Lupus Erythematosus Panniculitis in Pregnancy

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Abstract

A case of lupus erythematosus (LE) panniculitis in pregnancy without any lesions of discoid LE or systemic LE is being reported. There were no systemic symptoms. Her ANA, anti-dsDNA, anti-Ro/SSA, and anti-La/SSB antibodies were within normal limits. Diagnosis of lupus panniculitis was considered on clinical and histopathological grounds. The condition responded favorably to systemic steroid therapy.

Key Words: *Lupus erythematosus panniculitis, pregnancy, systemic steroid*

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What was known?
Lupus erythematosus profunds or panniculitis is a chronic relapsing condition in which painful lesions may heal with significant lipoatrophy and scarring. It often responds to treatment with antimalarials and systemic steroids should be reserved for resistant and widespread lesions.

Introduction

Lupus erythematosus (LE) panniculitis is a quite rare clinical entity characterized by one or several firm, asymptomatic, often fairly large subcutaneous nodules, as a manifestation of systemic LE (SLE) or discoid LE (DLE). The inflammatory reaction in LE profundus (LEP) takes place primarily in the deep corium and the subcutaneous tissues leading to deep indurated nodules or sharply defined plaques. The overlying skin usually appears normal, but there may be erythema, atrophy, ulceration or poikilodermatous or hyperkeratotic changes.[1] The lesions are most frequent on cheeks, but other sites of predilection are face, upper arms, hands, chest, buttocks, and thighs.[2]

Case Report

A 24-year-old primigravida of 6th month gestation presented asymptomatic, multiple ulcerated lesions situated bilaterally over cheeks just in front ears. The lesions first developed as erythematous, painless nodules, oval-shaped, and firm in consistency; gradually increased in size to reach the present size and ulcerate. There was no history of preceding trauma. She had no complaints of fever, arthralgia, oral ulceration, or photosensitivity. None of her family members was having similar illness. Examination revealed multiple erythematous, indurated plaques of about 2 cm × 3 cm over both the cheeks with ulceration at base [Figure 1]. Few atrophic scars with thickened, infiltrated, and raised erythematous margins were present. There was no lymphadenopathy. No associated lesions of DLE were found. Except mild degree of pallor, the rest of the general and systemic examination were within normal limits.

Besides routine investigations, special investigations, except anemia, like ANA, anti-dsDNA, anti-Ro/SSA, and anti-La/SSB antibodies were within normal limits. Mantoux test, rheumatoid factor, VDRL, and ELISA for HIV were negative. Direct immunofluorescence staining was not done because of nonaffordability. Ultrasonography showed normal fetal growth. Skin biopsy showed a moderately dense lymphoplasmacytic lobular panniculitis with septal thickening. The fat lobules showed patchy infiltrate with lymphocyte and plasma cells, and the adipocytes showed signs of coagulative fat necrosis with microcyst formation. The overlying dermis showed sparse

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superficial and deep perivascular lymphocytic infiltrate with mucin deposits [Figures 2 and 3]. Diagnosis of LEP was made on a clinical and histopathological basis. The patient was started oral prednisolone 20 mg daily, which led to significant improvement. This was given for 2 weeks and then gradually tapered over a period of 2 months. Obstetrician reference for fetal monitoring was done at regular intervals. After 2 months, lesions healed with atrophy [Figure 4] and did not show the occurrence of any new lesions till delivery. Newborn was healthy, and blood was negative for ANA, anti-dsDNA, anti-Ro/SSA, and anti-La/SSB antibodies.

Discussion

Kaposi first described subcutaneous nodules in LE in 1883 but the term "LE profundus" first used by Irgang in 1940.[3] LEP is unusual but distinct clinical variety of LE, commonly present in the third to sixth decades of life with a female: male ratio of 2:1. The typical lesions are multiple indurated nodules or plaques involving the arms, face, buttocks, legs, chest, and less frequently the abdomen, back and neck, and breast. The lesions heal leaving depressed atrophic scars.[4] The histopathology is sufficiently characteristic to establish the diagnosis in the absence of other cutaneous or systemic lesions of LE.[3] Histopathologically, LEP is classified as a lobular panniculitis. However, it is frequently accompanied by septal involvement; and therefore, termed as "mixed panniculitis."[5] Lupus panniculitis is often difficult to diagnose as other form of panniculitis may present similarly. The differential diagnoses of LEP include panniculitis due to other connective tissue disorders like dermatomyositis or scleredema and Weber-Christian panniculitis or Jessner's lymphocytic infiltration, lyrnphocytoma cults, and sarcoidosis. LEP may develop in association with DLE or SLE or may occur as an isolated phenomenon.[2] Raksha Patel *et al*. reported a case of lupus panniculitis as an initial manifestation of SLE. Aggarwal *et al*. reported a case of LEP with associated mastitis but without any lesions of DLE or SLE.[4] We found only three cases of LEP in pregnancy even after extensive Medline-based literature search.[6-8] Nitta reported a case of lupus panniculitis associated with neonatal LE.[6] Grob *et al*. reported a case of Fibrosing LE panniculitis in a pregnant woman with anti-RO/SSA antibodies in mother and child.[8] Treatment of LEP is challenging as it has a chronic and relapsing course.

