Our Dermatology Online

Case Letter

Papillon–Lefèvre syndrome

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Sir,

Papillon–Lefèvre syndrome (PLS) is an uncommon hereditary disorder characterized by erythemato-squamous palmoplantar keratoderma present in childhood coupled with severe periodontitis leading to a heightened risk of premature tooth loss [1]. Herein, we present the case of a thirteen-year-old child from a consanguineous marriage with erythemato-squamous lesions manifested since the age of two years, progressing to periodontitis one year previously, causing mobile teeth and premature loss of temporary teeth. A clinical examination revealed well-defined palmo-plantar hyperkeratosis with a red band (Figs. 1a and 1b), while an endobuccal examination showed severe and widespread gingival inflammation, recessions, and tooth mobilities (Fig. 2), indicative of severe periodontal destruction.

A biopsy was performed to eliminate other differential diagnoses and returned without any particularities. Radiographic examination confirmed marked alveolar bone lysis around the remaining teeth. On the basis of the clinical and radiological findings, the diagnosis of LPS was retained, and the patient underwent retinoid treatment and was subsequently referred to ortho-dental surgeons for a removable prosthesis.

Papillon–Lefèvre syndrome is a rare genotypic condition with an incidence of 1 to 4 cases per million [2]. LPS manifests with consistent dermatological and dental signs, including erythematous hyperkeratosis of the palms and soles appearing between the first and fourth year of life in relapses and remissions. Buccal manifestations involve gingival inflammation and alveolar bone lysis, leading to temporary teeth exfoliation around the age of 4 to 5 years, repeating with permanent teeth eruption. Genetically, the transmission is autosomal recessive, often associated with parental consanguinity. The gene responsible for LPS is located at 11q14/q21, encoding cathepsin C. SPL results from a mutation in this gene, suppressing its activity and affecting epithelial structural organization and immunological properties supporting the teeth [3]. Our case's clinical signs were so indicative that genetic testing was not pursued.

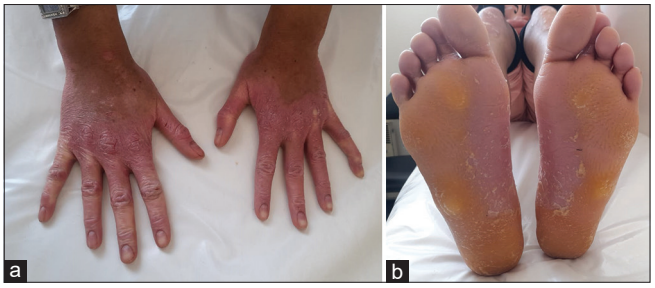

Figure 1: (a and b) Palmo-plantar hyperkeratosis (note the well-limited red band).

Figure 2: Loss of the teeth and severe generalized gingival inflammation.

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The treatment for skin lesions involves oral retinoids to reduce keratoderma and slow alveolysis.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

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