

Tender indurated plaque with ulceration on the chin

A 35-year-old lady presented with a one-year history of an erythematous, painful lesion on the left side of the chin. It started as a pea-sized papule on the chin, which gradually increased in size to become a plaque extending upto the left submandibular area. There was history of an occasional purulent discharge. There was no history of preceding trauma, dental infection, photosensitivity, joint pains, fever or other systemic complaints. Physical examination revealed a single erythematous indurated nodulo-plaque measuring 5×3 cm on left side of the chin extending upto submandibular area. Overlying skin had multiple ulcers covered with yellowish-brown adherent crust and atrophic scarring at places [Figure 1]. It was firm to hard in consistency, tender



Figure 1: An ill-defined erythematous indurated plaque measuring 5×3 cm with overlying skin showing adherent yellowish-brown crust over ulcerated areas on the left side of chin extending to the submandibular area

and free from underlying structures. Anti-nuclear antibody (ANA) was 1+ in a speckled pattern and anti-double stranded DNA (ds DNA) antibody was negative. On histological examination, the epidermis showed follicular plugging and basal cell vacuolization. In addition to perivascular and perifollicular lymphomononuclear infiltrate in the superficial and mid-dermis [Figure 2a]. A lympho-histiocytic infiltrate in the fat lobules with diffuse necrosis of adipocytes was also observed [Figure 2b].

WHAT IS YOUR DIAGNOSIS?

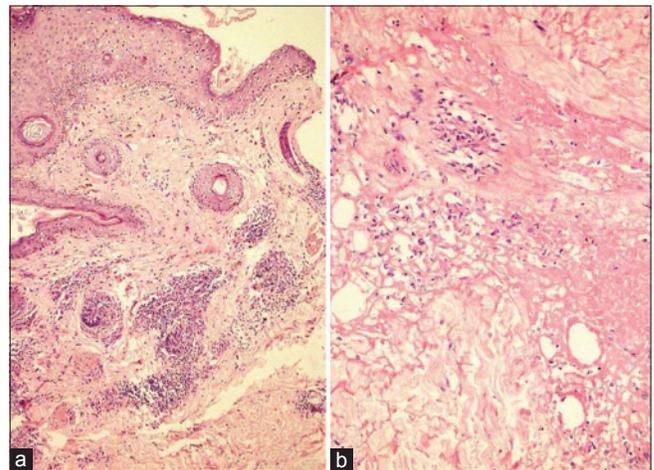


Figure 2: (a) Follicular plugging, basal cell vacuolization, perivascular and perifollicular lymphomononuclear infiltrate in superficial and mid-dermis (H and E, $\times 10$) (b) Lobular panniculitis with necrosis of adipocytes (H and E, $\times 40$)

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Answer:

Lupus erythematosus panniculitis

DISCUSSION

Subcutaneous lesions in patients with lupus erythematosus (LE) were first described by Kaposi in 1883 and were termed lupus erythematosus of Kaposi-Irgang by Arnold in 1956.^[1] LE panniculitis (LEP), a distinct variant of LE, commonly presents in the 3rd to 6th decade of life with female to male ratio of 2:1. The characteristic lesions consist of tender subcutaneous nodules and plaques. Ulceration is less frequent and described in about 28% cases of LEP.^[2] The association of LEP with discoid LE (DLE) varies from 33% to 60%.^[2,3] Although 10-42% patients may have associated systemic lupus erythematosus (SLE),^[4] LEP has been reported to occur in only 2-5% cases of SLE and is a marker of less severe forms of SLE.^[5]

