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Case Report

Lupus profundus limited to a site of trauma: Case report and review of the literature☆

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ABSTRACT

Lupus erythematosus profundus (LEP) is a rare form of chronic cutaneous lupus erythematosus. We report on a case of a 56-year-old Caucasian woman who presented with a single, persistent, painful rash on the left hip and lateral aspect of the left upper thigh, which had been present for 2.5 years. The patient had a history of previous injury to this area before the rash started. Clinical findings showed an inflamed, hyperpigmented, and indurated plaque with a linear skin invagination and no associated systemic symptoms. A skin biopsy test result confirmed the diagnosis of LEP and the clinical and laboratory examinations ruled out systemic lupus erythematosus. After 2 months of treatment with methotrexate 20 mg weekly and 1 month of prednisolone 7.5 mg daily, the skin rash improved considerably. We also present a brief review of the epidemiology, etiology, clinical features, histopathology, laboratory findings, differential diagnosis, and treatment of LEP.

Case report

A 56-year-old Caucasian woman presented with a persistent painful rash on the left hip and lateral aspect of the left upper thigh, which had been present for 2.5 years. The patient had a fall with blunt contusion to the same area 2 months before the rash started, which caused persistent left-sided tenderness and hip pain. The patient underwent magnetic resonance imaging and the scan demonstrated an extensive subcutaneous contusion with fat necrosis in the left gluteal region with no muscle or tendon injury. Over the past years, the patient reported only persistent left hip, gluteal, and upper lateral left thigh tenderness that was associated with the development of a hyperpigmented and indurated plaque and intermittently became very inflamed but no other systemic symptoms. In the year immediately prior to presentation, linear skin invagination had developed and was enlarging gradually on the upper border with more induration, allodynia, and tenderness around this area. The patient was otherwise healthy with a history of osteoporosis and no current treatment and she had no personal and family history of autoimmune diseases.

During a physical examination, the patient was determined to have a large, indurated, tender, erythematous-to-violaceous, poorly demarcated plaque on the left hip and lateral aspect of the left upper thigh. Inside the plaque was a large linear skin depression (Fig 1A). She had no other skin lesions or symptoms, except for arthralgias on the hand joints.

Laboratory test results disclosed normal levels of blood cell counts, urinalysis, complements (C3, C4, CH50), and renal function. Some liver function tests results showed slightly elevated levels (gamma-glutamyl transferase 63 U/L [normal, 0-30 U/L], aspartate aminotransferase 52 U/L and alanine aminotransferase 55 U/L [normal, <45 U/L], and erythrocyte sedimentation rate 30 mm/hr [normal, 0-20 mm/hr]). Serological study results showed low grade antinuclear antibody results (antinuclear antibody, titer 1:320, homogenous pattern; titer 1:80, nucleolar; 1:80, cytoplasmic) and anti-ribonucleoprotein/Sm antibody and anti-nucleosomes antibody test results were positive. Anti-ds-DNA antibody, anti-Sm antibody, rheumatoid factor, and anti-cyclic citrullinated peptide antibody test results were negative.

An examination of deep skin biopsy tissue of the lesion revealed an epidermis of normal thickness. The underlying dermis showed perivascular and interstitial lymphocytes with dermal edema and

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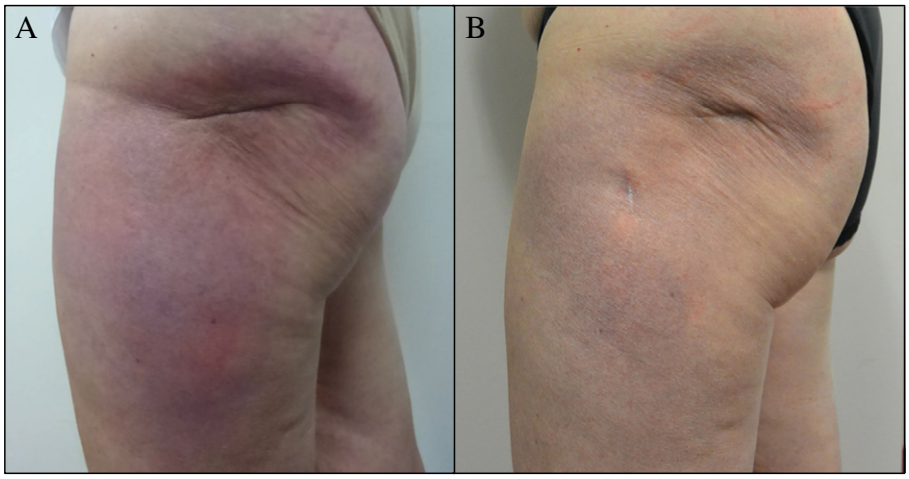

