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Case report

LUPUS PANNICULITIS - A CASE REPORT WITH LONG-TERM FOLLOW-UP

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Abstract

Lupus erythematosus panniculitis (LEP), also known as lupus erythematosus profundus, is a rare subtype of chronic cutaneous lupus erythematosus (CCLE), characterized by inflammation of the subcutaneous adipose tissue. It is estimated to affect 1-3% of patients diagnosed with cutaneous lupus. It can present as a distinct clinical entity or in association with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE).

We report a case of lupus panniculitis with long-term follow-up in a 54-year-old woman, initially presenting with unilateral periorbital erythema and edema. The patient's medical history revealed intermittent episodes of unilateral periorbital edema on the affected side over the previous three years along with the development of an erythematous, indurated lesion on the right cheek, which resolved with an atrophic, depressed plaque. During this period, the patient had been treated with oral antibiotics, corticosteroids, and antiviral therapy due to suspected clinical diagnoses of contact dermatitis, recurrent herpes simplex, and periorbital cellulitis. Considering the patient's history, clinical presentation, histological evidence of lymphocytic lobular panniculitis, and immunological findings, a diagnosis of lupus panniculitis was established.

The patient demonstrated an excellent response to treatment with hydroxychloroquine and prednisolone, achieving complete resolution of her dermatological symptoms. However, after a three-year period without medical supervision and discontinuation of therapy, she presented with new lesions on the facial region, including one on the nose suspected to be DLE and multiple atrophic, depressed plaques on the upper arms.

Keywords: lupus panniculitis, periorbital edema, discoid lupus erythematosus, hydroxychloroquine

Introduction

Lupus erythematosus panniculitis (LEP), also known as lupus erythematosus profundus, is a rare subtype of chronic cutaneous lupus erythematosus (CCLE), characterized by inflammation of the subcutaneous adipose tissue[1]. Initially it was described by Kaposi in 1883 and was subsequently recognized as a distinct clinical entity by Irgang in 1940[2,3]. It is estimated to affect 1-3% of patients diagnosed with cutaneous lupus[4].

The cutaneous manifestations of lupus panniculitis typically include persistent subcutaneous indurated plaques or firm nodules, which often heal with atrophy and residual scarring. Lesions most commonly appear on the upper arms, shoulders, breasts, buttocks, and thighs, as well as on the face - particularly the forehead and cheeks[5].

Lupus panniculitis generally occurs in the third to sixth decades of life, with a notable predilection for females. It may present as an isolated clinical entity or in association with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE)[6].

We report a case of lupus panniculitis with long-term follow-up in a 54-year-old woman, initially presenting with unilateral periorbital erythema and edema.

Case report

A 54-year-old female patient presented to our University Clinic for Dermatology with persistent erythema and edema in the right periorbital region over the past three months. The patient reported a history of intermittent unilateral periorbital edema on the affected side over the past three years, as well as an erythematous, indurated lesion on the right cheek that healed with the formation of a depressed, atrophic plaque.

On examination, there was pronounced edema of the right upper and lower eyelids, with a soft consistency on palpation and erythematous discoloration of the affected skin. The lesion itself was asymptomatic. Additionally, an atrophic, depressed plaque was present in the right mandibular region. The rest of the physical examination was normal, and the patient did not report any systemic complaints or other illnesses.

Over the years, the patient had undergone medical evaluation, with suspected diagnoses including contact dermatitis, periorbital cellulitis, and recurrent herpes simplex. Accordingly, she had been treated with oral antibiotics, corticosteroids, and antiviral therapy. A short course of prednisone had led to rapid regression of the edema, but recurrent exacerbation occurred upon discontinuation of treatment. The other mentioned treatment attempts had been ineffective.

Laboratory tests, rheumatological and ophthalmological evaluations, chest X-ray, sinus X-ray, MR imaging of the paranasal sinuses, and biopsies were performed.

Histopathological examination of the biopsies from the upper and lower eyelids revealed a dense mononuclear infiltrate within the lobules of subcutaneous adipose tissue. Adipocyte necrosis was observed in the lobules, and the infiltrate was composed entirely of mononuclear cells, including clusters of lymphocytes with hyperchromatic nuclei. Lymphocyte nuclear dust was also observed in certain areas. The epidermis appeared without pathological alterations. Perivascular infiltrate composed of lymphocytes in an edematous dermis was present. Immunohistochemical analysis showed focal aggregates of CD20+ B lymphocytes, diffusely distributed CD3+ T lymphocytes (CD4+/CD8+), and CD138+ plasma cells.

