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CASE REPORT

Scleroderma-Like Lupus Panniculitis: A Case Report and Literature Review

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Abstract: Sclerodermic or scleroderma-like lupus erythematosus panniculitis (SLEP) shares both clinical and histopathological features between lupus panniculitis and localized scleroderma. It is exceedingly rare. We herein report a case of SLEP manifested with a solitary, firm-to-hard, erythematous plaque in an Asian woman. This patient responded well to intralesional corticosteroid and antimalarials. We have reviewed the pathogenesis of fibrosis in patients with chronic cutaneous lupus erythematosus as well as documented cases of SLEP in the literature.

Keywords: cutaneous lupus erythematosus, scleroderma-like lupus erythematosus panniculitis, sclerodermic lupus panniculitis, localized scleroderma, lupus profundus, overlap syndrome

Introduction

Lupus erythematosus panniculitis (LEP) is a chronic inflammation of subcutaneous tissue affecting 1–3% of patients with chronic cutaneous lupus erythematosus (CCLE).[1] While it usually occurs independently, coexistence with discoid lupus erythematosus (DLE) occurs in approximately one-third of patients.[2–4] LEP typically manifests with tender subcutaneous nodules or plaques on the face, proximal extremities, and trunk.[1] It displays a female predilection with a female-to-male ratio ranging from 2:1 to 9:1, with onset in third to fifth decades, and a slightly younger onset in the Asian population.[1,3] Unique presentations, such as linear configuration or coexistence with localized scleroderma are exceedingly rare. The term "sclerodermic or scleroderma-like lupus erythematosus panniculitis (SLEP)" was used for 5 patients with overlap clinical and histopathologic features between lupus panniculitis and localized scleroderma.[5–8] Four of them displayed a linear configuration, and one was described concomitantly with sclerodermic DLE.[5,6,8] We hereby present a case of SLEP presented as a solitary, firm to hard erythematous plaque mimicking dermatofibrosarcoma protuberans in a middle-aged Asian woman.

Case Report

A 43-year-old Thai female presented with a solitary indurated erythematous hard plaque at the left lateral neck. The lesion was mildly pruritic, painless, and gradually expanded over the course of 15 months. She had no underlying diseases and denied family history of malignancy or other autoimmune connective tissue diseases. Symptoms of fever, weight loss, joint pain, hair loss, oral ulcer, or photosensitivity were not present. There was no history of trauma preceding the lesion. She also had no history of smoking or alcohol consumption.

Dermatological examination revealed a solitary indurated erythematous plaque with firm to hard consistency on the left lateral side of the neck (Figure 1A), and a faint erythematous plaque with telangiectasia at the left lateral nose. Other examinations including superficial lymph nodes, mucous membranes, scalp, hair, and nails were unremarkable. The initial diagnosis was dermatofibrosarcoma protuberans (DFSP). Other differential diagnoses were cutaneous pseudolymphoma, cutaneous Rosai-Dorfman disease, and lupus tumidus.

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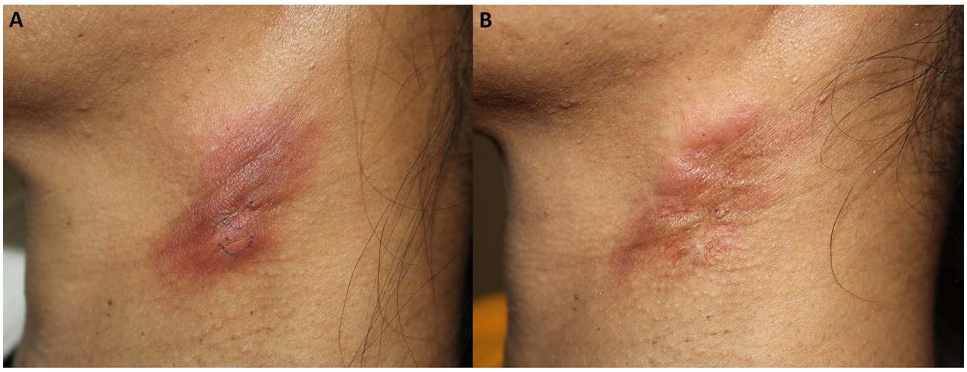

Figure 1 Clinical features and treatment response of SLEP. (**A**) A solitary firm to hard erythematous plaque mimicking dermatofibrosarcoma protuberans on the left lateral neck. (**B**) Partial improvement with indentation after treatment with intralesional steroid and hydroxychloroquine (200 mg/day).