Fig. 1. Clinical manifestation of lupus erythematosus profundus. A) Before treatment. A large, indurated, erythematous-to-violaceous, poorly demarcated plaque on the left hip and lateral aspect of the left upper thigh within a large linear skin invagination of lipoatrophy. B) Plaque with less edema, inflammation, and induration, after 2 months of therapy, showing signs of postinflammatory hyperpigmentation.

mucin. The inflammation was more marked in the deeper dermis and subcutis where there were lymphoid aggregates with focal germinal center formation. Fat lobules were reduced in size and associated with hyaline fat necrosis. The inflammation and lobular panniculitis extended the full depth of the biopsy tissue. Lymphocytes surrounded the vessels and permeated the walls but vascular destruction was not evident (Fig 2A and B). The test results of a direct immunofluorescence examination (DIF), and fungal and bacterial cultures of skin specimens were negative.

A diagnosis of lupus erythematosus profundus (LEP) was made on the basis of a combination of clinical (indurated erythematous-violaceous patch with hypodermis atrophy) and histological findings of lymphocytic cells infiltration over the dermis (more marked within the deeper dermis) and subcutaneous tissues, and hyaline fat necrosis in the context of antinuclear antibody positive test results. Treatment with hydroxychloroquine 200 mg twice daily was initiated but the patient developed a drug eruption within 2 weeks, which was confirmed by an examination of skin biopsy tissue. Treatment with hydroxychloroquine was stopped and methotrexate 5 mg/week was initiated with gradual increases up to 20 mg/week, along with prednisolone 7.5 mg daily for 1 month. After 2 months of methotrexate 20 mg weekly, the skin rash improved considerably, showing less induration, edema, and erythema (Fig 2B). The patient continued treatment with methotrexate 20 mg/week for 6 months without complications or flare-ups.

Discussion

LEP is an infrequent form of chronic cutaneous lupus erythematosus (Massone et al., 2005; Tuffanelli, 1971). The term *lupus profundus* is used with dermal and subcutaneous involvement. When there is solely subcutaneous involvement, it is called lupus panniculitis (Walling and Sontheimer, 2009). Studies have described the frequency of LEP at 1% to 3% of patients with cutaneous lupus erythematosus (Requena and Sánchez Yus, 2001; Walling and Sontheimer, 2009). LEP may manifest as a unique entity or can be associated with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE). A patient with LEP has approximately 50% of probability to develop SLE (Kundig et al., 1997). When LEP is present in combination with SLE, the prognosis of the systemic disease is often better because the patient usually develops a mild form of SLE with infrequent neurological and renal manifestations (Fraga and García-Díez, 2008; Kundig et al., 1997). Our patient did not fulfill the American College of Radiology criteria for SLE.

Epidemiology

LEP frequently occurs as a separate disease. However, 2% to 5% of patients with SLE and approximately 10% of those with DLE develop lupus panniculitis. LEP presents more frequently in women. The percentages of frequency are variable with a female/male ratio between

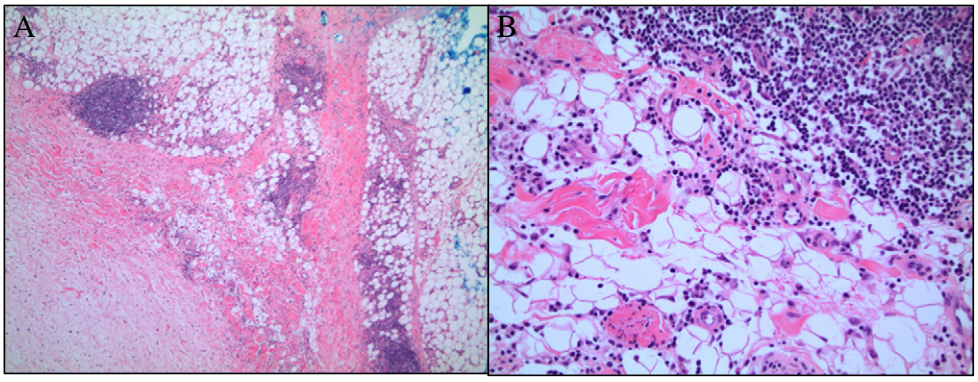

Fig. 2. Histopathology. A) Dermis with perivascular and interstitial lymphocytes, edema and mucin. The inflammation is more marked within the deeper dermis and subcutis where there are aggregates with focal germinal center formations. B) Subcutis with interstitial lymphocytes and plasma cells. Fat lobules are reduced in size and associated with hyaline fat necrosis.