Lupus Erythematosus Panniculitis Presenting as Palpebral Edema and Parotiditis

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Abstract. Lupus erythematosus panniculitis or lupus erythematosus profundus is characterized by inflammation of the deep dermis and subcutaneous tissue. It can occur in isolation or associated with chronic systemic or discoid lupus erythematosus. It usually consists of nodules and hardened subcutaneous plaques on the forehead, cheeks, proximal extremities, and buttocks. Periorbital and parotid involvement are rare and can lead to misdiagnosis. We present the case of a patient with lupus erythematosus panniculitis who presented with palpebral edema and involvement of the periocular fat and parotid gland.

Key words: lupus erythematosus profundus, lupus panniculitis, palpebral, edema, parotid gland.

Introduction

Lupus panniculitis or lupus erythematosus profundus is an inflammatory disease involving the deep dermis and subcutaneous cellular tissue. It may present in isolation or occur simultaneously or prior to manifestations of systemic or discoid lupus erythematosus.1

Clinical manifestation is characterized by nodules and hardened subcutaneous plaques, often adhered to the overlying skin, that heal after a long period of time with atrophy and residual scarring. These nodules and plaques are usually located on the forehead, cheeks, proximal limbs, and buttocks,1,2 though cases have occasionally been reported with orbital2-5 and salivary-gland6,7 involvement. Problems with the differential diagnosis tend to arise in these atypical sites, particularly when the disease is not associated with manifestations of systemic or discoid lupus erythematosus.

We present the case of a patient with involvement of the parotid gland and of the eyelid and extraocular fat of the right eye.

Case Description

A 63-year-old man was referred to our department with edema and erythema of the right upper eyelid that had appeared 3 years previously. The only history of interest was xerostomia and xerophthalmia that had begun 4 years previously.

Examination of the patient revealed edema in the right upper eyelid, which was slack when palpated and the affected skin had a violaceous erythematous appearance (Figure 1). The lesion was asymptomatic, persistent, and was not associated with displacement of the eyeball or other structural abnormalities.

There was an erythematous plaque in the right mandibular region that had appeared 6 years previously and revealed a scarred and infiltrated appearance on palpation (Figure 2).
The patient reported previous involvement of the left upper eyelid that had resolved spontaneously. The rest of the physical examination was normal and the patient presented no other symptoms. The lesion in the right mandibular region had already been studied by the rheumatology department and the ear, nose, and throat department using fine-needle aspiration, magnetic resonance imaging (MRI), and sialography, and the patient had been diagnosed with chronic parotiditis.

We performed skin biopsies of both the right upper eyelid and the infiltrated plaque on the jaw and obtained matching histologic findings. The biopsies revealed a dense, predominately lymphoid infiltrate in the deep reticular dermis that had spread to the subcutaneous cellular tissue, with a discrete nodular-periadnexal distribution (Figure 3). The lymphocytes displayed no morphological abnormalities and were accompanied by xanthomatous histiocytes. In the subcutaneous cellular tissue, the infiltrate was distributed in the lobules and septa, with hyaline necrosis of the fat (Figure 4). The epidermis appeared to be unaffected. The immunohistochemical study showed that the lymphoid cells consisted of B and T lymphocytes, with some germinal centers. Direct immunofluorescence was negative.

These findings were compatible with a diagnosis of lupus erythematosus profundus.

In the analyses requested, antinuclear antibodies were positive with a punctate pattern (titer, 1:80), as were antiribonucleoprotein antibodies.

A maxillofacial and orbital MRI scan revealed thickening of the eyelid and extracanal fat in the upper external area of the right orbit, with no displacement of the eyeball. Both parotid glands presented heterogeneous areas of increased uptake with no increase in size. These abnormalities were reported as compatible with a lupoid infiltrate.

