SHORT COMMUNICATION

Darier disease: a case report

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Introduction

Darier disease, also known as keratosis follicularis, dyskeratosis follicularis, and benign dyskeratosis, is a rare disorder of keratinisation that primarily affects the skin and, to a lesser extent, the oral mucosa. It was described independently by both Darier and White in 1889. It has a prevalence of 1:100,000 of the population and is inherited as an autosomal dominant trait.1

The age of onset is childhood or adolescence. Patients usually present with multiple small firm reddish-brown papules on the forehead, scalp, neck, shoulders, chest, and limbs. Other cutaneous signs include punctate keratotic pits of the palms and soles and dystrophy of the nails, which is characterised by a red and white sandwich of streaks associated with a V-shaped notch. The oral lesions are usually asymptomatic and are seen as multiple, normal-coloured or white flat-topped papules that predominantly affect the palate.

Case report

A 42-year-old white man was referred by his dentist with leukoplakia of the palate. He had a diffuse white leukoplakia-like change of the palatal mucosa. Similar white changes were noticed at multiple sites on the mandibular gingiva (Fig. 1). The patient was otherwise healthy. He smoked a few cigarettes a day, and drank less than two units of alcohol a day.

Under local anaesthesia a biopsy specimen was taken from the palatal lesion. Histological examination showed the presence of keratosis follicularis, also referred to as Darier disease.

The patient was interviewed again and examined. Although he initially denied the presence of