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SHORT COMMUNICATION

Case report

A case of lichen planus-lupus erythematosus overlap syndrome with eyelid involvement

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PURPOSE. *To report a case of lichen planus-lupus erythematosus overlap syndrome with eyelid involvement. Lichen planus and lupus erythematosus infrequently coexist in the same patients. Ocular involvement has rarely been reported for both diseases.*
Case report. *We describe a case of lichen planus-lupus erythematosus overlap syndrome with eyelid involvement. Histopathologic and immunofluorescent studies were done on buccal, lip, left conjunctival, malar, auricular and scalp lesions. The immunopathologic features of the conjunctiva, buccal mucosa and lip were consistent with lichen planus, while those of the malar, auricular and scalp lesions favoured lupus erythematosus.*
RESULTS. *The patient was successfully treated with hydroxychloroquine 200 mg/day and all lesions responded to therapy within weeks.*
CONCLUSIONS. *This is a rare example of two coexisting autoimmune disease entities: lichen planus of the oral mucosa, lip, eyelid and discoid lupus erythematosus of the skin. To our knowledge, this is the first reported case of lichen planus-lupus erythematosus overlap syndrome with eyelid involvement. (Eur J Ophthalmol 2002; 12: 244-6)*

KEY WORDS. *Lichen planus, Discoid lupus erythematosus, Overlap syndrome, Eyelid involvement*

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INTRODUCTION

Lichen planus-lupus erythematosus overlap syndrome (LP-LE) involves lesions that share concomitant clinical and histologic features of both LP and LE. The cause of this overlap syndrome is unknown, but evidence points to an autoimmune mechanism (1). To date, fewer than 50 cases have been reported (2). Most of these patients have atrophic discoid lupus erythematosus (DLE) lesions on the head, neck and trunk, and lacy white plaques on the oral mucosa (1). Eyelid involvement has been described rarely in isolated cases of LP and LE (3-7). We report a patient with LP-LE overlap syndrome, who had left lower eyelid involvement and immunohistopathologic features consistent with both LE and LP.

Case report

A 34-year-old woman presented with skin and mucosal lesions of nine years duration. On dermatological examination, she had violaceous, thickened, scaly lesions on scalp, arms, malar and auricular regions and her buccal and lip mucosa showed white papules with a reticular pattern (Figs. 1, 2). She had an erythematous, well-defined, mildly raised plaque on her left lower eyelid involving one-third of the lateral portion, of two week's duration (Fig. 3). Examination of the right eyelid revealed no abnormalities. Biopsies of the eyelid, lip, buccal mucosa, malar and scalp lesions were taken. A specimen of the lid revealed hydropic degeneration of the basal layer, moderate lymphocytic inflamma-

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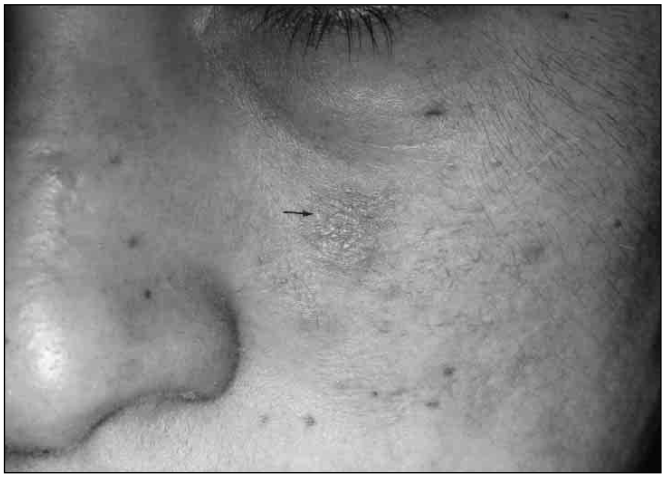

Fig. 1 - *Sharply demarcated, slightly indurated, erythematous discoid lesions on the malar aspect.*

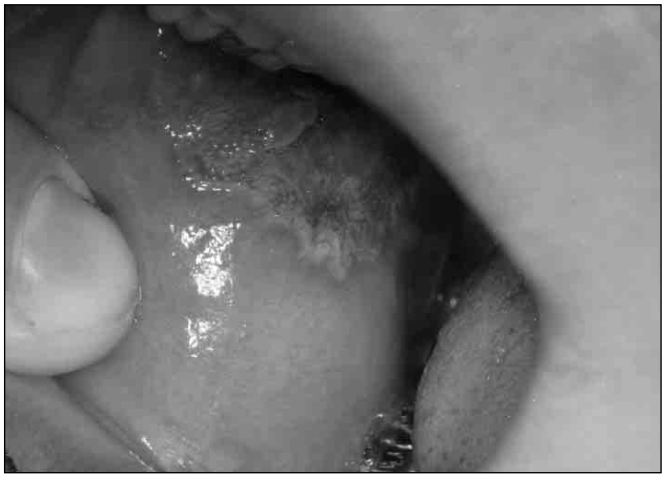

Fig. 2 - *Typical whitish, reticulated pattern of oral lichen planus lesions on the buccal mucosa.*

