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Oral Lichen Planus Pemphigoides: Three Cases of a Rare Entity

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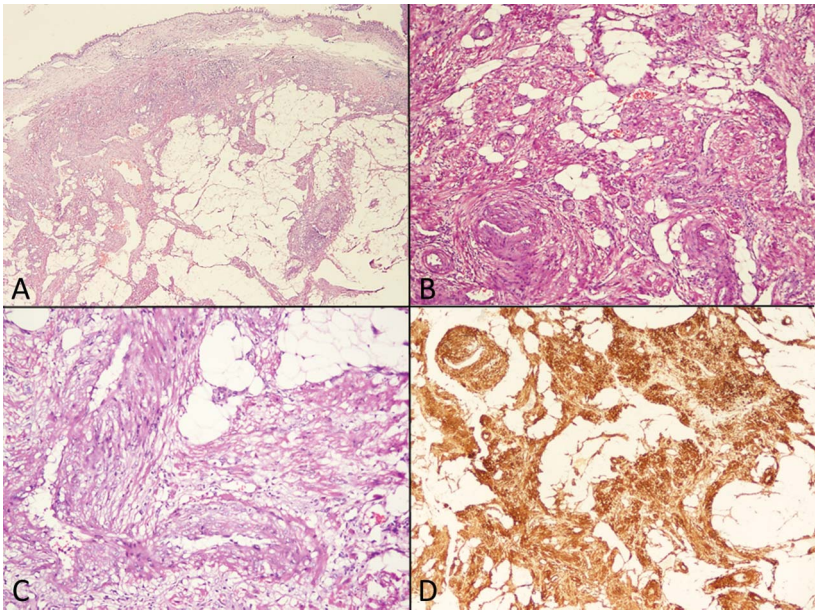

FIGURE 2. A, Biopsy shows a polypoidal mass covered with respiratory epithelium. A circumscribed tumor is identified in the subepithelium composed of fat and bundles of smooth muscle cells (hematoxylin and eosin, ×40). B, The tumor shows variable proportions of mature fat, smooth muscle cells and many thick-walled bleed vessels (hematoxylin and eosin, ×100). C, The smooth muscles cells are arranged in fascicles and shows bright eosinophilic cytoplasm (hematoxylin and eosin, ×400). D, The smooth muscle cells in the stroma and in the wall of the blood vessels show strong positivity for SMA (immunohistochemistry, ×200).

first described an unusual case of AML of the nasal cavity. Most cases have been reported from Asian countries including the Middle East,[5] Turkey,[4] Japan,[2] and Korea.[6,7] Clinically, patients usually present with nonspecific symptoms such as nasal obstruction, nasal polyp, recurrent epistaxis, and snoring, with some of them incidentally detected during CT scan of brain.[7,8] Sinonasal AML poses diagnostic difficulties because radiological and clinical examination findings are nonspecific; therefore, histopathological examination and immunohistochemistry are crucial for definitive diagnosis.[9] Pathologists should have a high index of suspicion to recognize this rare entity at unusual sites. Nasal AML is a benign tumor, and complete surgical excision is the treatment of choice. Recurrences have not been documented.[2,5]

We highlight the striking similarities between cutaneous and mucosal AML, both clinically and histologically. In fact, both together comprise a distinct disease entity and are better clubbed under the term "mucocutaneous AML."

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Oral Lichen Planus Pemphigoides: Three Cases of a Rare Entity

To the Editor:

Lichen planus pemphigoides (LPP) is a rare autoimmune dermatosis characterized by clinical and histopathologic features of LP and bullous pemphigoid (BP) or mucous membrane pemphigoid.[1–3] Herein are 3 cases of oral LPP followed by a brief discussion.

