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## Papillon-Lefevre Syndrome: A case report



### Síndrome de Papillon-Lefevre: descripción de un caso

Dear Editor:

Papillon-Lefevre Syndrome (PLS) is a rare autosomal recessive disorder characterized by rapidly progressive periodontal tissue destruction, early loss of primary and permanent teeth and widespread hyperkeratotic areas of palms and soles.<sup>1–3</sup> Cathepsin C gene mutation is responsible for its etiology.<sup>4</sup> Palmoplantar hyperkeratosis typically starts between 1 and 4 years of age.<sup>5</sup>

A 6-year-old boy with a PLS diagnosis was referred to our clinic from the dermatology department. He did not have any other systemic disease, and his parents had no consanguineous marriage. The patient underwent extraoral, intraoral and radiographic examination.

Extraoral examination showed dryness and hyperkeratosis in bilateral palms, knees and foot dorsum (Fig. 1a–c). Hair and nails had normal development. Our patient had normal development according to age; there was no physical and mental retardation.

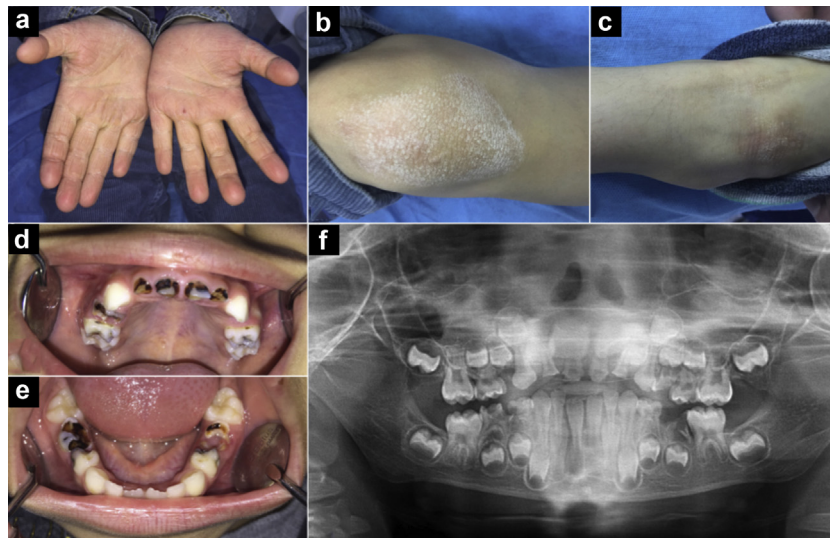
Intraoral examination showed deep dentin caries in the upper primary incisors, molars (55, 52, 51, 61, 62, 64 and 65), lower primary molars (74, 75, 84 and 85) and root of primary tooth number 54. It was also noted that the patient's oral hygiene was not good (Fig. 1d–e).

Panoramic and periapical radiographs from our patient supported our intraoral findings. They detected destruction in the alveolar bone at the root apical of teeth numbers 51, 75 and 84 and fracture of tooth number 75 (Fig. 1f).

Examination of the biopsy specimen taken from the patient's knee revealed microscopic findings compatible with PLS. In addition, normal brain computerized tomography findings were observed in the result of brain computerized tomography taken from the patient. Cathepsin C gene analysis was not requested from the patient again because the patient was diagnosed from the dermatology clinic.

Based on patient's history, clinical, radiographic and biopsy findings, a PLS diagnosis was supported and treatment planning was performed. It was decided to extract primary teeth numbers 51, 64, 75 and 84 and the root of primary tooth number 54; to fill primary teeth numbers 61, 62 and 74; and to apply root canal treatment to primary teeth numbers 55, 52, 65 and 85. Oral hygiene training of the patient and follow-up of the patient was requested.

PLS was originally described in 1924 by two French physicians, Papillon and Lefèvre. PLS is inherited as an autosomal recessive trait and has a reported prevalence of 1–4 cases per million. PLS usually appears in childhood. Males and females are equally affected, and patients are normal at birth. The disorder is characterized by diffuse palmoplantar keratoderma and rapidly progressing periodontitis leading to premature loss of both deciduous and permanent teeth. Skin lesions develop concurrently with oral lesions



**Fig. 1.** Hyperkeratosis of palms (a). Hyperkeratosis of knee (b). Hyperkeratosis of foot dorsum (c). Intraoral view—maxilla (d). Intraoral view—mandibular (e). Panoramic radiograph (f).

and may extend to dorsal surfaces of the hands and feet. Another form of disease associated with palmoplantar keratosis and severe aggressive periodontitis is Haim-Munk Syndrome. It differs from PLS with symptoms such as arachnodactyly, acroosteolysis and onychogryphosis.<sup>1</sup>

The cause of the development of these lesions is attributed to three main factors, namely genetic, immunologic and microbiologic. The genetic factor responsible for causing this disease is due to mutations of the cathepsin C gene (CTSC) in the region of chromosome 11q14–21. The immunological factor responsible for PLS mainly depends on the deterioration of neutrophil chemotaxis, phagocytosis, bactericidal capabilities, decreased cell migration, lymphocytic response and monocytic activity. Impairment in natural killer cells' cytotoxicity has been implicated in the development of PLS. A possible bacterial etiology has also been proposed. It is believed that *Actinobacillus actinomycetemcomitans*, *Porphyromonas gingivalis*, *Fusobacterium nucleatum* and *Prevotella intermedia* may be amongst the organisms involved not only in periodontal breakdown but also in the cutaneous lesions of PLS.<sup>2</sup>

In PLS, there is severe periodontal destruction and early loss of primary and permanent teeth. The permanent dentition starts to erupt at the proper time, but periodontal destruction begins to occur at around 8–9 years of age. All permanent teeth are usually lost before 14–16 years of age.

The management of cases with PLS requires a multidisciplinary approach with the active participation of the dental surgeon, dermatologist and pediatrician. Treatment of the dental component of the disorder is aimed at eliminating the reservoir of causative orga-

nisms. Oral retinoids such as acitretin and isotretinoin have proven to be beneficial in treating both the dental and cutaneous lesions of PLS.

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