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CASE REPORT

A case of lupus erythematosus/lichen planus overlap syndrome

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ABSTRACT

A case of lupus erythematosus/lichen planus overlap syndrome (LE/LP overlap syndrome) was reported. A 53-year-old woman developed violaceous erythema around the nostrils and the upper lips and atrophic scaly erythema on the cheeks and neck. Histopathological studies revealed that the patient had distinct discoid lupus erythematosus (DLE), LP, and a lesion with combined features of DLE and LP. Direct immunofluorescent (DIF) studies of the mixed lesion revealed both prominent immunoglobulin (Ig)G deposits in a granular pattern at the basement membrane zone (BMZ) and IgM deposits in the clusters of cytoid bodies; the former are more typical of LE and the latter more of LP. DIF features in combination were unique for LE/LP overlap syndrome. The patient was satisfactorily treated with topical tacrolimus. While reports support the effectiveness of tacrolimus in either LE or LP, this is the first case of LE/LP overlap syndrome treated with topical tacrolimus.

Key words: lupus erythematosus/lichen planus overlap syndrome.

INTRODUCTION

Lupus erythematosus (LE) and lichen planus (LP) are two common and distinct skin diseases that may infrequently coexist in the same patient or exhibit lesions that may not be readily distinguished from one another. This condition, characterized by mixed clinical and histopathological features of LE and LP, has recently been referred to as lupus erythematosus/lichen planus overlap syndrome (LE/LP overlap syndrome).[1–3] Its authenticity has been questioned in some studies, but there is an accumulating list of case reports demonstrating the unarguable coexistence of LE and LP.[4,5] Immunofluorescent studies are useful to distinguish LE from LP,[6] and they may also provide a powerful method for diagnosing LE/LP overlap syndrome. Here, we present a case that not only exhibited the typical histopathological and immunofluorescence features of both LE and LP, but also showed mixed features of the two diseases in the same lesion, which uniquely characterizes LE/LP overlap syndrome.

CASE REPORT

A 53-year-old woman presented to our outpatient clinic with a 5-year history of erythema on her face. Skin examination revealed violaceous erythema with faint whitish streaks (Wickham's striae) surrounding the nasal cavity and on the upper lip (Fig. 1a), slightly atrophic erythema with fine scales on the cheeks (Fig. 1b) and neck, and white lacework on the buccal mucosa. Indurated erythema was also present on the bilateral palms. She did not suffer from subfever, arthralgia or myalgia. No medications had been taken prior to the onset of skin lesions.

Laboratory examinations including blood count, erythrocyte sedimentation rate, liver function and routine urine tests were within normal limits. Antinuclear antibody was positive with a speckled and homogenous staining pattern at a 1:40 dilution, and antibodies to double-strand DNA were detected (29 IU/ml; normal limits <6 IU/ml). Complement C3 and C4 levels were slightly decreased (83 mg/dl, 11 mg/dl; normal limits 86–160 mg/dl, 17–45 mg/dl).

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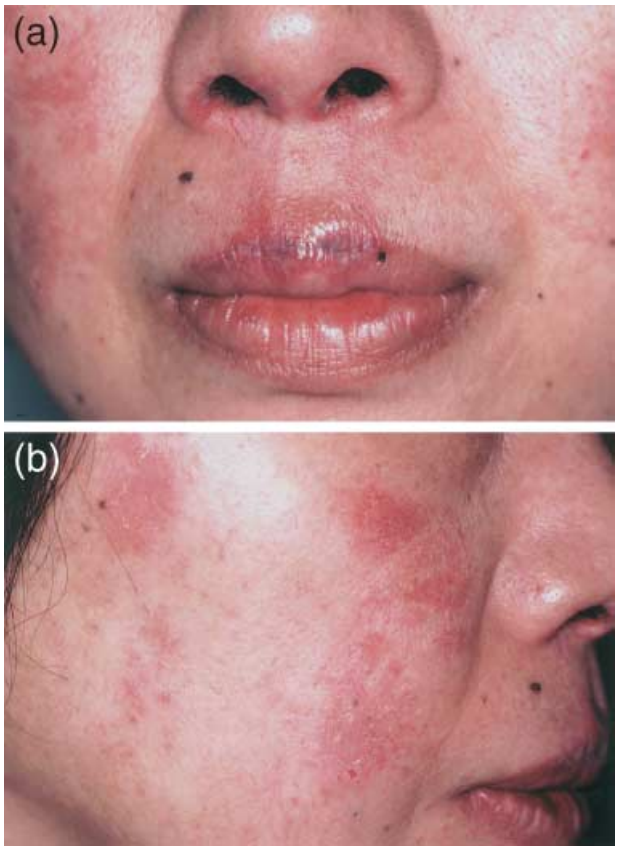

Figure 1. (a) Lichen planus (LP) lesions surrounding the nasal cavity and on the upper lip. (b) Lupus erythematosus (LE) on the cheeks presenting as slightly atrophic erythema with fine scales.