Immunological testing revealed positive ANA antibodies with a titer of 1:180 and nRNP/Sm antibodies at 93 (<10). Tests for SSA, SSB, anti-Scl70, RF, C3, and C4 were negative. MRI demonstrated edema of the periorbital soft tissues. Rheumatological and ophthalmological evaluations, and chest and sinus X-rays demonstrated normal findings.

Considering the patient's history, clinical presentation, histological evidence of lymphocytic lobular panniculitis, and immunological findings, a diagnosis of lupus panniculitis was established.

The patient was started on hydroxychloroquine at a dose of 400 mg/day and prednisolone at an initial dose of 50 mg/day, which was gradually tapered. After three months of treatment, complete resolution of dermatological manifestations was achieved. However, a relapse occurred

following the discontinuation of therapy. The patient was subsequently reinitiated on hydroxychloroquine at a dose of 200 mg/day, leading to complete clinical remission over the course of the following year (Figure 1).

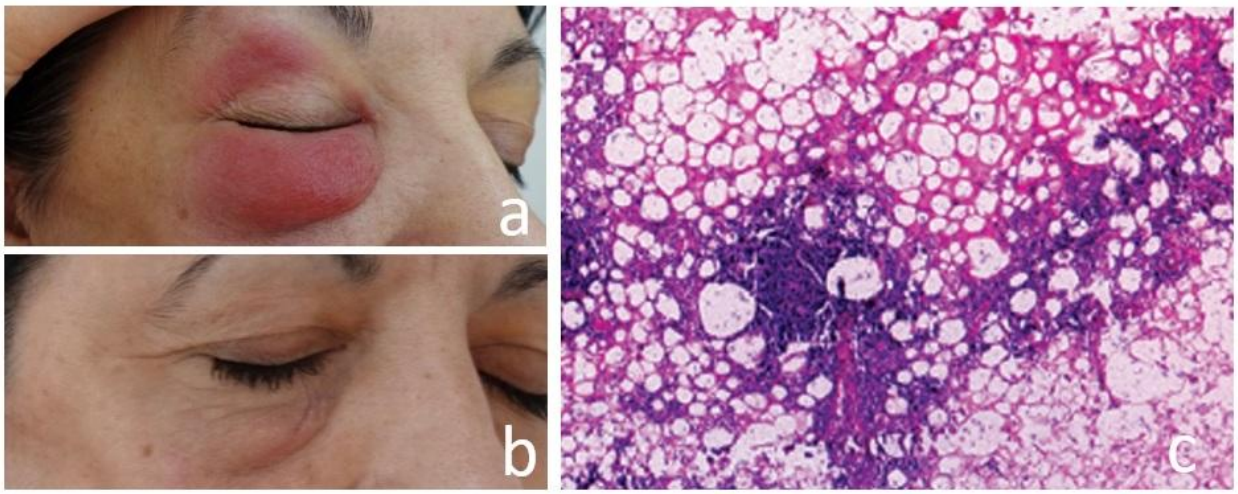

Fig. 1. (a) Patient initially presenting with unilateral periorbital erythema and edema (b) Complete clinical remission achieved after treatment with hydroxychloroquine, with no recurrence observed for ten months (c) Histopathological finding of lymphocytic lobular panniculitis

Three years later, the patient presented again with new lesions in the facial region, including one on the nose suspected to be discoid lupus erythematosus (DLE), along with multiple atrophic, depressed plaques localized on the upper arms. It came to our knowledge that the patient had discontinued the therapy, and the new lesions had developed over the course of the past year (Figure 2).

