus: a case report

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Lichen sclerosus et atrophicus (LSA) rarely coexists with morphea, especially when bullae occur in lesions of morphea. Here we report the case of a 15-year-old girl with this condition, who also fulfilled four out of 11 diagnostic criteria for systemic lupus erythematosus (SLE). Tissue biopsy of different skin lesions showed LSA in the regions of bullous morphea, that has rarely been reported in the literature.

Key words: Lichen sclerosus et atrophicus (LSA), bullous morphea, systemic lupus erythematosus (SLE)

Lichen sclerosus et atrophicus (LSA) is an uncommon disease of unknown etiology. It occurs most often in the anogenital region of perimenopausal women and is rarely noted in children. Warrington *et al* reported a high rate of coexisting sexual abuse in children with LSA [1]. LSA is difficult to clinically differentiate from morphea, a situation much discussed in the literature [2-9]. The occurrence of bullae in lesions of morphea is rare, with only few cases having been previously reported [10-14]. Here we describe a case of LSA with bullous morphea in a patient with systemic lupus erythematosus (SLE).

Case Report

A 15-year-old girl was hospitalized due to exertional dyspnea and chest discomfort over a 4-day period. She had visited another hospital twice during the previous 3 years due to progressive development of a skin lesion with recurrent induration and crust. The lesion had initially developed on the forearm and then spread to the trunk, lower extremities and face. About 1 year prior to this admission, mild dysphagia and exertional dyspnea had developed. Initial examination at another medical center had led to suspicion of scleroderma. Pathology of skin biopsy from lower back excision was reportedly compatible with bullous morphea at that time.

Physical examination at admission revealed hypopigmentation and sclerotic change of the facial skin, especially over the cheeks and nose. The skin lesions were variously sized, hyperkeratotic, and crusted with brownish plaques, a few with a bullous base, and extended to the trunk and extremities (Fig. 1). The skin was dry, with a fish-scales appearance on the extremities, especially on the lower legs. Friction rub and grade II systolic murmur were heard upon auscultation, and electrocardiography showed sinus tachycardia with low voltage. About 230 mL serosanguinous fluid with fibrin was drained out after demonstration of pericardial effusion by cardiac echography. The drained effusion was exudative in nature, with lymphocytes predominating. Symptoms of exertion dyspnea improved a great deal after pericardiocentesis. The following laboratory results showed: white blood cell count, 3650/mm^3; hemoglobulin, 9.1 g/dL; platelet count, 642000/mm^3; erythrocyte sedimentation rate, 112/h; IgA, 435 mg/dL, IgM, 772 mg/dL, IgG, 3080 mg/dL; IgE, 1456 kU/L; antinuclear antibody (ANA) titer, 1:5120(+), homogenous pattern; anti-double-stranded DNA antibodies, 41.25 IU/mL; C3, 79.9 mg/dL; C4, 26.5 mg/dL (normal value: anti-dsDNA antibody < 7 IU/mL; C3, 138.7 ± 20.7 mg%; C4, 40.2 ± 12.7 mg%); positive direct Coombs' test but negative indirect test; rheumatoid factor, 1:20480(+); anti-ENA (including SM, RNP, SSA, SSB, SCL-70) antibody, antimicrosomal antibody, anticardiolipin antibody, antiphospholipid antibody, lupus anticoagulant, and VDRL/RPR test were all negative. Raynaud's phenomenon was absent and the lymphocyte subset percentage was normal. She complained of dry eye sensation but the results of Schirmer's test was in the