Patch testing for metals were all negative. A photosensitivity test using ultraviolet (UV) irradiation revealed a normal minimal erythema dose.

Three biopsies were taken from erythema on the cheek, neck and upper lip, respectively. Histopathology of the biopsy specimen from her right cheek showed thinning of the epidermis, liquefaction degeneration and a patchy inflammatory infiltrate consisting of mononuclear cells around blood vessels and skin appendages in the dermis consistent with discoid LE (Fig. 2a,b). A biopsy specimen from her lip showed wedge-shaped areas of hypergranulosis and a band-like mononuclear infiltrate at the superficial dermis consistent with LP (Fig. 2c,d). A biopsy specimen from the neck showed follicular plugging, a band-like lymphocytic infiltration and patchy mononuclear cell infiltrates in the dermis (Fig. 2e). Eosinophilic colloid bodies (cytoid bodies) were present at the dermoepidermal junction (Fig. 2f). Direct immunofluorescent study of the biopsy specimen from the neck showed deposits of immunoglobulin (Ig)G, IgM and C3 forming a granular pattern in a line along the basement membrane zone (BMZ) and clustering of IgG and IgM-positive (IgM > IgG) cytoid bodies at the dermoepidermal junction (Fig. 2g,h).

From the clinical, pathological, and immunological findings, the lesion on the cheeks was diagnosed as discoid LE, the upper lip as LP, and the neck as a mixture of both. Overall, the patient was diagnosed as having LE/LP overlap syndrome. Because the patient did not suffer from any symptoms of systemic involvement, she was treated topically with 0.1% tacrolimus ointment (Protopic, Fujisawa Pharmaceutical, Tokyo, Japan) with moderate but satisfactory effect.

DISCUSSION

Lupus erythematosus and LP are well established and distinct entities, but cases arise in which the two conditions coexist or cannot be readily distinguished from one another. The latter condition had been reported as an unusual variant of LE or LP,[1,2] but is recently referred to as LE/LP overlap syndrome,[3–5] now also including the former condition. In some cases of the overlap syndrome, the histological features and direct immunofluorescent (DIF) are more consistent with LP, while in other cases with LE.[3] In yet another subset of patients, the lesions of LE and LP coexist, rather than overlap, raising the controversial issue of the authenticity of LE/LP overlap syndrome.[3] Interestingly, in addition to distinct lesions of LE and LP, our case presented a lesion with combined features of LE and LP that was confirmed by both histopathological and immunofluorescence studies. It appears that our case represents a true form of "overlapped" LE and LP.

As is true in clinical features, histopathological findings usually enable us to draw the distinction between LE and LP in most cases, although they may be of some difficulty in less typical or overlapping cases. Because characteristic immunoglobulin deposits are known to be found in the skin lesions of LE and LP, DIF studies are known to be helpful in distinguishing between the two diseases.[3] In LE, immunoglobulins (IgG, IgA and IgM) and complement components deposited in a continuous granular line