During follow-up, the patient underwent a cornea transplant and was administered oral corticosteroids to prevent rejection; this led to the resolution of the cutaneous lesions and improvement of the xerostomia. Subsequent follow-up examinations revealed persistence of the atrophic plaque in the right mandibular region. The palpebral edema and erythema resolved completely. Follow-up MRI scans confirmed that the orbital infiltrate had disappeared, with persistence of discrete inflammatory changes in both parotid glands.

**Discussion**

The clinical findings of this form of lupus were first reported in 1883 by Kaposi, although the terms lupus erythematosus profundus and lupus panniculitis were first introduced by Irgang in 1940.

Lupus panniculitis is a rare form of lupus that is characterized by involvement of the deep dermis and subcutaneous cellular tissue. It usually affects middle-aged women, though cases of pediatric patients have been published. The condition may appear in isolation or in association with manifestations of discoid or systemic lupus. Approximately 2%-7% of patients with systemic lupus develop lupus panniculitis. It has generally been observed that patients with lupus erythematosus lesions occasionally develop systemic manifestations and present abnormal test results.
Clinical findings include indurated subcutaneous plaques and nodules that, although initially movable, adhere to deep layers. These are persistent lesions that do not usually become ulcerated, though when ulceration does occasionally occur, they are painful.

The overlying skin may be intact or show discoid lupus erythematosus lesions in 20%-70% of cases. Our patient presented only erythema on the eyelid, whereas the skin of the right mandibular region showed erythema and areas of atrophic skin adhering to deep layers but there were no findings compatible with chronic discoid lupus erythematosus.

The disease usually affects the cheeks, proximal limbs, and the buttocks, though involvement of the breast (lupus mastitis), the eyelid, in the form of persistent unilateral edema, the orbit of the eye, beginning with proptosis, and the salivary glands have also been reported. The principal problem in these atypical cases is confusion essentially with neoplastic processes and connective-tissue diseases. In fact, the few reported cases of periorcular and parotid involvement are found in the ophthalmology and otorhinolaryngology literature, respectively. Differential diagnosis in the case of our patient included cutaneous infiltrate due to cancer, sarcoidosis, lymphoma, paraneoplastic dermatomyositis, and lupus erythematosus.

Our patient did not meet the criteria for systemic lupus erythematosus at any time during follow-up and parotid involvement was asymmetrical and without size increase. The patient had only presented xerostomia and xerophthalmia and the only abnormalities in the analyses were positive results for antinuclear antibodies and antinucleoprotein antibodies.

Another entity to be considered in the differential diagnosis is panniculitis-like T-cell lymphoma, which may occasionally produce very similar clinical and histologic findings. Histology reveals lobular lymphocytic panniculitis with hyaline necrosis of the fat and presence of lymphoid follicles adjacent to the septa, occasionally with germinal centers. Older lesions may contain calcium deposits. More than half of cases may present changes characteristic of lupus erythematosus in the dermis and epidermis, with epidermal atrophy, vacuolar degeneration of the basement membrane, and perivascular and periadnexal lymphocytic infiltrates. These findings are more frequent when the lesions are in association with discoid lupus. In our patient, the epidermis was unaffected; there were no apparent abnormalities in the epidermis, no mucin deposits, and no degeneration of the basement membrane.

Laboratory assays for antinuclear antibodies are positive in 70% of patients, as was the case with our patient. Cases have been reported of lupus erythematosus profundus in association with a congenital deficit of C2 and C4. Naturally, if the patient presents an associated systemic lupus, we will find the usual abnormalities in the analyses.

The treatment of choice consists of antimalarial drugs such as hydroxychloroquine at a dosage of 200 to 400 mg/d, which provides a clinical response in 6 to 8 weeks. Oral
corticosteroids and dapsone may also be used. Our patient improved rapidly following administration of oral corticosteroids to prevent rejection of the cornea transplant. When faced with a patient with persistent palpebral edema, ocular proptosis, or involvement of the salivary glands, lupus panniculitis should be considered in the differential diagnosis.

Conflicts of Interest
The authors declare no conflicts of interest.

References