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VENEREOLOGY – NOT TO BE FORGOTTEN
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QUIZ SECTION

Ulcerated Nodules on the Oral Mucosa and Fingers: A Quiz

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An 85-year-old man presented with a 1-cm ulcerated nodule with a slightly elevated border on his tongue (Fig. 1a) and oral vestibule. He also had 0.5-cm ulcerated papules on 3 of his fingers (Fig. 1b). The patient reported that the lesions were asymptomatic and had developed slowly over 18 months prior to his presentation; first the lesion in the oral cavity and then the lesions on the fingers. A complete skin examination revealed no other lesions. The lymph nodes were normal. He had a medical history of diabetes mellitus, hypertension and hyperlipidaemia, controlled with amlodipine, irbesartan, hydrochlorothiazide, metformin, simvastatin and torasemide. Multiple punch biopsies were taken from the tongue and fingers (Fig. 1c).

What is your diagnosis? See next page for answer.

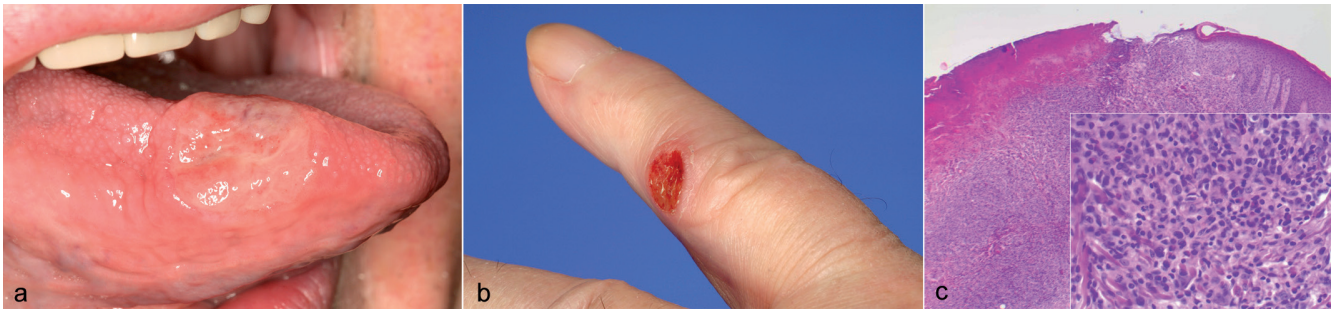

Fig. 1. (a) Clinical examination revealed a 1-cm ulcerated asymptomatic nodule with slightly elevated border on the tongue. (b) 0.5-cm ulcerated papules on the finger. (c) Histological examination showed a diffuse mixed nodular infiltrate formed by medium-to-large-sized anaplastic lymphocytes admixed with small lymphocytes, histiocytes and large numbers of eosinophils (H&E staining, × 10, insert × 40). doi: 10.2340/00015555-1488

ANSWERS TO QUIZ

Ulcerated Nodules on the Oral Mucosa and Fingers: Comment

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Diagnosis: Lymphomatoid Papulosis, Type A

Fig. 1c shows a dense nodular lymphoid infiltrate with central ulceration. The infiltrate was composed mainly of medium-to-large-sized, anaplastic lymphocytes admixed with small lymphocytes, histiocytes and large numbers of eosinophils. By immunohistochemistry the tumour cells expressed CD3 and CD4, and 30% of them expressed CD30. Analysis of the T-cell receptor (TCR) γ-chain genes identified a monoclonal rearrangement, with an identical clone in the skin and tongue. Total computed tomography (CT) scan and laboratory studies revealed no abnormalities. These findings are consistent with a diagnosis of lymphomatoid papulosis, type A with mucosal involvement. The lesions resolved spontaneously within 6 weeks of initial presentation.

Regarding the World Health Organization – European Organisation for Research and Treatment of Cancer (WHO-EORTC) classification of primary cutaneous lymphomas, lymphomatoid papulosis (LyP) and primary cutaneous anaplastic large cell lymphoma (C-ALCL) are classified as primary cutaneous CD30-positive lymphoproliferative disorders (1). LyP was first described by Macaulay in 1968 (2) and is characterized by chronic, recurrent, self-healing papulonecrotic and nodular lesions.

The lesions are typically localized on the trunk and limbs, while oral or mucosal involvement is extremely rare. The first case of oral mucosal involvement in LyP was described in 1997 (3). To our knowledge, 7 previous definite cases of mucosal involvement in LyP have been reported to date (4–8). Oral lesions also appear as solitary or multiple papules or indurated nodules with a tendency to ulcerate. In all patients the ulcers regressed spontaneously within 2–4 weeks. Usually they develop in patients with previous skin lesions of LyP. In only 1 case were the oral lesions the first sign of LyP (8). No differences in the clinical course of the patients were noted. The differential diagnosis of oral LyP includes squamous cell carcinoma, granulomatous diseases, lupus erythematodes, Behçet's disease, infectious disorders, especially viral infections such as herpetic infections, drug eruption, angiolymphoid hyperplasia with eosinophilia and Riga–Fede disease.

In mycosis fungoides oral involvement is also rare, but in contrast to LyP it is considered to be a marker for poor prognosis and a manifestation of advanced disease (9). However, mucosal involvement is a common feature in primary cutaneous CD8-positive epidermotropic cytotoxic T-cell lymphoma or nasal-type extranodal natural killer/T-cell lymphoma.

In conclusion, oral involvement in LyP is rare, but it is important to recognize this uncommon clinical presentation in order to avoid misdiagnosis and overtreatment.

The authors declare no conflicts of interest.

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