Net Letter

Lupus erythematosus profundus

Sir,

In 1883, Kaposi,[1] first, described the association of subcutaneous nodules with lupus erythematosus. This finding has subsequently being recognized as LE profundus, more accurately, LE panniculitis (LEP). In 1956, Arnold.[2] reviewed the literature and helped substantiate the existence of this entity. LEP is an unusual but distinct clinical variety of lupus erythematosus. The inflammatory reaction in 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.[3] The lesions are most frequent on cheeks but other sites of predilection are face, upper arms, hands, chest, buttocks and thighs.[4]

A 39-year-old female presented with history of development of a single, asymptomatic, erythematous plaque in the upper part of left arm since four years. The lesion, initially, was about 1 × 2 cm^2 in size, oval in shape and firm in consistency. It gradually increased in size to about 8 × 5 cm^2 was firm to hard in consistency. There was no history of preceding trauma. No history of taking injection. Patient had myalgia, but no other systemic symptoms. Examination revealed a large erythematous plaque about 8 × 5 cm^2 present over the lateral aspect of the left arm [Figure 1] with well-defined margins, sharply demarcated from the surrounding normal skin, firm to hard in consistency and non-tender. There was no lymphadenopathy.

The following laboratory tests were done which showed normal results: complete blood cell count, differential cell count, ESR, total serum proteins with A:G ratio, blood sugar, blood urea, serum creatinine, serum electrolytes, liver function tests and urinalysis. X-ray chest was normal. Rheumatoid factor was negative. Blood for LE cells was normal. Connective tissue profile showed antinuclear antibodies positive in 1:100 titre by direct immunoflourescence method, but anti ds DNA and Anti Sm, Anti RNP, Anti Ro, Anti La, Scl antibodies were negative.

Skin biopsy showed septal and lobular panniculitis [Figure 2] without vasculitis with widened septae and infiltration by moderately dense infiltrate of lymphocytes. The infiltration is spilled into the periphery of the lobules forming lace like network,

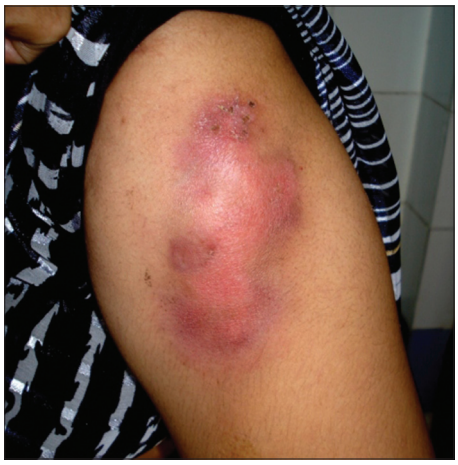

Figure 1: Large erythematous plaque present over the lateral aspect of the left arm

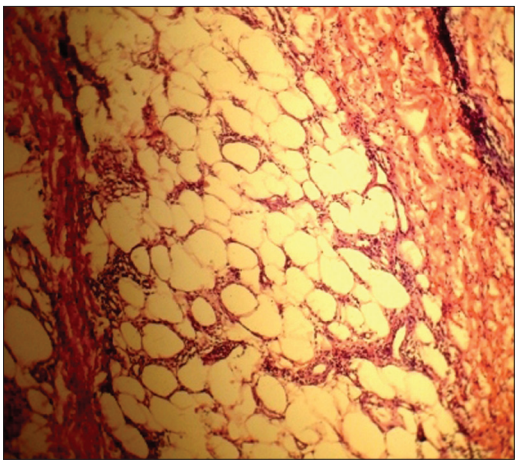

Figure 2: Features suggestive of septal and lobular panniculitis with infiltration spilled into the periphery of the lobules forming lace like network (H&E, ×40)

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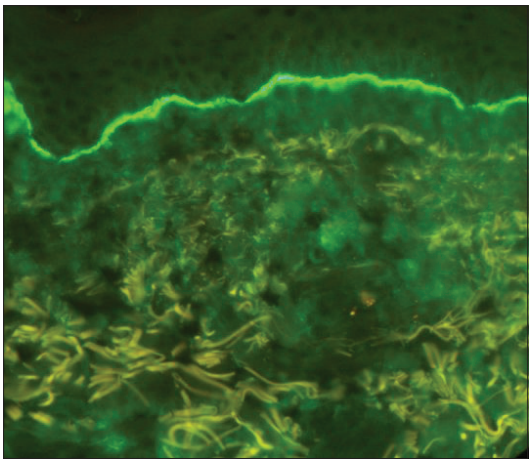

Figure 3: Linear depositis of IgG,IgM and C3 along the basement membrane zone

particularly close to the septa. Overlying dermis is uninvolved. The dermoepidermal junction is infiltrated by lymphocytes. The basement membrane zone is thickened and basal layer showed vacuolar type of degeneration. Mucin staining was not done.

Immunofluorescence studies were done on both lesional and non lesional skin, which showed linear depositis of IgG, IgM and C3 along the basement membrane zone in the lesional skin [Figure 3] and negative results in the non-lesional skin.

LEP may develop in association with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE) or may occur as an isolated phenomenon.[5]

It is to be differentiated from panniculitis due to other connective tissue diseases, like dermatomyositis and scleroderma and from Weber-Christian disease, protease inhibitor deficiencies, erythema nodosum, lipodystrophies and sarcoidosis.[6] Most of the reported patients are middle aged but some childhood cases (7-18 years) have been recorded.

In our patient, there was no history of photosensitivity, oral ulceration, arthralgia or neuropsychiatric manifestations. Family history was negative. The antinuclear antibody was positive and lesional lupus band test was positive. This case is reported for its difficulty in diagnosis.

The patient was given 200 mg of Hydroxychloroquine twice daily, orally, along with 40 mg of prednisolone daily, orally, which led to significant improvement after six months.

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