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Oral and Skin Lichen Planus Pemphigoides.

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ABSTRACT

We report a case of a 40-year-old male patient who was referred to our department with oral and skin lesions. The skin lesions were pruritic vesicles with clear contents on the erythematous base and were present for the past 9 months. The patient was otherwise healthy and was not taking any medication. Oral lesions were multiple erosions covered with pseudomembranes. Histology finding suggested lichenoid reaction. Direct immunofluorescence showed presence of IgG along the dermo-epidermal border, and the indirect immunofluorescence was negative. Enzyme linked immunoassay was positive for BP-180. The diagnosis of lichen planus pemphigoides was established.

Keywords: lichen planus pemphigoides, oral lesions, skin lesions.

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INTRODUCTION

Lichen planus pemphigoides (LPP) is a rare, controversial disease with partially known causative factors. Traditionally, it is thought that LPP is a simultaneous presentation of two diseases, i.e. lichen planus and bullous pemphigoid. Recently, a new antigen of 200 kDA has been identified suggesting that LPP might represent a distinct entity from bullous pemphigoid (BP), i.e. that LPP is a variant of BP with the same immunopathologic mechanism [1]. LPP immune deposits might mimic BP, cicatricial pemphigoid or epidermolysis bullosaaquisita, therefore supporting the theory that LPP is a heterogeneous disease [2].

Usually, LPP manifests on the skin as pruritic erythematous areas on the body with blistering which heal without scarring [3].LPP lesions in the oral cavity have usually been described as keratotic with presence of bullous/erosive forms within the erythematous mucosa. Oral lesions might evolve on any part of the mucosa, more often on the buccal mucosa and the presence of desquamative gingivitis is seen in some cases.

LPP has been associated with malignancies such as lymphoma, colon cancer and Castleman's disease, medication intake (weight reduction drugs, ramipril, captopril, simvastatin, furosemide, Chinese herbs, cinarizine, ibuprofen and paracetamol), phototherapy (UVA/UVB) and viral infections such as hepatitis and varicella [4-11].

Histopathological finding in LPP can be confusing as in some cases it will show lichen planus or lichenoid reaction while in others pemphigoid one. Direct immunofluorescence and finding of IgG deposits with complement C3 along the dermo-epidermal junction are often sufficient for the diagnosis of LPP and indirect immunofluorescence is frequently negative. Enzyme-linked immunoassays (ELISA) to desmogleins 1 and 3 as well as on the BP-230 and BP-180 are sometimes found [12, 13].

The treatment is mainly based on the local and/or systemic use of steroids, although some recalcitrant cases have been treated with other immunosuppressive drugs such as azathioprine, dapsone, tetracycline, isotretinoin, cyclosporine, mycophenolate mofetil and intravenous immunoglobulins [14, 15].

CASE REPORT

A 40-year-old male patient was referred to our department due to the painful lesions in the oral cavity. The patient had a skin lesion on his back which had been continuously present for 9 months before we saw him. Otherwise he was healthy and was not taking any medications, however, he smoked 30 cigarettes and drank 2 litres of beer daily. He took Lysobact (lyzosime and B vitamin) for treatment of his oral lesions. Oral lesions (Figures 1-3) consisted of multiple erosions covered with pseudomembranes. Most extensive lesions were on the lips, buccal mucosa bilaterally and ventral surface of the tongue. Pruritic vesicles with clear contents on the erythematous base could be seen on his skin (Figure 4). After cracking, the skin lesions were covered with yellowish and haemorrhagic crusts whose healing left behind residual pigmentation (skin on the back and groin area). On the right sole and on the lumbosacral skin those erosions were covered with haemorrhagic crusts. The thumb nails on the feet were hyperkeratotic. Histopathological finding of the oral mucosa was highly suggestive of oral lichenoid reaction. DIF finding showed presence of IgG deposits along the epidermo-dermal border. IIF finding did not reveal circulating pemphigus or bullous pemphigoid antibodies. ELISA was negative for Dsg 1, Dsg 3 and BP-230, however, BP-180 was positive. Local therapy was initiated and included a cocktail consisting of hexetidine, D-panthenol and lidocaine 3% mouthwash to be used three times a day as well as gentamycin and bethametasone drops to be used four times a day (five drops). Systemic steroid therapy was included, i. e. 30 mg of prednisone which was tapered according to the clinical picture. We tried to exclude systemic therapy on several occasions; however, the lesions would recur. After continuous intake of systemic steroid therapy during the period of one year, the therapy stopped. The patient was examined five years after the therapy had stopped and he never had any recurrences of blistering either on the skin or oral mucosa. There were only asymptomatic residual hyperkeratotic plaques on the buccal mucosa (Figures 5-6).

