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CASE REPORT

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# A Non-Erythematous and Depressed Panniculitic Lesion in the Gluteal Region: Lupus Profundus or Subcutaneous Dermatofibroma?*

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Lupus profundus (lupus erythematosus panniculitis) is an unusual clinical variant of lupus erythematosus (LE) in which the cutaneous inflammatory reaction occurs primarily in the deeper corium. The common clinical features of lupus profundus include asymptomatic, firm, and sharply defined nodules. The histologic findings are characterized by non-specific panniculitis composed of lymphoid cells, plasma cells, and histiocytes with varying degrees of necrobiotic changes with fibrinoid deposits (1). Subcutaneous dermatofibroma also presents with a depressed large subcutaneous nodule, in which fibrohistiocytic proliferation, storiform arrangement of spindle cells, sclerotic collagen, and interspersed trapping of collagen at the periphery of the lesion were the histopathologic findings (2). We report here a woman who had a hard and depressed gluteal lesion, which was clinically mimicking subcutaneous dermatofibroma, but histopathologically diagnosed as lupus erythematosus panniculitis.

## Case Report

A 43-year-old woman presented to our outpatient clinic with a 2-month history of a non-tender, hard, plaque-like lesion in her left gluteal region. On inspection, there was only a non-erythematous depressed area, under which an indurated nodule could be palpated (Figure 1). Her histopathologic examination revealed orthokeratosis, mucinous edema in the superficial dermis, perifollicular mononuclear inflammatory cell infiltrate, a dense lobular infiltrate of lymphocytes with expansion to the septal area in subcutaneous tissue, hyaline necrosis, and vasculitis (Figures 2a and 2b). Direct immunofluorescence (DIF) was negative at the dermoepidermal junction, and around superficial blood vessels and the follicular basement membrane.

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Laboratory tests showed a normal complete blood cell count, routine blood chemistry analysis, immunoglobulins, complement levels, and antinuclear antibodies. As all the serologic tests for LE were negative in our patient, she did not fulfil the criteria for systemic lupus erythematosus (SLE).

Although her lesion was clinically mimicking subcutaneous dermatofibroma, the clinical and histopathological findings supported a diagnosis of lupus profundus.

It is known that in the treatment of lupus profundus, surgical debridement or resection of individual lesions may be attempted when all other modalities have failed and there is appreciable debilitation. Interestingly, after the biopsy, our patient's lesion regressed without needing any other modalities. Moreover, no recurrence was detected

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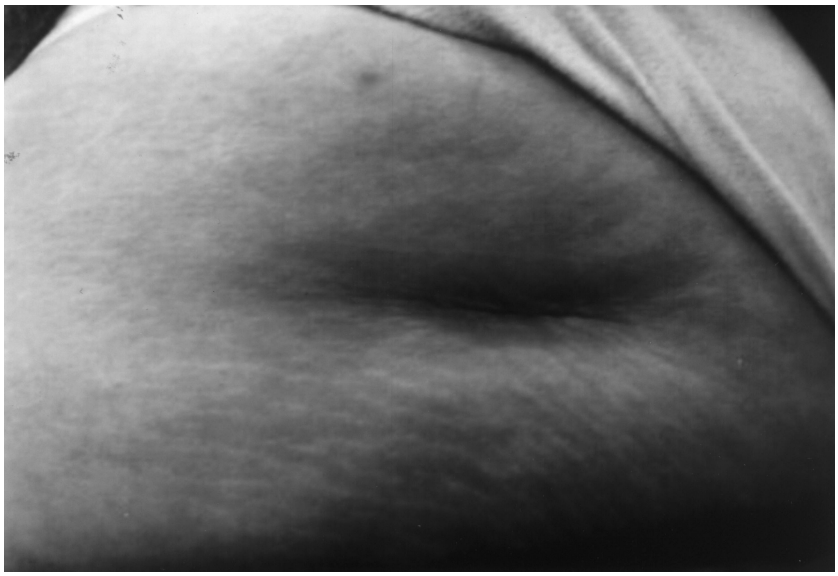

Figure 1. Non-erythematous, depressed and indurated area in the left gluteal region.

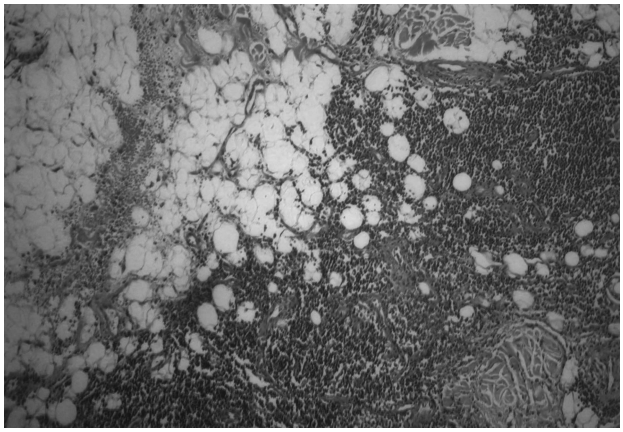

Figure 2a. Lupus panniculitis (H&E x100).

