

# Papillon-Lefevre Syndrome

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Inflammation that is associated with periodontium and its related structures can be managed in most of the patients but unfortunately, not all forms of periodontal disease respond to treatment. Periodontitis that is resistant to conventional periodontal therapy are recognized in certain medical conditions.<sup>1</sup> Certainly one of the most challenging condition is Papillon-Lefèvre Syndrome.

Papillon-Lefèvre Syndrome is a rare inherited autosomal recessive disorder which was first described by two french physicians, Papillon & Lefèvre in 1924.<sup>5</sup> It usually causes severe destructive periodontal damage affecting both deciduous and the secondary dentition which can result in early loss of teeth. It affects the skin which is characterized by redness, hyperkeratosis of palm and sole and is caused by mutation in cathepsin-C gene.<sup>1</sup> Hence it is a genetic defect that is located on chromosome 11q14.1-q14.3, which involves mutation of gene CTSC.<sup>3</sup> The syndrome has an estimated prevalence of 1 to 4 cases per million. Incidence is seen in patient's with consanguineous marriages.<sup>4</sup> Such patients suffer from severe periodontal destruction as the primary teeth are lost as early as 4 years and most permanent teeth by age of 14 years.<sup>3-4</sup> The etio-pathogenesis of PLS is not completely understood however anatomic, microbial and viral agents as well as host response are suspected causative factors. Male and female are equally affected with no racial predominance.<sup>5</sup> Here we report a case with clinical findings of such complex syndrome.

A 19-year-old male patient presented to the Prosthodontics department with the chief complaint of difficulty in chewing and bad breath. Past dental history revealed that the patient had early loss of deciduous teeth by the age of 5-6 years. He got his mandibular central incisors extracted a couple of years ago because he had difficulty in chewing with these teeth. Clinical examination revealed marked hyperkeratotic patches on dorsum of aspect

hands (Fig 1). The nails of the hands were keratotic but were not malformed. His feet were not examined as the patient did not give consent to removal of his shoes; but informed that they were similar in appearance to his hands. On intra-oral examination, there was generalized inflammation of the mucosa and grade III mobility in almost all the teeth (Fig 2). The Orthopantomogram (OPG) revealed advanced bone loss and a typical Papillon-Lefèvre Syndrome finding of floating teeth (Fig 3). No ultimate treatment exists for the prevention or management of periodontitis associated

**Fig. 1:** Hyperkeratotic patches on hands



**Fig. 2:** Intraoral view



with PLS.<sup>3</sup> After discussing the various management options, patient opted for provision of conventional complete dentures after extraction of all the teeth. Owing to young age and better motor skills, such patients usually adapt easily and quickly to the prosthesis.<sup>4</sup> More recently, implant-supported removable or fixed prosthesis, with and without bone augmentation, have been used as definitive treatment modality to rehabilitate such patients. However, only a few cases have been reported in literature and long-term clinical outcomes need to be established.<sup>5,6</sup>

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**Fig. 3:** Advanced bone loss  
(Teeth floating in air)



Periodontitis is the more serious component of this disease. The deciduous teeth erupt at the usual time but then the onset of disease is heralded by gingivitis and a florid Periodontitis.<sup>2,4-8</sup> There is no definitive treatment available for preventing Periodontitis although oral-hygiene instructions, root planning and scaling along with antibiotics can improve condition. In later stages, Patients with this disease may loose all of their teeth. Hence managing such a patient should involve a multidisciplinary team approach that involves a Periodontist, Prosthodontist, Dermatologist and Psychologist for treatment.<sup>6,9</sup>

#### CONFLICT OF INTEREST

None declared

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