CASE REPORT

LUPUS PANNICULITIS INVOLVING THE PAROTID/PERIPAROTID REGIONS AND BREAST; A RARE PRESENTATION

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INTRODUCTION

Kaposi first described lupus erythematosus panniculitis, also known as lupus panniculitis and lupus profundus, in 1869, and Arnold established it as a subtype of lupus erythematosus in 1956.[1] It is a chronic relapsing panniculitis that occurs in about 1 to 3% of patients with cutaneous lupus erythematosus and it is characterized by the development of deeply situated subcutaneous nodules and/or plaques with a predilection for the upper arms, face, shoulders and buttocks.[2,3] Rarity of involvement of the lower extremities distinguishes this condition from other forms of panniculitis, particularly erythema nodosum[4] Lupus panniculitis may occur as a separate entity or in association with systemic lupus erythematosus or discoid lupus erythematosus.[5] As the serological abnormalities are rare, the diagnosis is based on the clinical and histological findings. Antimalarials and corticosteroids are the mainstay of treatment but their real worth is still to be determined.[4] We report a chronic case of lupus pannicultis in a middle-aged woman affecting her face including the left parotid/periparotid region, left upper arm and right breast. Breast and parotid/periparotid areas are less commonly involved in lupus panniculitis.[6,7] To the best our knowledge, only a few cases of lupus panniculitis involving the parotid/periparotid region have previously been reported in the literature.[8]

CASE REPORT

A 45-year-old woman presented at the Ayza Skin & Research Centre, Pakistan for the evaluation of her facial disfigurement and abnormal contours of the left upper arm and right breast. She had a twelve-year history of developing multiple nodular swellings over the face, right breast and left arm associated with pain and tenderness. The nodules healed with progressive loss of superficial tissue and subsequent furrowing of the face over a few years. She also complained of multiple joint pains, myalgia and fatigue from the onset of her cutaneous symptoms. Her past medical history was unremarkable and there was no history of fever or Raynaud's phenomenon. Moreover, there was no family history of any autoimmune diseases. Clinical examination revealed a middle-aged woman, with normal vital signs. Her face was hollowed to the contours of her facial bones due to the presence of two large irregular atrophic plaques, one over the right malar area (Fig-1) and the other one over the left outer cheek extending onto the parotid/periparotid region (Fig-2). Patchy loss of subcutaneous tissue was also found involving the outer aspect of left arm and the upper outer quadrant of right breast.

Fig-1: Atrophic plaque over right malar area

Fig-2: Atrophic plaque left cheek extending onto the parotid/periparotid region

The histopathology of the biopsy from the trunk lesions showed a lobular panniculitis with a dense infiltrate of lymphocytes and macrophages. Focal hyalinization of the adipocytes was also evident.

The erythrocyte sedimentation rate was 20-mm/hr. Complete blood count with differential analysis, blood urea nitrogen, serum creatinine, liver function tests, urinalysis, and chest radiograph were normal. Antinuclear antibodies were positive but antibodies to double stranded DNA was negative. She was treated with oral indomethacin, hydroxychloroquine 250 mg x OD and topical fluocinolone acetonide.

DISCUSSION

Lupus panniculitis is a variant of lupus erythematosus that primarily affects subcutaneous fat. It occurs in 1 to 3% of patients with cutaneous lupus erythematosus.[2] First described by Kaposi in 1869, lupus panniculitis is characterized by the development of deeply situated subcutaneous nodules or plaques which have a predilection for the upper

arms, shoulders, face, and buttocks.[3] Less commonly involved areas include subcutaneous fat of the breast, orbital, and periparotid fat tissue.[6-8]Lupus panniculitis characteristically may involve the face but specific parotid and periparotid involvement is rare.[8]

The lesions may be tender and painful and frequently heal with atrophy and scars. In most of the patients with lupus panniculitis, there will be preceding, subsequent, or concomitant lesions of discoid lupus erythematosus.[5] Most patients are adults between 20 and 60 years of age, with a female to male ratio of approximately 2:1.[4]

Physical examination reveals deeply situated subcutaneous nodules or plaques of variable sizes. The surface of the nodules may show the classic features of discoid lupus erythematosus or may appear as normal skin. When the lesions regress, they leave atrophy and swelling of the involved area. Persistent areas of lipoatrophy on the shoulders and upper arms are characteristic of lupus panniculitis.