CASE 1

A 75-year-old Caucasian woman presented with symptomatic gingival lesions of 7 months duration. Generalized erythema, edema, and desquamation were observed on the maxillary and mandibular gingiva (Fig. 1). Histopathologic analysis revealed lichenoid inflammation on routine staining (Figs. 2 and 3) and linear C3 deposition at the epithelial–connective

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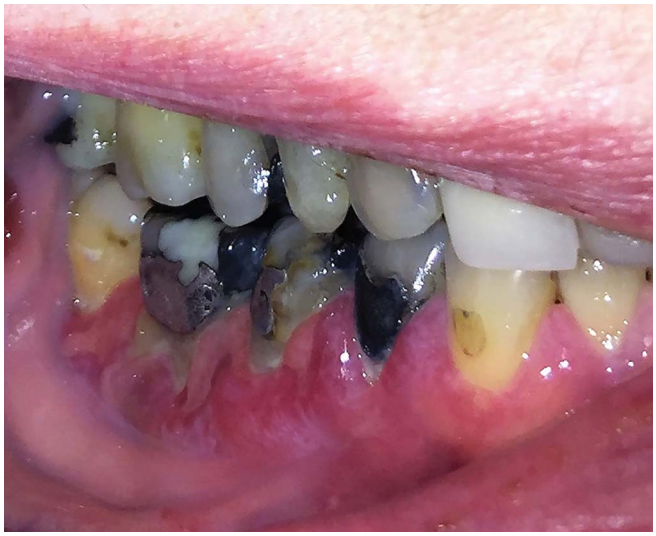

FIGURE 1. Oral LPP. This photo demonstrates generalized erythema, edema, and desquamation associated with oral LPP affecting the right posterior mandibular gingiva.

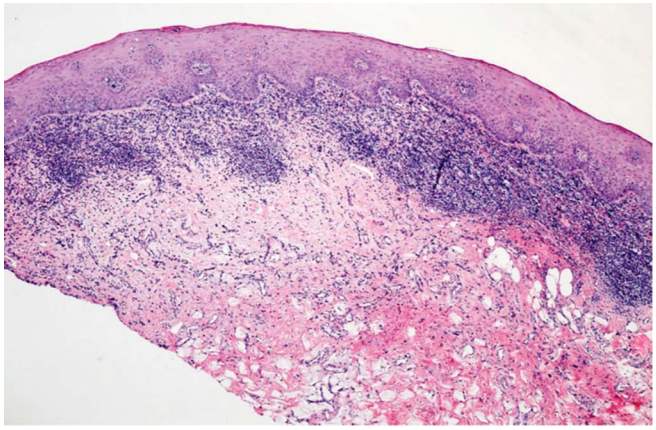

FIGURE 2. Hematoxylin and eosin, ×2. Oral LPP. This photo demonstrates lichenoid inflammation of the affected tissue. Note the parakeratotic epithelium exhibiting sawtooth rete ridge formation and degeneration of the basal cell layer and a dense band-like infiltrate of lymphocytes.

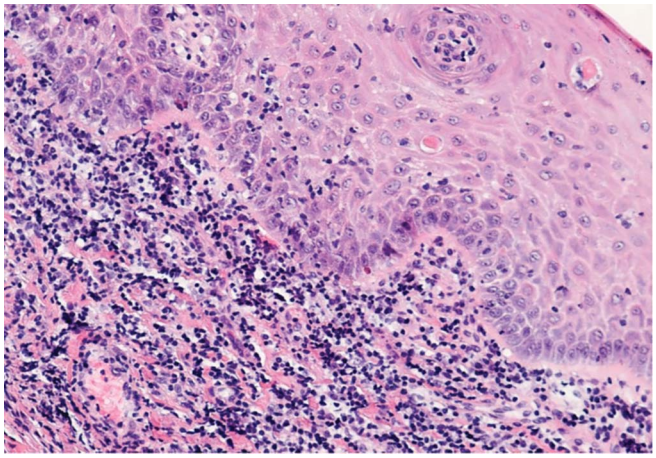

FIGURE 3. Hematoxylin and eosin, ×10. Oral LPP. This photo demonstrates a lichenoid tissue reaction of the affected gingival tissue.

tissue interface with direct immunofluorescence (DIF) (Fig. 4). Clinical and histopathologic findings were consistent with oral LPP.

CASE 2

A 71-year-old Caucasian man presented with a 1-year history of symptomatic gingival and concurrent cutaneous lesions. Desquamation and severe erythema were observed on the mandibular and maxillary gingiva with positive Nikolsky sign. Histopathologic analysis demonstrated a lichenoid interface reaction on routine staining with linear IgG, IgA, C3, and fibrinogen deposition at the mucosal–submucosal interface with DIF. Clinical and histopathologic findings were consistent with oral LPP.