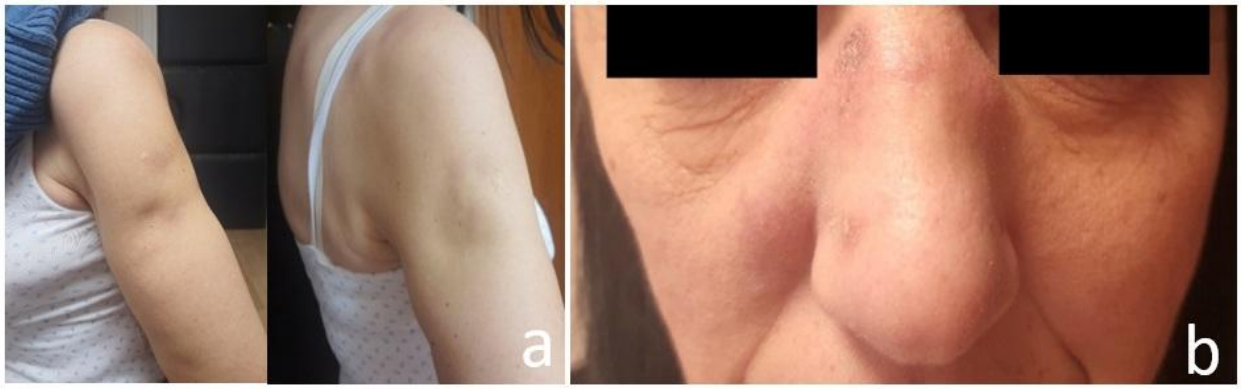

Fig. 2. (a) After three years without medical follow-up and complete discontinuation of therapy, the patient presented with: multiple atrophic, depressed plaques on the both upper arms
(b) New lesions on the facial region, including one on the nose suspected to be DLE

Discussion

Lupus panniculitis is a rare subtype of chronic cutaneous lupus erythematosus, characterized by inflammation of the subcutaneous adipose tissue[1]. Typically, lesions tend to develop in the specific predilection areas[7]. Nevertheless, they may also present in other locations, as observed in our patient, who exhibited a rare initial manifestation of lupus panniculitis, characterized by unilateral erythema and edema in the periorbital region. According to the available literature, there have been reported only five cases of lupus panniculitis with an initial presentation of unilateral periorbital erythema and edema[5,8-12].

The differential diagnoses considered included infections (cellulitis, lupus vulgaris, Kikuchi disease), thyroid orbitopathy, idiopathic orbital inflammation, sarcoidosis, eyelid malignancy (lymphoma, sinus histiocytosis, M. Castleman disease, sarcomas), IgG4-related orbital inflammation, granulomatosis with polyangiitis and lupus panniculitis[13-15]. After conducting the paraclinical investigations, we ruled out other differential diagnoses and confirmed the diagnosis of lupus panniculitis with histological analysis.

The gold standard of lupus panniculitis diagnosis is the histopathology examination result from a deep skin biopsy of the lesional area[16,17]. It shows lobular panniculitis with dense lymphoid cell infiltrate and mucin deposits as well as fat necrosis and histiocytes with phagocytosed nuclear debris in the deeper dermis and subcutaneous tissue. According to Ackermann *et al.,* and other authors, the presence of lymphocytic nuclear dust within a patchy lymphoplasmacytic infiltrate in the lobules of the subcutaneous fat is a clue for the histopathologic diagnosis of lupus panniculitis [4]. The key histopathological findings necessary to establish the diagnosis were present in our patient's biopsy. Ideally, a biopsy should have been performed promptly after it became evident that the patient did not respond to treatment for the previously suspected diagnoses.

With regard to the laboratory findings, approximately 75% of patients show elevated ANA, although specific autoantibodies are usually not detectable[5]. As seen in our patient, ANA antibodies were also found to be positive.

Around 30-50% of patients have evidence of systemic lupus erythematosus (SLE), and lupus panniculitis may precede the development of SLE by several years[1,18]. Our patient did not meet the criteria for systemic lupus erythematosus at any point during the follow-up until this day.

Also, lupus panniculitis lesions can appear with surface changes including erythema and discoid lupus erythematosus (DLE) features as observed in the new lesion in our patient. Approximately 70% of patients with this type of CCLE also have typical DLE lesions, often overlying the panniculitis lesions[5]. Some have used the term lupus profundus to designate those patients who have both lupus panniculitis and DLE lesions, and lupus panniculitis to refer to those having only subcutaneous involvement[19].

The primary treatment approach for lupus panniculitis involves the use of antimalarials, as first-line therapy[6,19]. Therefore, we selected hydroxychloroquine, as a treatment of choice, which yielded satisfactory results in our patient. Alternative treatment options include oral corticosteroids, thalidomide, dapsone and immunosuppressive agents like methotrexate [6,9,17,19]. Furthermore, there has been a documented case demonstrating successful therapeutic outcome with tacrolimus[9].

Conclusion

This case highlights the diagnostic challenges, the importance of early identification and timely treatment of the disease, and emphasizes the significance of patient education, adherence to treatment, as well as the necessity for long-term and regular follow-up as key factors in effective disease management and prevention of further progression.

Conflict of interest statement. None declared.

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