Laboratory data showed hemoglobin level of 11.4 g/dL, white blood cell count of 5300/mm^3 composed of 59% neutrophils, 34% lymphocytes, and 5% monocytes, and platelet count of 262,000/mm^3. Liver function test revealed total and direct bilirubin levels of 0.3 mg/dL and 0.1 mg/dL, respectively, aspartate aminotransferase 20 IU/L, alanine aminotransferase 16 IU/L, alkaline phosphatase 82 IU/L, and gamma-glutamyl transferase 16 IU/L. Renal function test showed blood urea nitrogen of 14 mg/dL and creatinine of 0.84 mg/dL. Urinalysis was within normal limits. Antinuclear antibody titer was 1:80 with coarse speckle pattern.

After obtaining informed consent, a punch biopsy from the lesion at the neck was performed. Histopathology revealed normal epidermis, dense superficial and deep perivascular and periadnexal lymphoplasmacytic infiltrates with lobular lymphocytic panniculitis. In addition, thickened homogenized collagen in the lower dermis extending to the broadened fibrous septum and loss of adipocytes around eccrine structures were noted (Figure 2). These findings shared histopathologic findings among lupus panniculitis and scleroderma.

Based on the histopathological features, the diagnosis of SLEP was made. The patient was treated with intralesional corticosteroid along with hydroxychloroquine (200 mg/day). Moderate potency topical steroid was applied twice daily on the lesion at the nose. Strict photoprotection was also advised. At 2-month follow-up, the lesions showed partial improvement with some indentation (Figure 1B). She remained free from signs and symptoms of systemic lupus erythematosus (SLE) after a 2-year follow-up period.

Discussion

Concomitant CCLE and localized scleroderma is a rare overlap syndrome of connective tissue diseases. The first case was described in 1978 by Umbet and Winkelmann with the clinicopathologies of both DLE and scleroderma.[9] Later, in 1994, an unusual presentation of lupus panniculitis coexisting with localized morphea was reported.[7] Sclerodermic or SLEP is an umbrella term proposed by Marzano for these groups of patients.[8] They also share some histopathological findings such as lymphocytic panniculitis, lymphocytic vasculitis, dermal sclerosis, and broadening of fibrous septa.[5]

Due to the rarity of SLEP, the presence of lymphoid follicles with germinal center or dermal sclerosis can mislead to the diagnosis of cutaneous lymphoma or scleroderma. Previous documented cases are summarized in Table 1. Of six patients, half of them required repeated skin biopsy for the diagnosis of SLEP. Of note, all cases apart from ours were reported from Western countries with age of onset varying from 9 to 63 years. The predominant features are the female predilection and upper body distribution. Four of six lesions displayed a linear configuration. Association with SLE was reported in 2 cases.[8] The first developed severe renal involvement 9 years after the onset of lesions, and the second had autoimmune thrombocytopenia 5 years prior to cutaneous presentation and experienced spontaneous abortion a few months before the lesions occurred.[8] Marzano proposed that this rare variant could be a warning sign of impending SLE, and long-term follow-up on systemic symptoms are required.[8] Most of SLEP lesions responded well to antimalarials and resolved with localized lipodystrophy. There was only one pediatric case that had almost complete healing without lipodystrophy.[5] Our case presented with a hard indurated plaque mimicking dermatofibrosarcoma protuberans (DFSP),