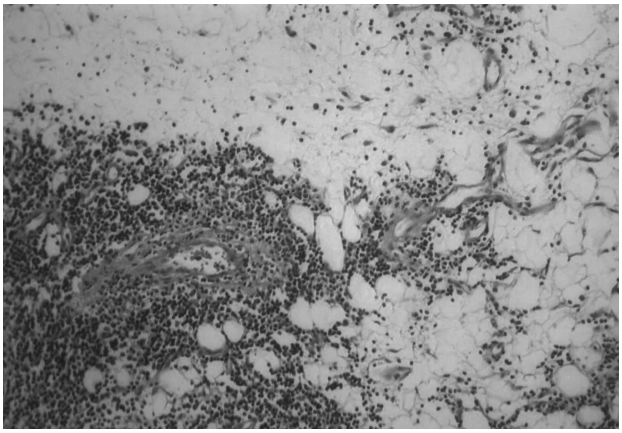

Figure 2b. Lupus panniculitis with vasculitis (H&E x200).

during the 2-year follow-up. As lupus panniculitis may precede the development of associated LE in some of these patients, we checked our patient for systemic signs, symptoms and serologic markers of LE again during this period.

Lupus profundus is a rare skin condition originally described by Kaposi in 1883 and later by Irgang in 1940. It is a clinical variant of LE in which the deep dermis and subcutaneous fat are predominantly affected. It may occur on its own or in association with discoid lupus erythematosus (DLE) or SLE (3). In cases associated with SLE, it may precede SLE by several years. In the majority of cases, it tends to have a mild chronic course marked by recurrent nodules or plaques (4). In our case, there was only one depressed nodule, which did not recur during the follow-ups. Moreover, there were no systemic symptoms or signs of SLE.

Dermatofibroma (DF) is usually confined to the dermis and in most instances is not difficult to diagnose. One of the peculiar types that has only been briefly alluded to is subcutaneous DF (5). Only one article cited these forms of distinct clinical manifestations of purely subcutaneous DF (5), although the histological variants of DF with deep subcutaneous extension, so-called deep penetrating DF,

with or without a depressed surface, is more commonly encountered (6-8). Histopathologically, there is a well-circumscribed, purely subcutaneous nodule composed of fibrohistiocytic proliferation. The storiform arrangement of spindle cells, sclerotic collagen, and interspersed trapping of collagen at the periphery of the lesion is consistent with subcutaneous DF. Dermatofibrosarcoma protuberans is a rare dermal and subcutaneous spindle cell tumor, which may be difficult to differentiate; however, the tumor is positive for CD34 (2). There was neither fibrohistiocytic proliferation nor mitosis with positivity for CD34 on punch biopsy in our patient.

In most previous studies, lupus profundus has been reported to affect the middle-aged (40-50 years) with female preponderance (9,10). In some recent studies, lupus profundus appears to affect a slightly younger age group, between 20 and 40 years (4,11). There is a predilection for the face, upper limbs, buttocks and scalp. Furthermore, very few had truncal involvement (4). Both the age and gender of our patient, and the localization of her lesion were consistent with lupus profundus.

In some studies, on histopathologic examination of lupus profundus all patients had lobular panniculitis with supporting features of either hyaline fat necrosis, which is characteristic of lupus profundus (9,10,12), and/or epidermal and dermal changes diagnostic of DLE. In another study, the majority of cases showed paraseptal and lobular inflammation, ranging from focal lymphocytic inflammation to diffuse lymphocytic inflammation in the fat (4). Common features in our patient were hyaline fat necrosis, predominant lymphocytic inflammation mostly in fat lobules and paraseptal regions, and mucin deposition.

Lupus profundus is a subset of LE with distinctive clinicopathologic features. It tends to have a mild disease course, similar to DLE, in the majority of cases. DIF is usually positive in 50% of the patients. A positive ANA appears to indicate a high probability of systemic involvement (4). Occasionally, subcutaneous nodules of lupus profundus are the only clinical manifestation of LE (11). Trauma to subcutaneous fat seems to be a precipitating factor for lupus profundus lesions. Some patients develop the lesions at the point of previous injections, and aggravation of the process at the site from which a biopsy is taken or recurrence of the subcutaneous nodules along scars of previous excisions are distinctive clinical features of lupus profundus (9). In our patient, the clinicopathologic features were consistent with lupus profundus, DIF and ANA were negative, and there was no history of trauma or previous injection. On the contrary, her lesion regressed following biopsy.

We report this case in order to bring the clinicopathologic difference between lupus profundus and subcutaneous dermatofibroma to the attention of clinicians, and to review the literature about these 2 entities.

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