Darier disease: A case report

Article type: Article.

Abstract:

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.

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