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Case Report

Lupus erythematosus panniculitis: a case report

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Abstract A case of lupus erythematosus profundus (LEP), with classical involvement of cheeks and upper arm, is reported in a patient of discoid lupus erythematosus (DLE). The skin overlying lesions of LEP also showed features of DLE.

Key words
Lupus erythematosus panniculitis, lupus erythematosus profundus.

Introduction

Lupus erythematosus panniculitis or profundus (LEP), an uncommon variant in the clinicopathological spectrum of lupus erythematosus (LE), is characterized by chronic inflammation and hyaline necrosis of subcutaneous tissue.[1] LEP may develop in association with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE)[2] or may occur as an isolated phenomenon. In most of the patients with lupus panniculitis, there will be preceding, subsequent, or concomitant lesions of discoid lupus erythematosus.[3] The lesions are most frequent on cheeks but other sites of predilection are face, upper arms, hands, chest, buttocks and thighs.[2] We report a classical presentation of LEP.

Case report

A 38-year-old woman presented to our outdoor patient department for the evaluation of a tender swelling on the right cheek since last three months. About 3 years back, she had a similar swelling on her left cheek which subsequently shrunk and receded till it formed a depression. She also had a similar nodular swelling on right upper arm that resolved spontaneously with progressive loss of superficial tissue and atrophy. She denied any fever, arthralgia, myalgia, fatigue, fever, Raynaud's symptoms or gastrointestinal symptoms. There was no family history of any autoimmune diseases.

Clinical examination revealed a middle-aged woman, with normal vital signs. There was an indurated swelling (4cmx4cm) on the right cheek which was mildly tender. Marked lipoatrophy was noted on the left cheek (till contour of maxillary bone appeared prominent) and a skin-coloured, atrophic, scarred plaque was noted on the right upper arm. She had a large patch of scarring alopecia on right temporal region (**Figure 1** and **Figure 2**).

Complete blood count with differential analysis, blood urea nitrogen, serum creatinine, liver function tests, urinalysis, chest radiograph and echocardiograph were normal. The serology of the patient's ANA was 1:160 but her dsDNA antibody was negative. We

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Figure 1 Scarring alopecia on right temporal region, indurated swelling on right cheek and lipoatrophy on right upper arm.

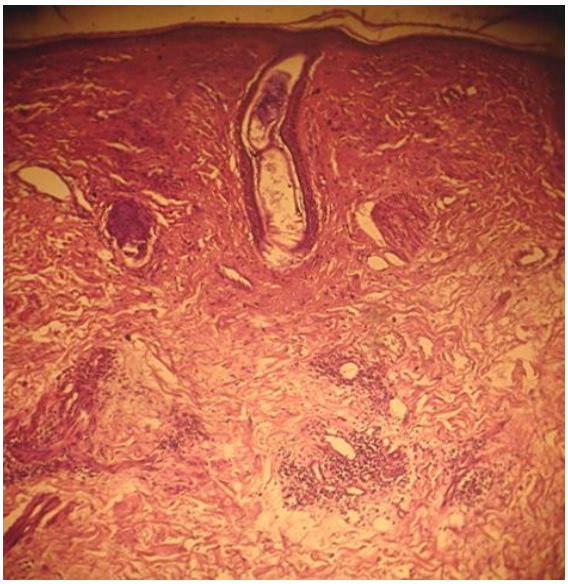

Figure 3 Epidermis shows atrophy and a thickened basement membrane. The dermis shows perivascular and periappendageal inflammatory infiltrate of lymphocytes (H&E, X 10).

Figure 2 Mildly tender swelling on right cheek and a receded left cheek due to lipoatrophy.

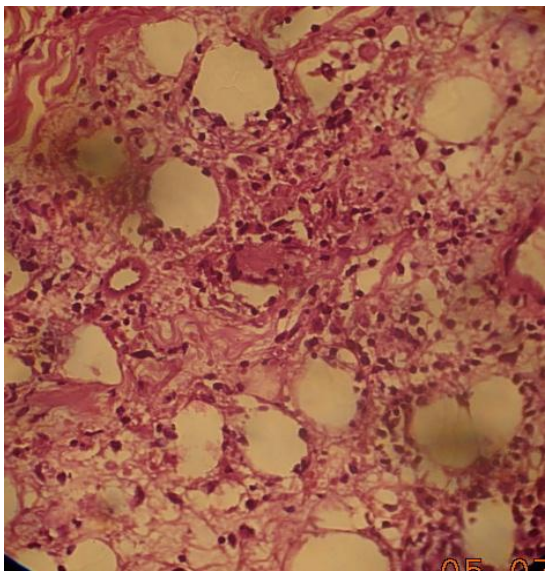

Figure 4 Subcutaneous tissue shows lobular panniculitis with dense infiltrates of lymphocytes and macrophages, and focal hyalinization.