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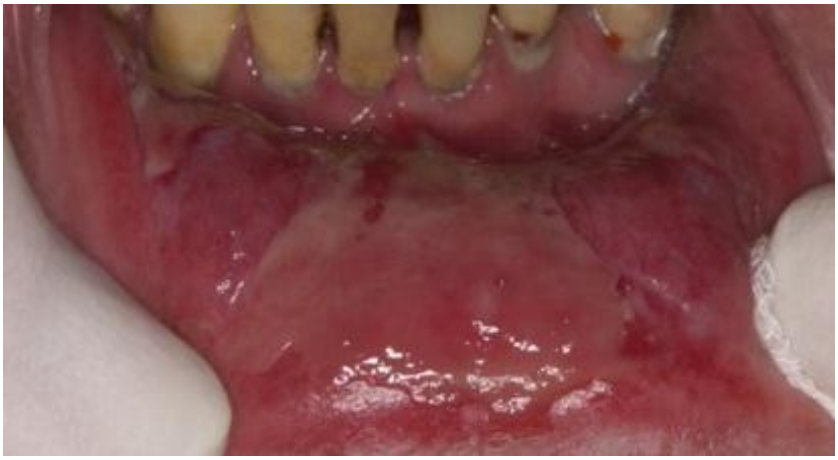

Figure 1: Bullous/erosive/ulcerative lesions covered with pseudomembranes and hyperkeratotic plaque on the labial mucosa.

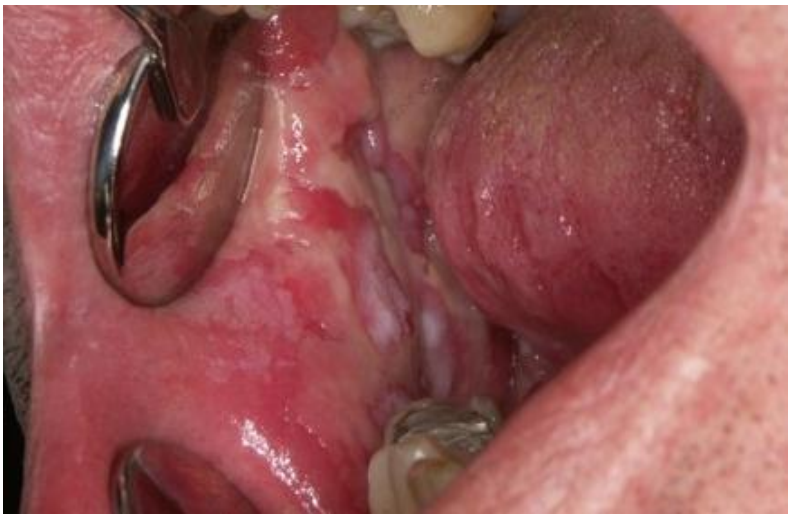

Figure 2: Bullous/erosive/ulcerative lesions covered with pseudomembranes and hyperkeratotic plaque on the inflamed right buccal mucosa.