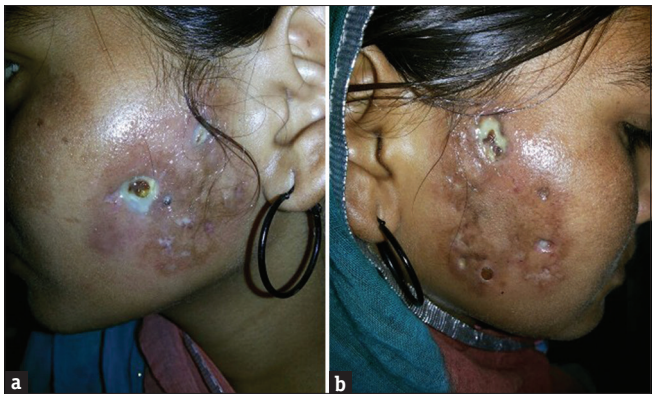

Figure 1: (a and b) Multiple erythematous ulcerated lesions situated bilaterally over the cheeks

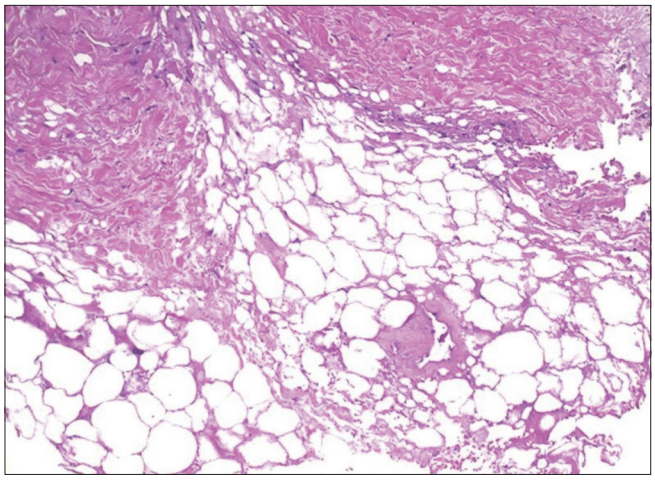

Figure 3: Fat lobules showing patchy infiltration and signs of coagulative fat necrosis with microcyst formation (H and E, ×10)

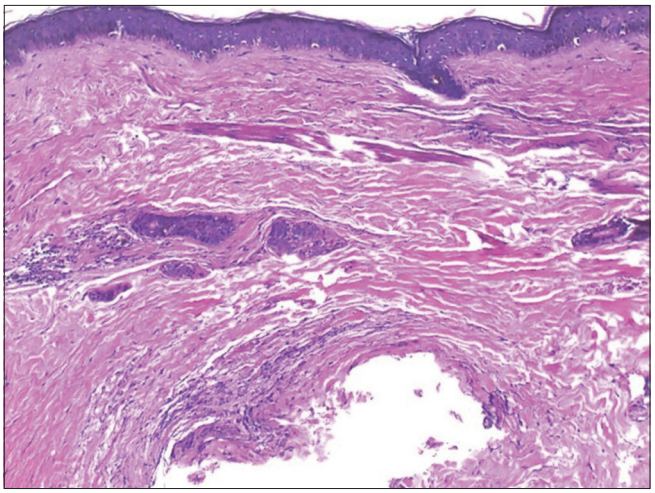

Figure 2: Moderately dense lymphoplasmacytic lobular panniculitis with septal thickening (H and E, ×10)

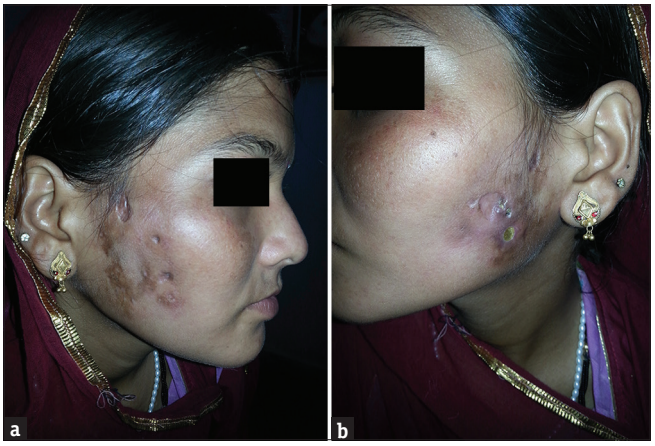

Figure 4: (a and b) Lesions healed with atrophy after treatment

Anti-malarials are the mainstay of treatment. Intralesional injections of triamcinolone (5 mg/mL) may be helpful.[3] Systemic steroids are generally not recommended for lupus panniculitis unless it is indicated for other manifestations of SLE. It is generally reserved for widespread and resistant lesions.[9] As anti-malarials are contraindicated in pregnancy, our patient was treated with steroids and responded well.

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Nil.

Conflicts of interest

There are no conflicts of interest.

What is new?
Very few cases of LEP with pregnancy have been reported in the literature. We reported an isolated case of LEP with pregnancy which responded well to oral steroids.

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