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Case Report

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Disfiguring lupus panniculitis: case report and review of the literature

Abstract

Lupus panniculitis also called lupus profound is a rare variant of chronic cutaneous lupus. This entity share histological characteristics with subcutaneous T cell lymphoma, which require a lot of times, a clinical biological and histological correlation. This entity is difficult to treat and oral corticoids seem to be the mainstay therapy. We report a case of a woman who presented systemic lupus erythematous with a cutaneous involvement as panniculitis and a good response to antimalarials.

Keywords: lupus, panniculitis, atrophy, lymphocytic infiltrate

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Abbreviation: LP: lupus panniculitis

Introduction

Lupus panniculitis or lupus profound is a rare variant of chronic cutaneous lupus, it can be isolated, or associated with a chronic erythematous or systemic lupus, the diagnosis requires a clinical, biological and histological correlation. This entity is difficult to treat and oral corticoids seem to be the mainstay therapy. We report a case of a woman who presented systemic lupus erythematous with a cutaneous involvement as panniculitis.

Case observation

A woman of 52 years old, was followed for chronic anemia, and receiving martial supplementation, with no history of photosensitization. Presented for four years, painful lesions on her face, arms, buttocks, consulting, one year later, those lesions have regressed with facial deformities, weight loss. Dermatological examination noted several Subcutaneous nodules of different sizes with erythematous and pigmented surfaces sitting in the proximal part of the arms and limbs, the lower back and buttocks. Diffuse atrophic plaques were noted in the buttocks, back, and thighs (Figure 1). The face was marked by deep atrophy of cheeks with the dissolve of 'boule de Bichat' (Figure 2).

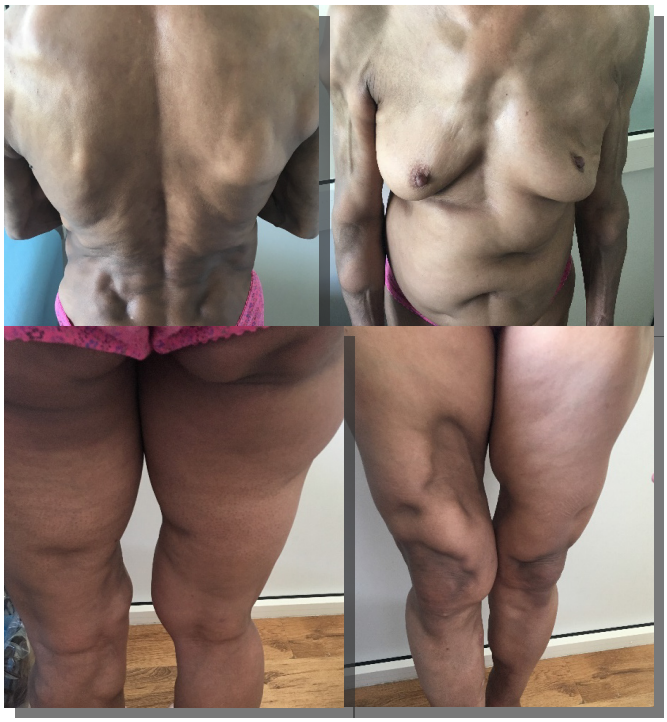

Figure 1 Deep atrophy on different fatty locations.

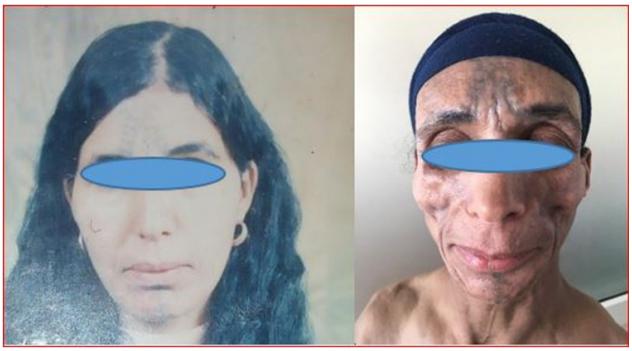

Figure 2 Dissolve of 'Boule de bichat' with deep cheeks atrophy (figure A) in comparing with a patient's picture before disease onset.