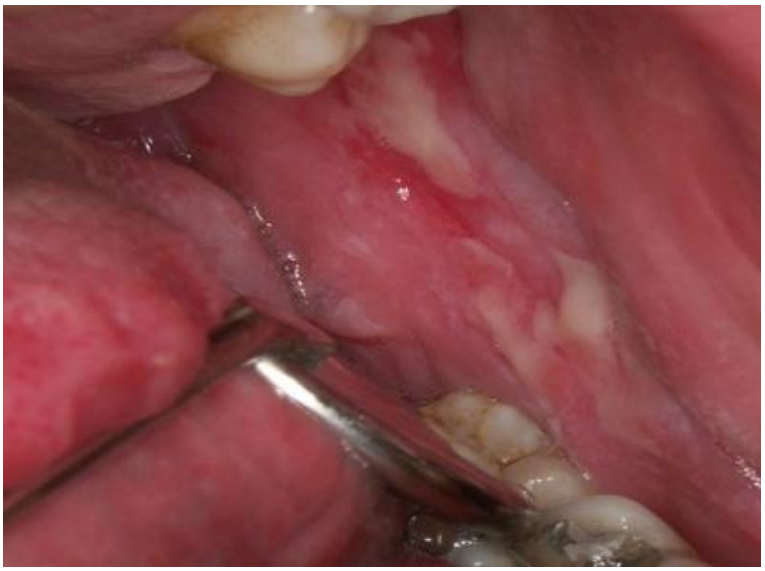

Figure 3: Bullous/erosive/ulcerative lesions ulcers covered with pseudomembranes and hyperkeratotic plaque on the inflamed left buccal mucosa.

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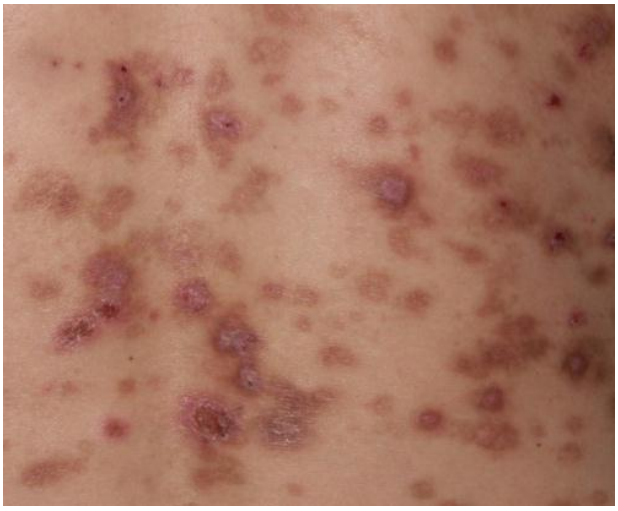

Figure 4: Cutaneous lesions of lichen planus pemphigoides.

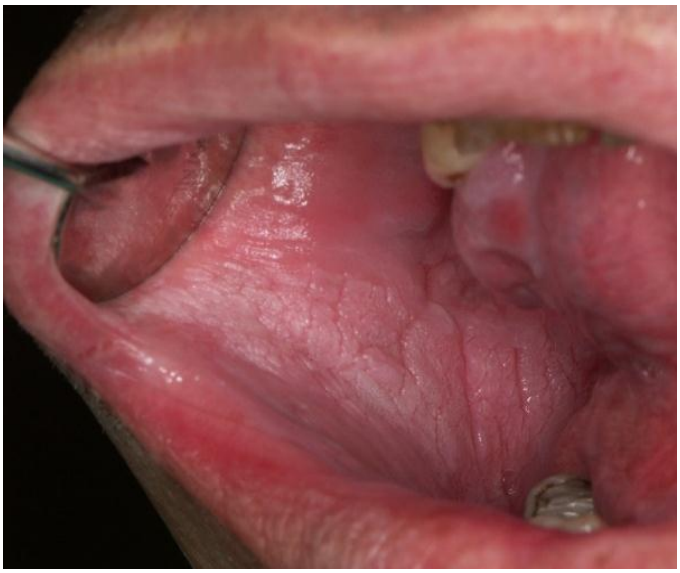

Figure 5: Residual hyperkeratotic plaque on the right cheek.