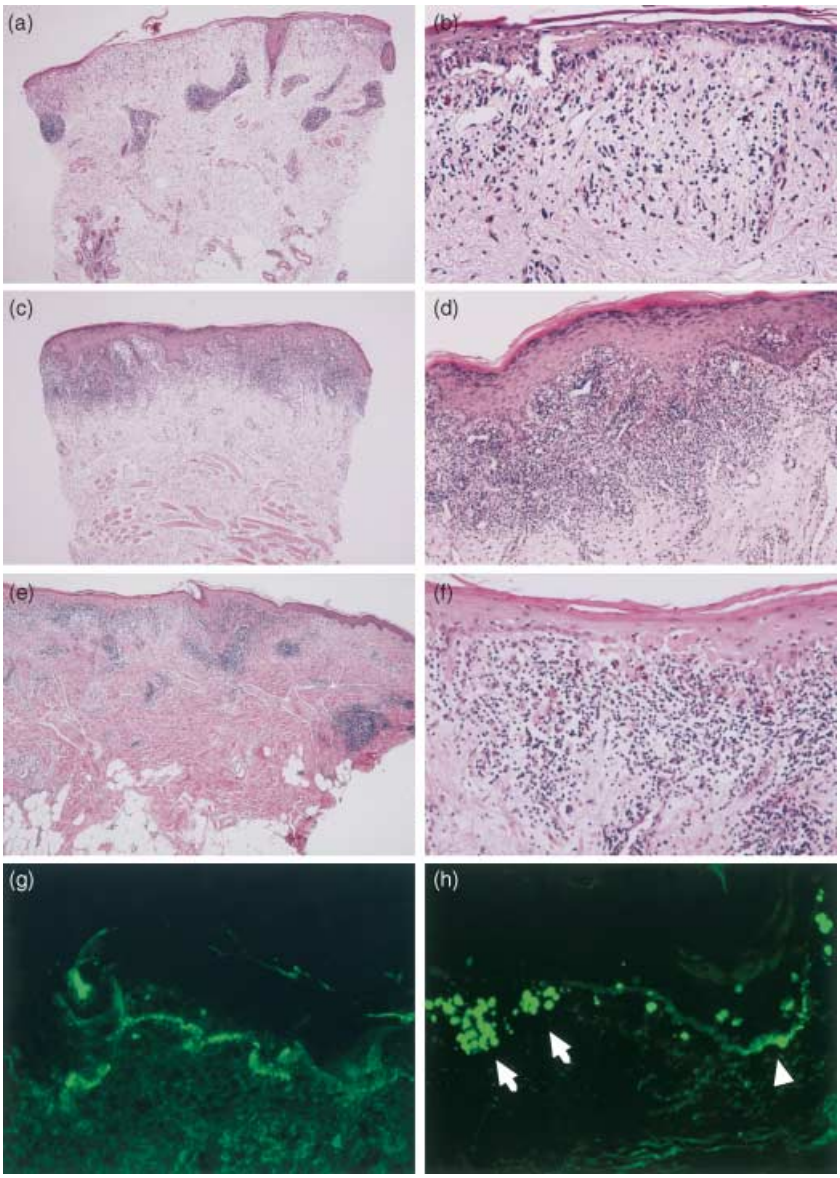

Figure 2. Biopsy from the LE lesion on the right cheek reveals atrophy of the epidermis and patchy inflammatory infiltrates in the dermis with slight liquefaction degeneration at the dermoepidermal junction (a, original magnification ×20; b, ×200). LP lesion from the upper lip shows a dense band-like lymphocytic infiltrate at the dermoepidermal junction and superficial dermis with wedge-shaped areas of hypergranulosis (c, ×20; d, ×200). The overlapped skin lesion from the neck presented follicular plugging, atrophic epidermis with many cytoid bodies at the dermoepidermal junction, and a band-like lymphocytic infiltrate as well as patchy lymphocytic infiltrates in the dermis. (e, ×20; f, ×200). Direct immunofluorescent (DIF) of (e) and (f) reveals granular deposits of immunoglobulin (Ig)G in line at the dermoepidermal junction (g, ×200) and clusters of IgM-positive cytoid bodies (arrows) with linear IgM deposition along the dermoepidermal junction (arrow head) (h, ×200).

or band along the dermoepidermal junction have been observed.[3] On the other hand, DIF in LP frequently reveals clusters of colloid bodies mainly staining with IgM and fibrin depositions in a fibrillar or band-like pattern along the dermoepidermal junction.[3] Nieboer[6] suggested that the presence of a granular band pattern of IgG at the BMZ is an important clue to differentiating DLE from LP. In our case of LE/LP overlap syndrome, DIF showed deposits of IgG, IgM, C3 and fibrin in a granular line at the BMZ, with intense IgM deposits corresponding to colloid bodies in the same lesion, exhibiting a unique character with mixed features of LE and LP. Although IgM deposits of colloid bodies may also be seen in LE, they are more prominent in LP. It is of interest that chilblain lupus, a cutaneous manifestation of LE, presents similar histological features to LP. Linear IgG and C3 deposits at the BMZ, but not IgM to colloid bodies, is seen by DIF, and its categorization as LE is justified. Not only is DIF useful to distinguish LE from LP, but our case also suggests that it may also be an alternative method for diagnosing LE/LP overlap syndrome.

Topical tacrolimus, which is a class of topical immunosuppressive agents originally developed for

atopic dermatitis,[7] is reported to be effective in various inflammatory skin disorders including LE[8] and erosive LP,[9] but its effectiveness has not been reported for LE/LP overlap syndrome. Although it did not eradicate the skin lesions, 0.1% tacrolimus ointment satisfactorily suppressed the active LE, LP and overlapping lesions in our case.

In conclusion, a case of typical LE/LP overlap syndrome is reported. DIF studies showing unique deposition pattern of immunoglobulins with a mixture of both LE and LP is of diagnostic value for LE/LP overlap syndrome. As our case implies, the contribution of systemic autoimmunity, as detected by positive antinuclear antibody and anti-dsDNA antibody cannot be denied, but the question of whether LE/LP overlap syndrome is a manifestation of LE remains yet to be addressed. Whereas treatment for LE or LP is often challenging, topical tacrolimus ointment gave acceptable results in the present case of LE/LP overlap syndrome. More cases need to be accumulated to confirm the efficacy of this drug.

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