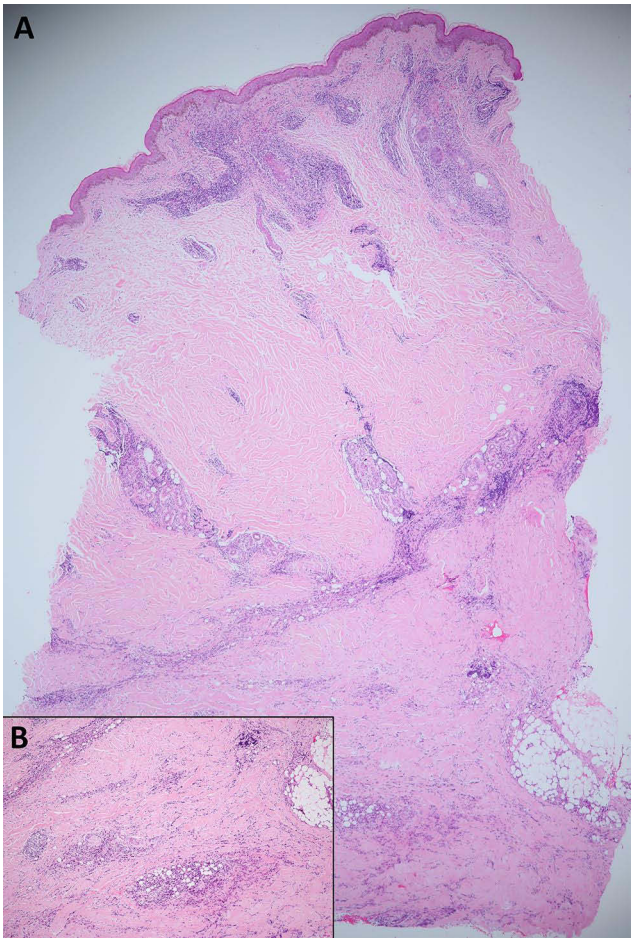

Figure 2 Histopathologic features of SLEP. (**A**) dense superficial and deep perivascular and periadnexal infiltration composed of lymphocytes and plasma cells (H&E, ×40) (**B**) thickened homogenized dermal collagen with lobular panniculitis (inset, H&E, ×100).

which is a unique clinical manifestation compared with previous reports on SLEP. The histopathology shared features of both lupus panniculitis and morphea. The superficial and deep perivascular and periadnexal lymphoplasmacytic infiltration and lobular lymphocytic panniculitis were compatible with lupus panniculitis while the sclerotic dermal collagen extending to the broadened fibrous septum were consistent with morphea. The patient had no signs and symptoms of SLE or systemic sclerosis.

The pathomechanism of fibrosis in lupus panniculitis is still unknown. Recent evidence reveals that active SLE neutrophils extracellular traps (NETs) bearing tissue factor and interleukin-17 had a potential role in promoting fibrotic activity in cultured human skin fibroblasts.[10] The amount of neutrophil producing NETs was significantly higher in scarring lesions such as lupus panniculitis and DLE than the non-scarring subtypes, such as subacute cutaneous lupus erythematosus, suggesting that NETs might be associated with scarring and tissue damage.[11] Current data mostly exhibited possible pathways involving transforming growth factor (TGF)-β to induce fibrosis in CCLE. A microarray study suggested TGF-β-dependent fibrosis formation in DLE.[12] An increase of B lymphocytes in scarring DLE was also proposed for a pathogenic role in fibrosis by upregulating collagen production in a TGF-β1-dependent manner.[13,14] Julia et al reported a case of CCLE coexisting with morphea and speculated that a lichenoid reaction may be a trigger in stimulating abnormalities in fibroblast-dependent reparative mechanisms leading to tissue sclerosis.[15] Further studies on the pathogenesis of SLEP are required.

We report a case of SLEP mimicking DFSP in a middle-aged Thai woman presenting with a slowly growing firm to hard erythematous plaque on the lateral neck. The histopathological findings showed a characteristic pattern of lupus panniculitis as well as thickened homogenized collagen, consistent with morphea. To the best of our knowledge, this is the first report in an

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Table 1 Demographic Data, Clinical, Laboratory, Histopathological, Immunopathological Characteristics, Treatment and Outcome in Scleroderma-Like Lupus Erythematosus Panniculitis Cases

