CASE 1

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Case

A 7 – year - old Caucasian girl presented with a 3-month history of nodules on her thighs and buttocks. During a one-month period, the lesions gradually increased in size and number. They became indurate and tender. Similar nodules appeared on forearms after 4 months. The lesions gradually healed with a depressed scar. On physical examination, we found many indurate areas 10-40 mm in diameter .The surface of some of them was erythematous and scaly. The lesions were not ulcerated and did not have a discharge (figure 1).

On examination, she was afebrile and healthy, organs such as heart, lungs, abdomen , joints and muscles were normal. Laboratory evaluation findings including complete blood cell count, liver and renal function test values, ESR, CRP,VDRL, BUN, Creatinin, complement were all normal .The titers for FANA, Anti ds DNA, Anti Sm, Anti Scl-70, Anti –RNP, Anti-Ro, Anti-La, Anti –Jo1, Anti Phospholipid Ab, Anti cardiolipin Ab, c- ANCA , p- ANCA were negative. A skin biopsy was taken (figures 2).

What is your diagnosis?

Move on to next page for the answer and discussion.

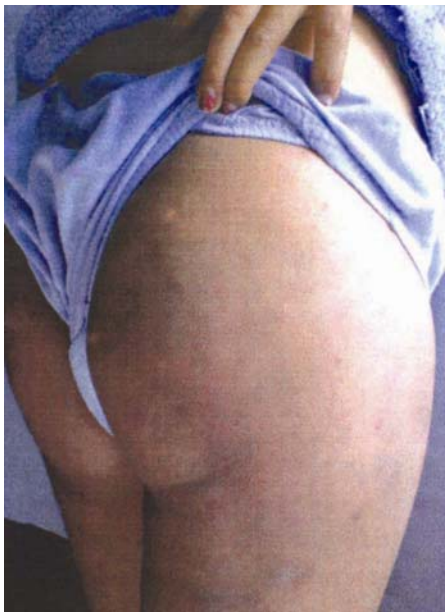

Figure 1

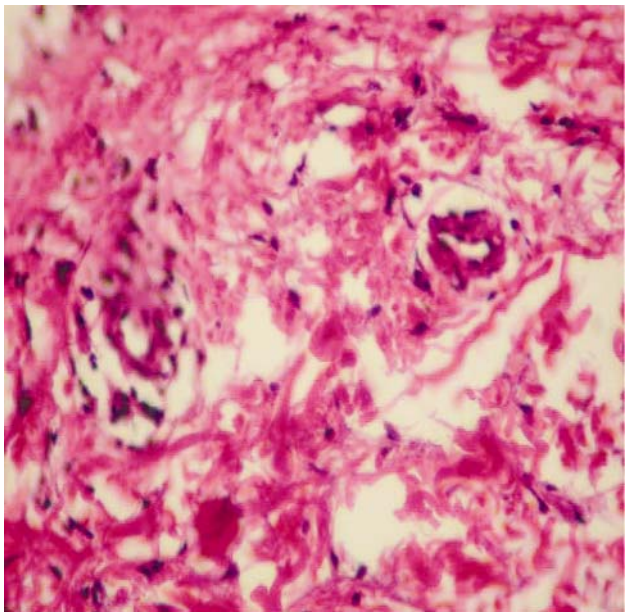

Figure 2

Diagnosis: Lupus Profoundus (Lupus Paniculitis)

Histologic findings:

A skin biopsy specimen taken from nodule on the thigh demonstrated hyperkeratosis and keratotic plugging, atrophy of the stratum malpighi, hydropic degeneration of the basal layer and a patchy infiltration composed of lymphoid cells, nonspecific panniculitis consisting of lymphoid cell, plasma cells and histiocytes, necrobiotic changes with fibrinoid deposits and eosinophilic collagen in the subcutaneous tissue, vessels with inflammatory infiltration and thickening of their wall with narrowing of their lumen (Figures 2).

Discussion

Lupus erythematous profoundus /LE panniculitis is a rare form of chronic cutaneous lupus erythematous typified by inflammatory lesions in lower dermis and subcutaneous tissue[1] with a chronic and recurrent clinical course.

In this type of LE, deep dermal and subcutaneous nodules are 1 to 3 or 4 cm in diameter, rubber-firm, sharply defined, persistent and nontender,[2,4] patients have multiple painful deep subcutaneous nodules.[3,4] The overlying skin usually appears normal; although histologically, there are changes at the dermo-epidermal junction in 61 %[5]. It most often occurs beneath normal skin of the head, face or upper arm. Chest, buttocks, and thighs may also be involved. This form of LE is characteristically chronic and occurs most often in women between ages 20 and 45. The diagnosis is more likely to be made if it is known that the patients have LE, many patients have DLE at other sites, most frequently on the cheeks, but they can occur on the face, arms, hands, breasts, buttocks, legs or less typically in the overlying skin. The lesions may heal with a deep depression, from loss of the panniculus, which may take years to fill in[2,5] and healing rarely leads to soft slightly pink areas of anetoderma up to 4 cm in diameter.[5]

Lupus profundus confined to the breast has been called lupus mastitis and may herald SLE. It may be confused with carcinoma.[5] LE profundus may affect the periorbital tissues and cause severe localized edema. It may occur with eyelid plaques. Whenever it occurs, the nodules are persistent, and lesions in the cheeks may lead to marked disfigurement.[5]

The histology shows lymphocytic panniculitis, hyaline degeneration of the fat, hyaline papillary bodies, and dense, sharply circumscribed lymphocytic nodules in the lower dermis and fat. The overlying epidermis may show basal liquefaction and follicular plugging, or may be normal. In one study with direct immunofluorescence, seven of nine cases showed granular deposition of immunoglobulin and C3 at the dermoepidermal junction.[4]

Associated diseases: Lesions can occur after trauma or surgical biopsy and have been precipitated by electromyography. Monoclonal ammopathy has been reported in LE profundus. LE profundus followed thrombocytopenic purpura in one case.[5]

Treatment with Clobetazole propionate cream and intralesional injections of Triamcinolone (5mg/ml) may be helpful. Antimalaria drugs are helpful, especially in children.[4, 5] Oral thalidomide can resolve resistant lesions.[5] Lupus panniculitis can successfully be treated with dapson.[6]

References

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