Histopathologic findings, which closely mimic those of discoid lupus erythematosus, in more than half of the patients, consist of atrophy of the epidermis, vacuolar change at the dermoepidermal junction, thickened basement membrane, interstitial mucin between collagen bundles of the dermis, and superficial and deep perivascular inflammatory infiltrate of lymphocytes involving the dermis.[9,10] In the rest of the cases, the epidermis and dermis remain uninvolved and the changes are confined to the subcutaneous fat, and are characterized by mostly lobular panniculitis with predominantly a lymphocytic infiltrate.[10]

The diagnosis is confirmed primarily by both clinical and histologic findings. When a biopsy specimen of clinically typical lesions of lupus panniculitis does not show definitive findings, the positive lupus band test along the dermoepidermal junction supports a diagnosis of lupus panniculitis.[11]

Although often normal, serological analysis may show a positive antinuclear antibody titer. Less frequently anti-double stranded DNA antibodies will be present. Syphilis serology may be falsely positive. Other possible laboratory findings are lymphopenia, anemia, reduction of C4 levels, and positive rheumatoid factor.[12]

The exact pathophysiologic mechanism responsible for the production of lupus panniculitis remains unknown. Trauma to subcutaneous fat seems to be a precipitating factor for the lesions of lupus panniculitis. The lesions may develop at the point of previous injections, and the aggravation of the process at the site of a biopsy or recurrence of the subcutaneous nodules along scars of previous excisions have been reported as distinctive clinical features of lupus panniculitis.[11]

Clinically, the lesions of lupus panniculitis can resemble other forms of panniculitis, but the rarity of involvement of the distal extremities[4] helps to distinguish it from conditions such as erythema nodosum or erythema induratum of Bazin. Other types of subcutaneous inflammation, which are associated with lupus erythematosus, include erythema nodosum, thrombophlebitis and pancreatic panniculitis.

Histopathologically, morphea panniculitis, traumatic panniculitis, or localized lipoatrophy, closely resemble lupus panniculitis. The presence of lymphoid follicles seems to be a common feature of both morphea and lupus panniculitis, but these are more abundant in case of lupus panniculitis. Moreover, hyaline necrosis is hallmark of lupus panniculitis.[13] In cases of traumatic panniculitis, a nidus of subcutaneous inflammation and other features of extrinsic injury are usually present.

To summarize, the characteristic histopathologic features of lupus panniculitis including lipoatrophy, hyaline necrosis and lymphoid follicles, greatly help distinguish lupus panniculitis from other forms of panniculitis.

Local treatment with potent corticosteroids under occlusion has been reported as being helpful in lesions of lupus panniculitis, but often a systemic course of corticosteroids or hydroxychloroquine is necessary.[4] Dapsone has also been reported as effective in the management of lupus panniculitis[4,14]

Lupus panniculitis often responds to treatment with antimalarials, such as hydroxychloroquine (200 mg once or twice a day).[13] Some cases respond to a combination of antimalarials (for example, hydroxychloroquine 200 mg and quinacrine 100 mg daily) when monotherapy is ineffective.[13] Systemic glucocorticoids should be reserved for widespread and resistant lesions.[15] Intralesional glucocorticoids are usually ineffective and may exacerbate the atrophic healing process. Success with dapsone, azathioprine, and thalidomide has been described in isolated case reports.[4] Supportive measures include rest, avoidance of trauma, and ultraviolet light protection. When all other modalities have failed, surgical debridement or resection of individual lesions may be attempted.

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