tion and melanophages consistent with late-phase lichenoid dermatitis. There was intermediate immunohistochemical staining with IgG, consistent with LP. The samples from the oral mucosa and lower lip showed similar histopathological and immunohistochemical features. The epithelium showed lichenoid hyperplasia and band-like lymphocytic infiltration. In the sections from the nasolabial sulcus, scalp and auricle, there were epidermal atrophy, follicular plugging, perivascular lymphocytic infiltration and vacuolar alteration, consistent with DLE. There was strong immunostaining with IgG in the basement membrane zone, also consistent with DLE (band-test).

Routine complete blood count, urinalysis, blood chemistry, immunoglobulin G, A, M, C3 and C4 levels were within normal limits. Erythrocyte sedimentation rate was 30 mm/1st h. Antinuclear antibodies was positive and anti-dsDNA antibody level was high (22.6 mIL; normal limits: 0-7 mIL). Systemic examination was normal and the patient did not meet the American Rheumatism Association criteria for systemic lupus erythematosus.

Based on the clinical, histologic and immunofluorescent findings, LP-LE overlap syndrome was diagnosed. We prescribed hydroxychloroquine 200 mg orally twice daily. The eyelid, mucosal and skin lesions improved dramatically within two weeks. The therapy was gradually tapered over six months and the patient is still free of symptoms.

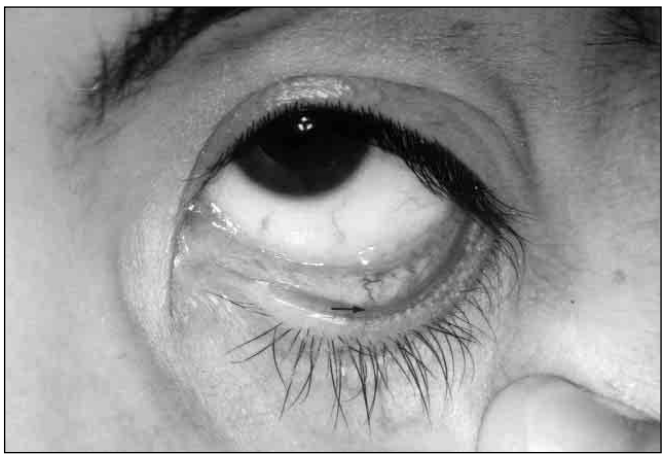

Fig. 3 - *Erythematous left lower eyelid lesion.*

DISCUSSION

LP and LE have been infrequently reported in the same patient, in what has been called the LP-LE overlap group (8). Ocular involvement has rarely been reported for both diseases. Chronic blepharoconjunctivitis, cicatrizing conjunctivitis and eyelid lesions have been defined for isolated cases of LP and LE (3-7). So far, no case of LP-LE overlap syndrome with eyelid involvement has been reported. Chung and Driebe presented a case of severe cicatricial conjunctivitis in a patient with LP, with conjunctival immunopathologic features consistent with LE rather than with LP.

However, their patient did not have active systemic or discoid LE (3).

Lichen planus of the eyelid is considered one of the rarest manifestations of the disease (6). Most reported cases of conjunctival LP have been in patients with oral LP (9). The immunohistochemical characteristics of the conjunctiva in ocular LP have not been defined (7). In some cases of LP-LE overlap syndrome, the histologic features and direct immunofluorescent findings are most consistent with LP while in others the immunofluorescent testing favours LE (10). In our case, the direct microscopic and immunohistochemical findings of the eyelid biopsy were in accordance with LP. Vacuolar alterations in the basal layer, moderate lymphocytic inflammation and melanophages detected in the histologic examination, and intermediate immunohistochemical staining with IgG of the eyelid biopsy, also suggested that the ocular involvement was associated with LP. Conjunctival inflammation associated with lichen planus must be differentiated from conjunctival involvement of other autoimmune diseases such as cicatricial pemphigoid, bullous pemphigoid, epidermolysis bullosa acquisita and lupus erythematosus. A careful clinical and histologic examination, searching for extraocular lichen planus lesions is mandatory in making the diagnosis (7).

To our knowledge, this is the first reported case of LP-LE overlap syndrome with eyelid involvement. We conclude that patients with LP-LE overlap syndrome should be questioned about ocular complaints by dermatologist and ophthalmologists, and LP and LE should both be considered in the differential diagnosis of eyelid lesions.

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