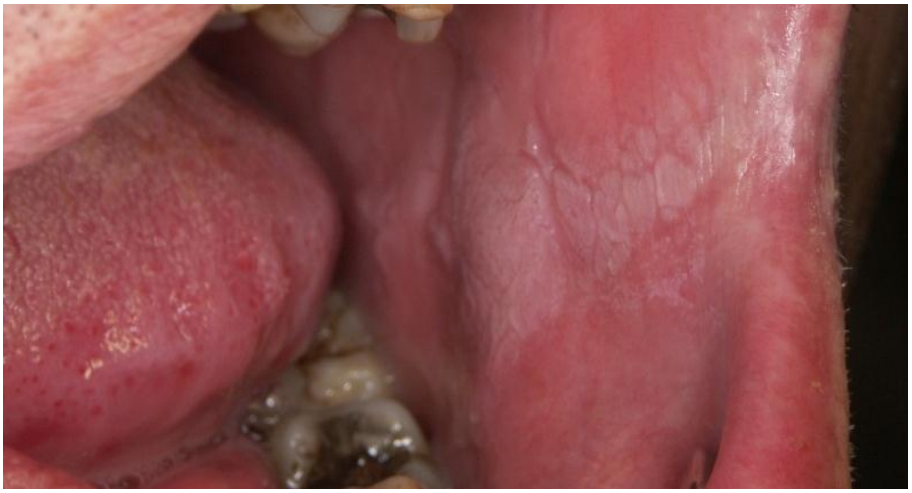

Figure 6: Residual hyperkeratotic plaque on the left cheek.

DISCUSSION

In this case, the histopathology finding indicated lichenoid reaction, however, the lesions were present bilaterally and the diagnosis of erosive or bullous lichen was more probable. Solomon et al. [13] concluded that IgG and C3 along dermo-epidermal junction are the hallmark of LPP on the direct immunofluorescence finding; however, it seems that positivity to IgG without C3 would also indicate LPP as suggested by Mignogna et al. [12], which can similarly be seen in our case report. As seen in other cases, indirect immunofluorescence was negative for circulating basement membrane zone antibodies. Circulating antibodies to BMZ components are seen only in 50-66% patients with pemphigoid [16].

ELISA showed positive finding to BP-180 which would indicate bullous or mucous membrane pemphigoid, however, bullous pemphigoid usually affects only skin whereas mucous membrane pemphigoid usually affects mucosa. It is known that pemphigoid variants cannot be distinguished clinically, histologically or by conventional immunological analysis.

LPP might be induced by certain drugs such as captopril, ramipril, simvastatin, hormone therapy, weight reduction drug, Chinese herbal drugs, paracetamol and ibuprofen [5-9] therefore it is of utmost importance to exclude, when possible, the drugs that induce bullous/lichenoid reactions in the oral cavity. Our patient was not taking any medication; therefore the drugs as a possible cause of LPP were excluded.

The differential diagnosis in patients with multiple chronic oral ulcerations might include: pemphigus variants, bullous and erosive lichen, pemphigoid, herpetiform dermatitis, epidermolysis bullosa, toxic epidermal necrolysis, linear IgA disease [17].

It is well known that LPP might have an underlying malignant disease [10] and therefore detailed medical history and systemic examination as well as tumour markers should be evaluated in certain cases. However, our patient did not have any other systemic symptoms and it was not prudent to perform tumour markers evaluation. Additionally, those are costly diagnostic procedures.

Further follow up of the patients with LPP is of utmost importance as in some patients an underlying malignancy is found. Regarding oral lesions, a regular follow up (3-6 months if asymptomatic) could be important as oral lichen planus lesions are considered premalignant.

CONCLUSION

LPP is a rare disease having clinical and histopathological features that combine lichen planus and bullous pemphigoid. Although known predisposing factors might be malignant diseases, infectious diseases, narrowband UVA/UVB and medication intake, in this case none of them could be identified. After one year of continuous steroid therapy which was tapered according to the clinical picture, the patient was free of any oral and skin lesions. However, patients should be seen at recalls since oral lichen planus is a premalignant condition, although there are no data on whether LPP itself might have premalignant potential.

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