Bilateral Facial Lipoatrophy-As Sole Manifestation of Lupus Panniculitis

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Abstract :

Lupus erythematosus can have varied manifestation in skin ranging from classical lesion of DLE (Discoid Lupus Erythematosus) and photosensitivity to Necrotic lesions of vasculitis or atrophic lesion. Panniculitis as histological finding in biopsy taken from any lesion of SLE (Systemic Lupus Erythematosus) is expected as associated finding.

We report one case with lipoatrophy as sole manifestation of lupus panniculitis in which patient presented to us with bilateral facial lipoatrophy. On further investigation patient was ANA (Antinuclear Antibody) positive and histopathology was also suggestive of lupus panniculitis. LE (Lupus Erythematous) presenting solely as lupus panniculitis is rare clinical manifestation and poses a considerable diagnostic challenge more so when presentation is lipoatropy.

Key Words : Bilateral Facial Lipoatrophy, Lupus Panniculitis

Introduction :

Lupus panniculitis is a chronic recurrent panniculitis that is more frequent in women, with a median age of onset being 30-40 year. It is a mild variant of LE (lupus erythematosus), and most patients show subcutaneous nodules as the only clinical manifestation of the disease. Lupus panniculitis often occurs prior to other manifestations of LE.[1] The lesions exhibit predilection for the face, shoulders, upper arms, back, and buttocks, areas infrequently involved in other forms of panniculitis. [2] The serum ANA (Antinuclear antibody) is usually positive in approximately 70% of cases. It arises spontaneously but can be triggered by injury such as immunization; local injections of glatiramer acetate for multiple sclerosis have induced lupus panniculitis-like histology in some instances. Hepatitis B vaccination has reactivated old lesions of lupus profundus, while interferon-beta has induced new ones. The lesions can be painful and may ulcerate, leading to atrophy and scarring after healing, even giving an anetoderma-like appearance. Lupus panniculitis can overlap with morphoea-like lesions, dermatomyositis or other forms of connective tissue panniculitis.[3]

The major histological criteria needed for a diagnosis of lupus panniculitis include hyaline necrosis of fat, lymphocytic aggregates or lymphoid follicle formation, periseptal or lobular lymphocytic panniculitis, and calcification. Minor changes (not necessary for diagnosis) include overlying changes of discoid LE, hyalinization of the subepidermal zone, mucin deposition, lymphocytic vascular inflammation and collections of plasma cells and eosinophils. Karyorrhexis from lymphocyte nuclear dust is common and a strong pointer.[3-5]

Connective tissue panniculitis is a rare form of panniculitis, affecting septae and lobules, and leading to prominent lipoatrophy. The condition has been described in female adults and children. Episodes of intense lymphocytic panniculitis lead to lipoatrophy. At various times in the illness, a circulating ANA and/or SSb (la) antibodies may be detected. The clinical spectrum of lipoatrophic panniculitis is thought to encompass connective tissue panniculitis. Histologically, in connective tissue panniculitis there is no evidence of hyalinization of fat and collagen, which is typical of lupus erythematosusprofundus.[3]

Case report

A 40 year old female presented to us with complain of bilateral hollowing of cheek for last few months (Photograph-one and two). She had history of tooth extraction of right lower molar and local anesthetic injection for the same six months back before starting the lesion. Patient was correlating the start of her cheek lesions following that. She gave history that lesion started on right side as painful swelling followed by loss of tissue and hollowing. Gradually lesion started on left side also. She was having history of pain in small joints of hands and in knee joint. She also gave history of weight loss around 10 kg in last three months. There was no other complains in terms of photosensitivity, oral ulcers or any other skin lesions. We put the differential diagnosis of Lupus Panniculitis and HIV associated lipoatrophy. All routine reports were within normal limits. Tests for HIV (Human Immunodeficiency Virus) were nonreactive. ANA was positive (+two). Ds-DNA (Double-stranded Deoxy-rebonucleic acid) was negative. On histopathological examination (Photograph three) from the biopsy taken from the indurated edge on the left side of her face showed basal cell vacuolization, perivascular mild lymphocytic infiltrate, lobular and septal lymphocytic infiltrate in subcutaneous tissue with hyaline degeneration of fat cells.

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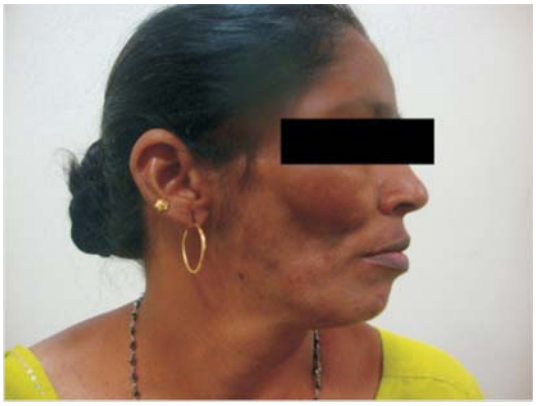
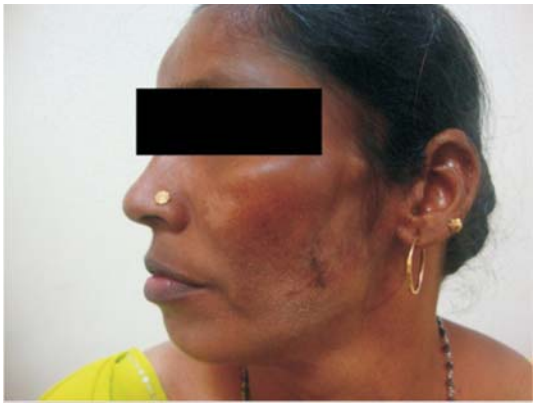
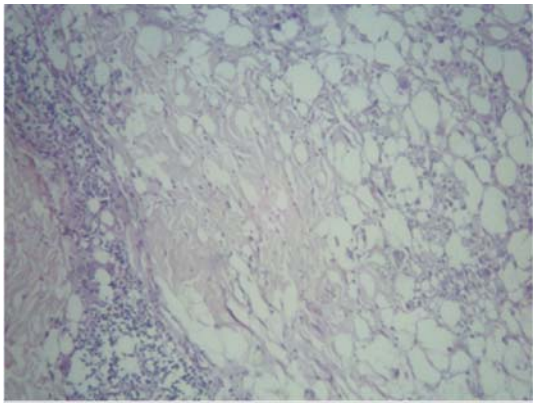
Photograph 1: Complete lipoatrophy on the right side of face



Photograph 2: Partial lipoatrophy with indurated edges on left side of face



Photograph 3: Septal and lobular panniculitis with hyalinization of fat



Discussion

Till date only few cases of facial lipoatrophy as sole manifestation of lupus panniculitis has been reported. Hawilo A et al [6] has reported a case with unilateral facial lipoatrophy whitch was suggestive of lupus panniculitis. Sometime lupus panniculitis can also present as initial manifestation of LE [7]. Usually lupus panniculitis presents as subcutaneous nodules along with typical features of discoid LE. Lipoatrophy has been common with other type of connective tissue panniculiitis other than lupus panniculitis. There was history of local anesthetic injection for tooth extraction in our patient, but patient's ANA was positive so it might be just association by chance or injection trauma being triggering factor. Our patient was started on oral steroids in dose of 1mg/kg which was tapered gradually and hydroxy chloroquine in dose of 200 mg 12 hourly and it was continued till patient lost to follow up. Disease progression was stopped further on treatment. Early identification of the disease is necessary as facial lipoatrophy is disfiguring.

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