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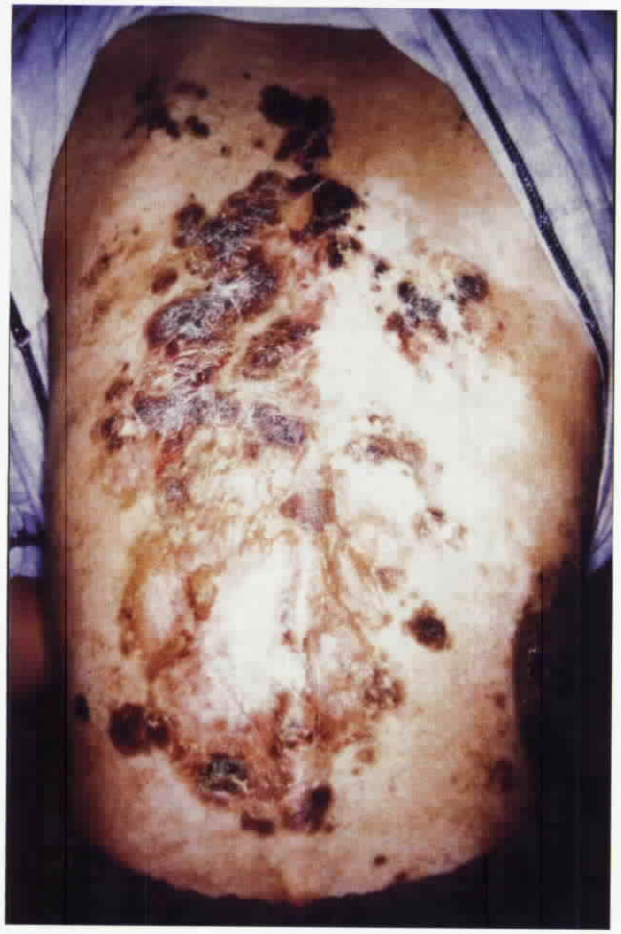

Fig. 1. Many variously sized, hyperkeratotic, and crusted brownish plaques, a few with a bullous base, were noted on the trunk.

normal range. She had mild symptoms of dysphagia, esophagogram and upper G-I series revealed antral gastritis and poor peristalsis of the duodenum and jejunum under fluoroscopy. Lung function test showed severe restrictive ventilatory defect, and moderate impairment of diffusing capacity.

Pathology reports of skin incisional biopsies from the scalp and flank areas showed LSA (Fig. 2). Pathologic findings included atrophy of the epidermis with hyperkeratosis, focal keratin plugs and basal vacuolation, severe edema in the papillary dermis, and sclerosis in the reticular dermis. However, skin incisional biopsies from the left upper arm and scapula area were compatible with morphea (Fig. 3), with findings of atrophy of the epidermis, and diffuse sclerosis in the superficial and deep dermis.

She was treated with oral azathioprine, application of prednisolone and naproxen and a mixture of hydrocortisone ointment and urea cream to the skin lesions. After a 4-month follow-up, her skin condition

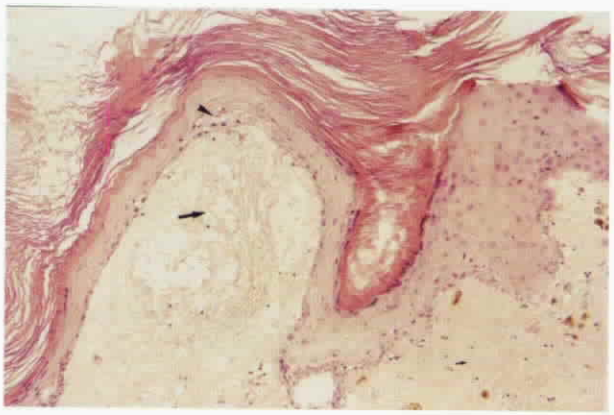

Fig. 2. Excisional biopsy from flank skin shows atrophy of the epidermis with hyperkeratosis, focal keratin plugs and basal vacuolation (arrow head). The dermis reveals severe edema in papillary dermis (arrow) and sclerosis in reticular dermis (small arrow), (H & E stain, 100 x).