could not perform immunofluorescent study because this facility was not available in our institute. Histopathology of biopsy from the

lesion of right cheek showed epidermal atrophy, hydropic degeneration of the basal cell layer, perivascular and periappendageal lymphocytic infiltration, a lobular panniculitis with dense infiltrates of lymphocytes and macrophages, and focal hyalinization of the adipocytes (**Figure 3** and **Figure 4**). Histopathology of biopsy sample from scarring alopecia was suggestive of DLE. Our case was diagnosed as lupus profundus. We treated her with chloroquine 250mg and prednisolone 20mg daily with complete subsidence of the nodule in 3 weeks. We, however, tapered the steroid after 6th week (completely stopped after 8 weeks) and continued with chloroquine only.

Discussion

Kaposi first described lupus erythematosus panniculitis, also known as lupus panniculitis and lupus profundus, in 1869. Arnold established it as a subtype of lupus erythematosus in 1956.[4] Clinically, the lesions of lupus panniculitis can resemble other forms of panniculitis, but the persistence of the lesions and the rarity of involvement of the distal extremities[5] help to distinguish it from conditions such as erythema nodosum or erythema induratum of Bazin. Lupus panniculitis with lipoatrophy should be distinguished from lipoatrophy caused by injections, factitial panniculitis, trauma and other forms of lipoatrophic panniculitis.

The histopathologic findings closely mimic that of discoid lupus erythematosus, in more than half of the patients. These consist of atrophy of the epidermis, vacuolar change at the dermoepidermal junction, thickened basement membrane, and superficial and deep perivascular inflammatory infiltrate of lymphocytes involving the dermis.[6,7] In rest of the cases, the changes are confined to the subcutaneous fat only, and are characterized by mostly lobular panniculitis. Hyaline necrosis is a hallmark of lupus panniculitis.[8] In our case, the histopathological features were consistent with those of DLE.

We assume that the swelling of her right cheek represented active lupus panniculitis while the depressed area on her left cheek represented localized lipoatrophy resulting from previous lupus panniculitis activity.

Antimalarials are the mainstay of treatment. Chloroquine or hydroxychloroquine (200 mg once or twice a day) is the first choice. Often a combination of antimalarials (for example, hydroxychloroquine 200 mg and quinacrine 100 mg daily) is used when monotherapy is ineffective.[8] However, appropriate time intervals (6 weeks to 3 months) must be given to allow these drugs to work maximally. Thalidomide can also be useful in rapidly controlling lupus panniculitis inflammation. Systemic steroids are generally not recommended for lupus panniculitis unless it is indicated for other manifestations for SLE. It is generally reserved for widespread and resistant lesions.[9] But as the panniculitis involving her left cheek persisted for three months and was gradually increasing, we decided to use a low dose of steroid (in combination with chloroquine) for the initial 1 month to prevent lipoatrophy. Intralesional steroid was not used as it might exacerbate the atrophic healing process.

Our patient was too poor to consider any cosmetic treatment for her shrunken left cheek. Autologous fat transfer or microlipoinjection would have been the best choice considering the depth of the lipoatrophy on her left cheek. The effects of this facial fat sculpting are very long-lasting and safe. They last significantly longer than those of synthetic fillers and there is no allergic reactions to worry about, because the fat transplantation is from own body. However, caution should be taken with any type of cosmetic manipulation in lupus

panniculitis for the fear of surgical trauma-induced ulceration or koebnerisation. It should be kept in mind that even therapeutic intralesional corticosteroid injections have been associated with ulcerative breakdown of lupus panniculitis lesions. Surgical correction of lipoatrophy should only be done after complete cessation of activity of the disease.

As the serological abnormalities are rare, the diagnosis of lupus profundus is confirmed primarily by both clinical and histologic findings. Aggarwal *et al*.[10] reported a case of LEP with associated mastitis, but without any lesions of discoid lupus erythematosus or systemic lupus erythematosus. When a biopsy specimen of clinically typical lesions of lupus panniculitis is not suggestive, the positive lupus band test along the dermoepidermal junction supports a diagnosis of lupus panniculitis.[11] In a very recent case report, the authors diagnosed a case of LEP with the aid of positive lupus band test.[12]

We report this case of lupus erythematosus profundus for its classical presentation with involvement of face and upper arm. It was associated with scarring alopecia of DLE.

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