Histopathologically, LEP is classified as a lobular panniculitis. However, it is frequently accompanied by septal involvement; and therefore, termed as 'mixed panniculitis'. The histopathologic criteria proposed for the diagnosis of LEP include major (important for the diagnosis) and minor criteria (not necessary for diagnosis).^[5] The major criteria are: (1) hyaline fat necrosis, (2) lymphocytic aggregates and lymphoid follicle formation, (3) periseptal or lobular lymphocytic panniculitis, and (4) calcification. The minor criteria are: (1) changes of DLE in the overlying skin, (2) lymphocytic vascular inflammation, (3) hyalinization of sub-epidermal zone, (4) mucin deposition, (5) histiocytes and small granulomas, and (6) infiltrates of plasma cells and eosinophils. Overlying dermo-epidermal findings of DLE may

be present in 50-75% cases even without clinical evidence of DLE. Direct immunofluorescence may show granular deposition of IgG, IgM, and C3 at the dermal-epidermal junction and blood vessels of deep dermis and subcutis in 50-70% cases,^[1,5] particularly when there is concomitant DLE.

Treatment of LEP is challenging as it has a chronic and relapsing course. Antimalarials are the first line of treatment. Oral corticosteroids though beneficial,^[5] should be used for severe cases with associated SLE due to their side effects. Thalidomide, dapsone, cyclosporine, intravenous immunoglobulins and rituximab have shown efficacy in intolerant or refractory cases.^[6] Our patient, was started on hydroxychloroquine (HCQ) 400 mg/day. Four days after commencing HCQ, she developed a maculopapular drug rash. HCQ was stopped and she was administered oral prednisolone 40 mg/day, which was tapered by 10 mg every month. At two months, the ulceration healed and there was significant reduction in tenderness and induration of the plaque [Figure 3]. Post-treatment skin biopsy showed normalization of basal layer with reduction in the dermal infiltrate [Figure 4a] and early collagenization of subcutaneous fat, suggestive of healing [Figure 4b].

Patients with LEP should be followed up regularly, lest they develop SLE. Repeat biopsy with immunohistochemistry and the T-cell receptor gene rearrangement studies may be required in refractory cases to rule out subcutaneous panniculitis-like T-cell lymphoma (SPTCL). Histopathologically, LEP encompasses a spectrum of features with small, mature lymphocytes showing



Figure 3: Marked decrease in erythema and induration of plaque and healing of ulceration with atrophic scarring after 2 months of oral prednisolone

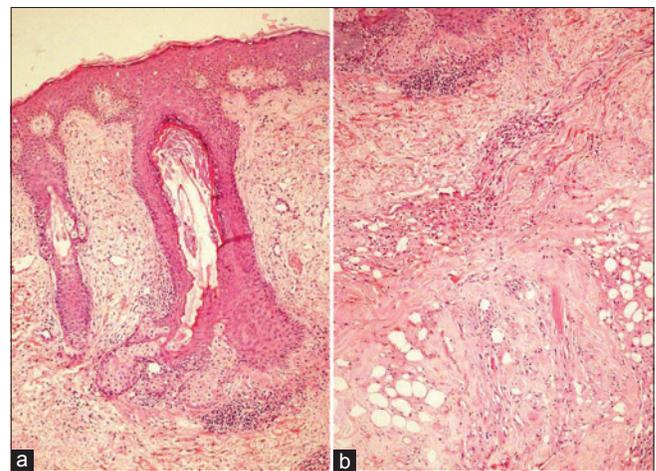


Figure 4: (a) Intact basal layer with reduction in dermal inflammation (H and E, $\times 10$) (b) Early collagenization of subcutaneous fat (H and E, $\times 40$)

polyclonality on one hand and pleomorphic lymphocytes with hyperchromatic nuclei demonstrating deletion of pan T-cell markers and monoclonal T-cell receptor gene rearrangement on the other. The term subcutaneous lymphoid dyscrasia has been proposed to include atypical cases of LEP, SPTCL and indeterminate lymphocytic lobular panniculitis.^[7] Further, vacuolar interface dermatitis and dermal mucinosis has been reported in SPTCL, thereby pointing to an overlap between the two entities.^[8] However, Pincus *et al.*,^[9] suggested that lymphoid follicles with reactive germinal centers and mixed infiltrate comprising of plasma cells may favor LEP over SPTCL.

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