

Lymphocytic lobular panniculitis – is it lupus or lymphoma?

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Abstract

Lymphocytic lobular panniculitis, or lymphocytic infiltration of the panniculus, is considered the hallmark of both lupus erythematosus profundus and subcutaneous panniculitis-like T cell lymphoma. Both these entities may be clinically indistinguishable, and in a proportion of patients there may be overlapping histological and immunohistochemical features. We present here a 12 year old female who had such a panniculitis with overlapping features, and responded to therapy with a benign course.

Case report

A 12 year old girl presented with painless subcutaneous nodules and indurated plaques on both thighs, face, upper limbs for 3 months duration. She also had low grade fever for the last 3 weeks with associated periorbital swelling. She was otherwise well and had no complaints of weight loss, anorexia, photosensitivity, joint pains or muscle weakness.

Examination showed a well-looking child with low-grade fever. She was not pale or icteric, and there was no lymphadenopathy or bone tenderness, malar rash, DLE or oral ulceration. There was bilateral periorbital swelling without erythema. There were non-tender indurated plaques and nodules on both thighs, upper limbs and submental region. Individual lesions were 1-2 cm in diameter. The lesions on the thigh were arranged in an annular configuration. Apart from mild hyperpigmentation there were no overlying changes such as ulceration, erythema or DLE. There was no hepatosplenomegaly or proximal muscle weakness.

Investigations showed ESR 20 with normal CRP. WBC was 2100/mm³ with Hb10g/dl and platelet 204000/mm³. Blood picture showed leukopenia with neutropenia, and no abnormal cells. Bone marrow aspirate and trephine biopsy showed only mildly hypocellular marrow and no abnormal cells. UFR, serum creatinine and ultrasound scan of abdomen were normal. LDH was mildly elevated (630.7). Lesional biopsy showed unremarkable epidermis and dermis with a lymphocytic lobular panniculitis. There was no vasculitis or lymphoid follicle formation. There was no cellular atypia. Immunohistochemistry revealed CD3 positivity with CD20, 56 and 4 negative.

The lupus band test was negative.

The patient was commenced on prednisolone 30mg/d and hydroxychloroquine 200mg/d. There was immediate defervescence of fever the following day, with improvement of WBC count within the next several days. No new skin lesions developed, and existing lesions as well as periorbital swelling subsided over the next 1-2 weeks.



Figure 1. Annular arrangement of panniculitis lesions on thigh.



Figure 2. Submental swelling and periorbital oedema.

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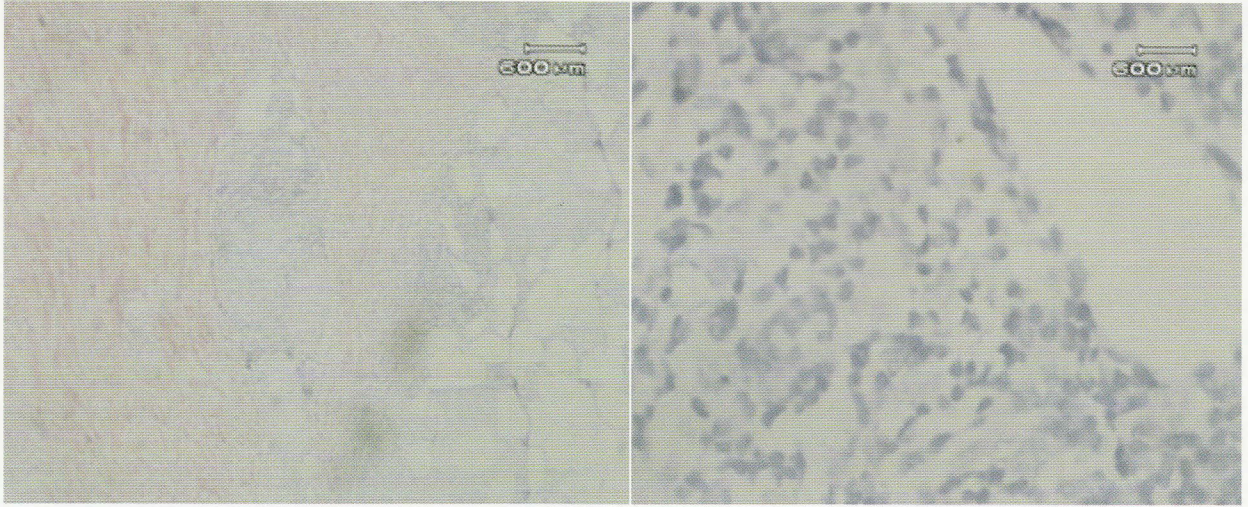


Figure 3 and 4. *Lymphocytic lobular panniculitis (10×4) and (10×40).*

Discussion

Lymphocytic lobular panniculitis is the term describing lymphocytic infiltration of the panniculus. It is considered to be the hallmark of lupus erythematosus profundus (LEP) and subcutaneous panniculitis-like T cell lymphoma (SCPTL)¹. However, intermediate forms have been described, termed indeterminate lymphocytic lobular panniculitis, indicating that the above represent a spectrum of subcuticular lymphoid dyscrasia, with LEP at the benign end and SCPTL at the malignant end of the spectrum². All of these may present in a clinically similar manner, with erythematous, violaceous plaques and nodules involving proximal extremities, trunk and buttocks. The literature reports many cases of SCPTL which had initially been misdiagnosed as LEP, or later changed from LEP to SCPTL^{3,4}. Furthermore, some cases of SCPTL such as the SCPTL-AB type, may respond to steroids in the same way as LEP⁵.

In the absence of characteristic clinical features, LEP and SCPTL may be clinically indistinguishable. Histologically also there may be few distinguishing features⁶. Both show lymphocytic panniculitis, and fat necrosis, rimming of adipocytes and eccrinotrophism may be seen in both⁷. Lymphoid atypia has been reported in LEP, contributing further to the diagnostic confusion⁸. However erythrophagocytosis indicates neoplasia. Immunohistochemistry and T cell receptor gene rearrangement studies are useful to indicate the loss of CD markers and clonality of the infiltrate.

Our young female patient did not show any

characteristic clinical features or serological evidence of lupus, and the lupus band test was negative. She had fever and cytopenia, with mildly elevated LDH level. Histologically there was no cytological atypia, but immunohistochemistry indicated monoclonality. However her clinical course was benign and she showed an immediate and sustained remission to hydroxychloroquine and steroid therapy. Therefore it is possible that this patient represents an intermediate site on the lymphocytic lobular panniculitis spectrum. Despite her good response, close follow up of this patient is mandatory as she may later revert to a less benign course.

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