CASE 3

A 67-year-old Caucasian woman presented with a 2-year history of symptomatic oral lesions, dysphagia, unintentional weight loss, and vaginal lesions. Examination revealed extensive erythema and areas of erosion on the buccal mucosa, gingiva, and tongue. Histopathologic analysis revealed lichenoid inflammation on routine staining and linear IgG, C3, and shaggy fibrinogen with DIF. Enzyme-linked immunosorbent assay (ELISA) was positive for BP180 and BP230 antibodies. These findings supported the diagnosis of oral LPP.

Management of all patients included dexamethasone 0.5 mg/5 mL solution (U.S.FDA off-label use), 5 mL swish and spit twice daily, and nystatin 100,000 units/mL, 5 mL swish and spit 3 times daily. Two patients (cases 1 and 2) were also prescribed topical clobetasol gel 0.05% (U.S.FDA off-label use) twice daily. In addition, the patient in case 2 was prescribed doxycycline 50 mg (U.S.FDA off-label use) daily. All patients received substantial benefit from therapy.

DISCUSSION

LPP was first introduced by Kaposi in 1982 and thought to be a variant of BP or LP.[1,2] Growing evidence suggests it is a distinct entity characterized by lichenoid and bullous lesions that develop in the context of autoantibodies targeting type XVII

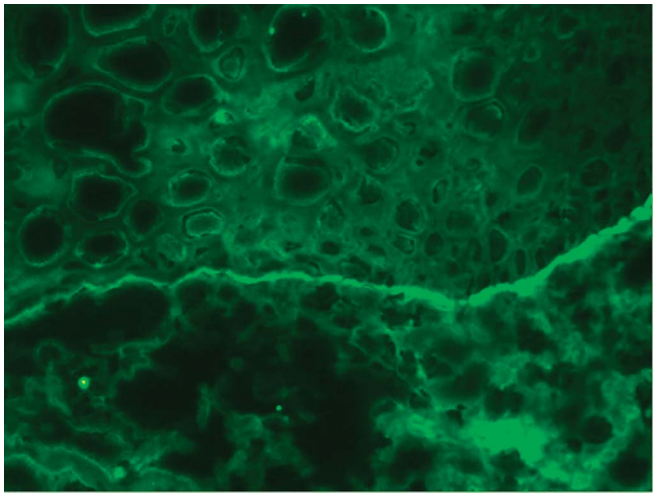

FIGURE 4. Direct immunofluorescence. Oral LPP. This photo demonstrates linear C3 deposition at the epithelial–connective tissue interface via immunofluorescence.

collagen COL17.[1,3] Prevalence of LPP is estimated at 1 per 1,000,000 patients. LPP has no gender predilection and commonly affects adults in the fourth or fifth decades, but has also been described in childhood.[1,4] Etiology is primarily idiopathic, but may be associated with infection-related and drug-induced causes.[4] Diagnosis of LPP may be challenging and made by characteristic clinical, histopathological, and immunopathological features.[1–4] Clinical features include blisters and erosions that appear on pre-existing lichenoid lesions or uninvolved skin or mucosa, frequently affecting upper and lower extremities; however, it may present in other body areas and limited to the oral cavity.[1–4] Oral LPP frequently affects the gingiva and buccal mucosa, presenting as desquamative gingivitis, Wickham striae, ulcerations and/or bullae. Histopathologically, LPP demonstrates findings of lichenoid tissue reactions with subepithelial bullae on routine histology and linear deposits of IgG, IgA, and C3 along the basement membrane zone with DIF of perilesional skin or mucosa.[1–4] Treatment of LPP comprises topical or systemic steroids and steroid-sparing agents.[1,2] Oral LPP management includes minimizing trauma from sharp teeth and food, topical corticosteroids (U.S.FDA off-label use) with or without occlusive dental trays and concomitant topical antifungal prophylaxis for oral candidiasis.[2] Prognosis of LPP tends to be better compared with BP, mucous membrane pemphigoid, and LP with an estimated recurrence rate of 20%.[3,4]

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