When examining the scalp, we noted a diffuse low hair density, with linear comedian scars, at the left temporal-frontal boss (Figure 3). Periungual dermoscopy did not show any abnormalities, and the rest of the somatic examination was correct. This clinical presentation made us evoke the lupus panniculitis, panniculitis-like T Cell lymphoma, and Weber-Christian disease. Our patient has benefited from a skin biopsy that showed a lymphohistiocytic infiltrate of the lobules with negative immunohistochemistry, this aspect was compatible with an LP. The Biological assessment of systematization was negative. The patient received hydroxychloroquine at the dose of 400mg per day without new lesions.

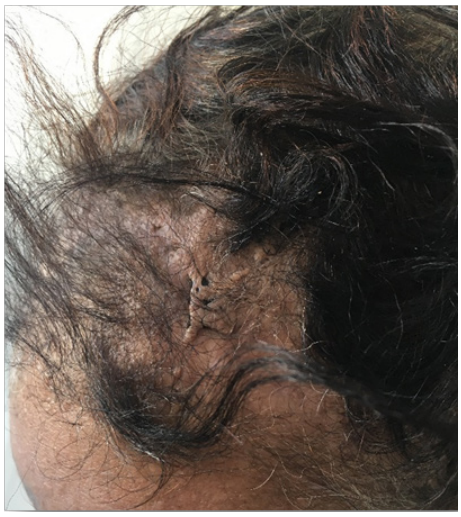

Figure 3 Comedonian scars after regression of a nodule of the scalpe.

Discussion

LP is a chronic recurrent inflammation of subcutaneous fat. In 1988, Kaposi described nodules in a patient having lupus with systemic manifestations.[1] In 1956, it was described as a patient presenting lupus panniculitis in the absence of an underlying discoid lupus erythematosus.[2] It's defined as a variant form of lupus erythematous that may be the unique manifestation, be associated with discoid lupus erythematosus or systemic lupus erythematosus.[3] This entity affects more females than males with a female-male ration variant between 2:1[3] and 9:1[4]referring to different series.[5] The age of a rise of this disease is variable between 30 and 60 years,[5] pediatric forms are rare[6] and neonatal lupus associated with lupus panniculitis is exceptional.[7,8] The classical presentation is an erythematous plaque or nodule that arose most frequently in proximal extremities, particularly the lateral aspects of the arms and shoulders, buttocks, trunk, face, scalp, and breast.[9,10] Legs are an unusual location for lupus panniculitis, that helps to differentiate it from other panniculitis affecting specifically this body site[11] in pediatric patients the facial location is most frequent.[12] For instance, lesions appear in a previous site of traumatism, injection or biopsy site.[13] After resolution of erythematous nodules, it persists lipoatrophic areas that help for a retrospective diagnosis[5,14] as was the case of our patient. The histologic features of lupus panniculitis are the presence of focal or diffused lymphocytic infiltrate on the dermis and lobular fat, with the peri-adnexal disposition.[15] Referred to the author's opinion, there are two major criteria for histological diagnosis of LP: f lymphocytic infiltrate involving fat lobules and hyaline necrosis of the fat lobule.[5] The management of LP is difficult; this profound variety is usually resistant to antimalarials. The use of thalidomide is limited by its teratogenicity and other severe side effects. Then oral corticotherapy consists of the principal effective treatment of this disease despite its side effects.[5] There is some anecdotal use of immunosuppressive therapy to maintain remission, we report azathioprine, mycophenolate mofetil, cyclophosphamide, and cyclosporine.[16] Recently, a report of LP refractory to diverse therapies has been successfully managed with Rituximab.[17] After long-term remission, a cosmetic procedure such as fat grafts may be suggested with prudence because of the risk of unhealing ulcers.

Conclusion

LP is a rare variant of Lupus erythematous, with clinical and histological specificities, this disease presents a therapeutic challenge and atrophic lesions are definitive, making a retrospective diagnosis.

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