Author, Year	Age/ Sex	Clinical Presentation	Location(s)	Disease Duration	Histopathology	DIF	Systemic Involvement	Laboratory Findings	Treatment and Outcome
Our case	43/ F	Solitary erythematous subcutaneous hard plaque	Lateral aspect of left neck	15 Mo	Vacuolar interface dermatitis, superficial and deep perivascular lymphoplasmacytic infiltrate, lobular panniculitis and thickened homogenized collagen	Not done	No	ANA; positive 1:80 coarse speckled	Intralesional steroid monthly. Hydroxychloroquine 200 mg/day. Improvement with indentation
Stork, 1994[7]	22/ F	Subcutaneous nodules and irregular, indurated plaques with tethered skin	Lateral aspect of right upper arm, breasts, buttocks, and face	2 Y	Normal epidermis, sparse lymphocytic infiltrate, lymphocytic panniculitis with hyalinized fat necrosis, hyalinized collagen around adnexae in the deep dermis and within broadened fibrous septa	Granular deposits of IgM at the DEJ	No	ANA; negative Anti-dsDNA; negative ESR; normal C3 and C4; normal	Chloroquine 500 mg/day. Complete resolution of subcutaneous nodules leaving depressed area
Marzano, 2005[8]	21/ F	Linear sclerotic erythematous plaques	Forehead and parietal area	NA	Noted that "Consistent with LE of the tumidus variant"	NA	Yes; diffuse membranoproliferative glomerulonephritis	ANA; positive 1:1280 homogeneous Anti-dsDNA; positive RF; positive, C3 complement; low CBC leukopenia	Intralesional steroid injection. Resolution with deep depression
Marzano, 2005[8]	32/ F	Linear subcutaneous nodules merged into an irregular indurated erythematous plaque	Lower half of right arm extending to upper third of forearm	3 Y	Lymphoplasmacytic infiltration in the dermis, fat lobules and septa, lymphoid follicles within the fat lobules, and fibrosis in the middle dermis extending to the subcutaneous tissue	Granular IgM and C3 deposits at the DEJ	Yes; autoimmune thrombocytopenia, spontaneous abortion	ANA; positive 1:640 speckled Antiplatelet antibody; positive Anti-cardiolipin IgG and IgM; positive Anti β2-glycoprotein I IgM; positive Lupus anticoagulant; negative CBC, ESR, LFT and urinalysis; within normal limit	Prednisolone 32 mg/ day and Hydroxychloroquine 200 mg/day. Improvement in erythema and sclerosis of the lesion with mild lipodystrophy and depression

Elbendary, 2016[5]	9/ F	Linear erythematous subcutaneous nodules	Medial aspect of left forearm	19 Mo	Superficial and deep perivascular, periadnexal and interstitial lymphocytic infiltrate, thickened collagen bundles, lymphoid follicles, and mild widening of fibrous septa, diminished CD34 expression in sclerotic area	Negative	No	ANA; negative Anti-dsDNA; negative Serum aldolase; elevated Anti-histone antibody; weakly positive	Pulsed intravenous methylprednisolone monthly and subcutaneous methotrexate weekly. Minimal residual discoloration, and slightly residual firmness overlying 1 lesion
Schwartz, 2020[6]	63/ F	Subcutaneous nodules coalescing into linear thickened skin	Medial aspect of right upper arm extending to axilla	1 Mo	Epidermal atrophy, interface dermatitis involving dermoepidermal junction and hair follicles, lymphocytic lobular panniculitis with hyalinized fat necrosis, deep dermal sclerosis, diminish CD34 expression within dermal fibrosis	NA	No	ANA; negative, Anti-CCP; negative	No treatment initiation. No improvement

Abbreviations: ANA, antinuclear antibody; CBC, complete blood count; CCP, cyclic citrullinated peptide; DEJ, dermoepidermal junction; dsDNA, double-stranded DNA; DIF, direct immunofluorescence; ESR, erythrocyte sedimentation rate; F, female; LFT, liver function test; M, months; NA, data not available; RF, rheumatoid factor; Y, years.

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Asian patient presenting with a unique feature of SLEP. Due to its rarity, we encourage physicians to keep in mind the coexistence of lupus panniculitis and morphea. The pathogenesis is largely unknown. Whether SLEP is a distinct entity or a coincidental overlapping disease between lupus panniculitis and localized scleroderma remains to be determined. Future studies are warranted.

Conclusion

We report a case of SLEP presented with a solitary firm to hard erythematous plaque, mimicking DFSP and biopsy results were consistent with lupus panniculitis and scleroderma. Due to its rarity, there are limited data on whether SLEP holds a prognostic value for systemic or recurrent diseases. We encourage physicians to keep in mind the coexistence of these two distinct entities.

Ethical Statement

Written informed consent was provided by the patient to have her case details and images published. Institutional approval was not required to publish the case details.

Disclosure

The authors report no conflicts of interest in this work.

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