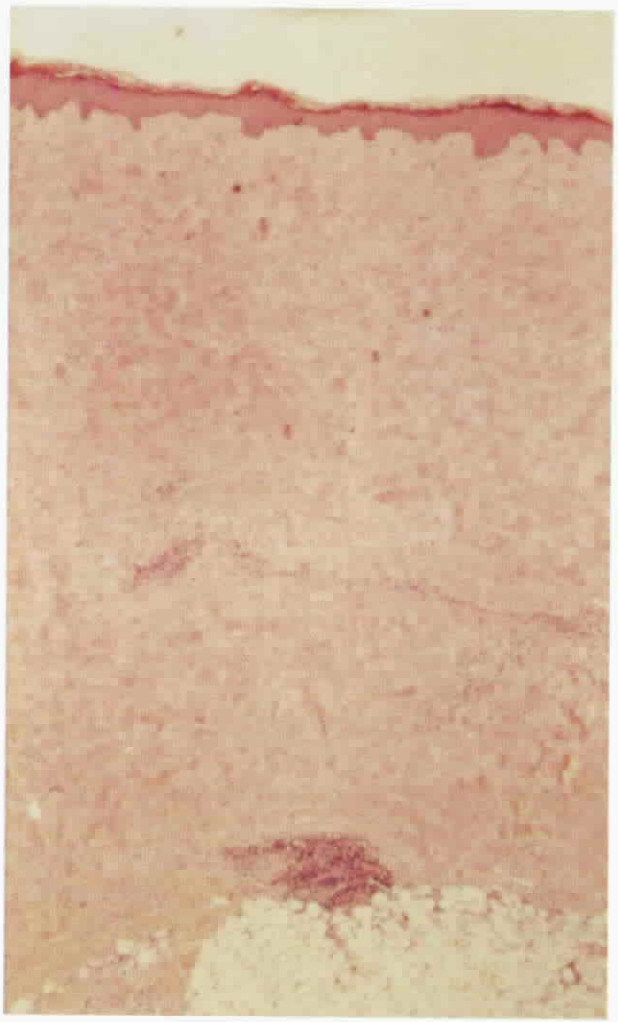

Fig. 3. Excisional biopsy from left upper arm shows relative atrophy of the epidermis with diffuse sclerosis in the superficial and deep dermis. Mild mononeuclear infiltration in the dermal-subcutaneous junction is also present, (H & E stain, 40 x).

had improved greatly and no systemic organ involvement was found.

Discussion

Bullae are rare in the lesions of scleroderma. The pathogenesis of bullae in morphea and whether bullous morphea should be included in the spectrum of clinical variants of morphea are controversial. The mechanism of bullae development on a sclerotic area of skin remains a missing link in understanding the etiology of bullae formation. Many different theories have been proposed to explain blister formation in morphea. Templeton [10] first reported bullous morphea in 1941, and suggested that bullous lesions in scleroderma were caused by lymphatic obstruction. Carb and Sims found 25 cases of bullous scleroderma in the world literature upon reviewing their case report in 1959 [11]. Tuffanelli described three patients with systemic scleroderma and vesicles and bullae in which dilated lymphatic channels were observed in the upper dermis, providing support for the lymphatic obstruction mechanism proposed by Templeton [12]. However, other authors have postulated that bullae may have been caused by trauma, "nerve factors," or vascular occlusion with enzymatic degradation of basement membrane.

Coexistence of LSA and morphea has been infrequently reported. Some authors have considered LSA and morphea to be different manifestations of a single disease spectrum [15,16], while others have thought that the association of LSA and morphea is one of the clues to a close relationship between two separate diseases [7]. Many authors have described coexistent LSA and morphea at all ages and in both sexes [3,5, 17]. The coexistence of morphea and LSA in the same biopsy specimen has been described [7,15]. In addition, several investigators have reported transition from LSA to morphea [7], or vice versa [16] using sequential biopsies. Borda *et al* (1968) considered LSA as a scleroderma of the papillary dermis and classified LSA as a subset of the greater body of scleroderma and "pseudo-scleroderma" disease [2]. Others have viewed LSA and morphea as manifestations of the same disease, with morphea having a greater association with systemic disease [8,9]. Acrodermatitis chronica atrophicans, well established as a spirochetal disease, is often considered the bridge between LSA and morphea, because it shows a degree of clinical and histological overlap [18,19]. On the contrary, early in 1910, Ormsby suggested that LSA should not be confused with sclerosis [20]. Patterson and Ackerman noted that many cases diagnosed clinically and histologically as LSA are actually morphea, regardless of how consistent with LSA the more superficial histological characteristics may be [21]. They suggested that the lack of significant infiltrate and minimal vacuolar interface changes in many of the cases in their series support the diagnosis of morphea.

The present case fulfilled four out of the 11 diagnostic criteria for SLE including neutropenia, serositis, elevated expression of anti-double-stranded DNA antibody and elevated ANA titer. Moreover, although the gross picture favored the diagnosis of scleroderma, skin biopsies taken from different locations showed bullae morphea in associated LSA. Kahana *et al* had reported a 25-year-old woman with a diagnosis of SLE, who had previously developed LSA on the elbow [22]. Although others have viewed this association as being two separate events [4], the concomitant presence of LSA and bullae morphea with SLE may not be merely coincidental, since SLE has a well-established autoimmune pathogenesis, and organ-specific autoantibodies have also been described in the literature [23,24]. Further study would be needed to elucidate this relationship.

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