

# Papillon-Lefevre Syndrome: Challenge for Periodontal Management

Dr. Krishna Prasad Lamichhane,<sup>1</sup> Dr. Shaili Pradhan,<sup>1</sup> Dr. Ranjita Shrestha Gorkhali,<sup>1</sup>  
Dr. Pramod Kumar Koirala<sup>1</sup>

<sup>1</sup>Periodontology and Oral Implantology Unit, Department of Dental Surgery,  
National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.

## ABSTRACT

Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive disorder associated with rapidly progressing periodontitis leading to premature loss of deciduous and permanent dentition and diffuse palmoplantar keratosis. Immunologic alterations, genetic mutations, and role of bacteria are some aetiologic factors. Patients present with early periodontal destruction, so periodontists play a significant role in diagnosis and management. This paper reports a case of Papillon-Lefevre syndrome with its clinical manifestations and challenges for periodontal management which was diagnosed in dental department.

**Keywords:** Diagnosis; genetic mutation; management; manifestations; palmoplantar keratosis; periodontitis.

## INTRODUCTION

Papillon-Lefèvre syndrome (PLS) is an autosomal recessive disorder of keratinisation, and severe destructive periodontal disease caused by mutations in cathepsin C gene,<sup>1</sup> expressed highly in epithelial regions like keratinised oral gingiva, palms, soles, and knees. An impaired chemotactic and phagocytosis of polymorphonuclear leukocytes and impaired reactivity to T and B cell mitogens.<sup>2</sup> Incidence is one to four per million with no sex and racial predominance.<sup>3</sup> Disease becomes apparent by two to three years of age leading to premature exfoliation of deciduous teeth. As permanent dentition erupts, same sequence of events recur, leading to early shedding.<sup>4</sup>

## CASE REPORT

Twenty-five years old male patient reported to the Department of Dental Surgery with the chief complaint of missing many teeth in both upper and lower jaws and a loose tooth in lower front jaw for five years. After taking consent, extraoral examination revealed palmar (Figure 1) and plantar (Figure 2) hyperkeratosis and hyperkeratosis on knees, elbows and on dorsum of hands and feet. Patient

gave a history of liver abscess and drainage six years back and also frequent skin infections. Patient was otherwise physically and mentally sound and reported no family history of similar condition. Intraoral examination revealed missing all teeth in both maxilla and mandible except all second molars and presence of grade III mobile canine on third quadrant and first premolar on both third and fourth quadrants (Figure 3a). Maxilla showed flat and atrophic alveolar ridge and mandible showed thin and atrophic ridge at edentulous sites (Figure 3 b and 3c).

Orthopantomogram revealed severe alveolar bone loss at edentulous sites with the floor of maxillary sinus close to alveolar ridge in maxillary posterior region and less alveolar bone height in mandibular posterior regions but well supported alveolar bone at all second molars (Figure 4). Patient also gave the history of early loss of deciduous dentition at the age of four to five years and permanent teeth were started to loss at the age of 14-15 years.

Patient was sent for dermatologic consultation and dermatologist did incisional skin biopsy on dorsum of right hand and on histopathologic examination revealed presence of hyperkeratosis on superficial layer of epithelium and thickening of all layers of epithelium and presence of

### Correspondence:

Dr. Krishna Prasad Lamichhane  
Periodontology and Oral Implantology Unit, Department of Dental Surgery, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.  
email: lmchkp@gmail.com

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Figure 1: Palmar hyperkeratosis. Figure 2: Plantar hyperkeratosis

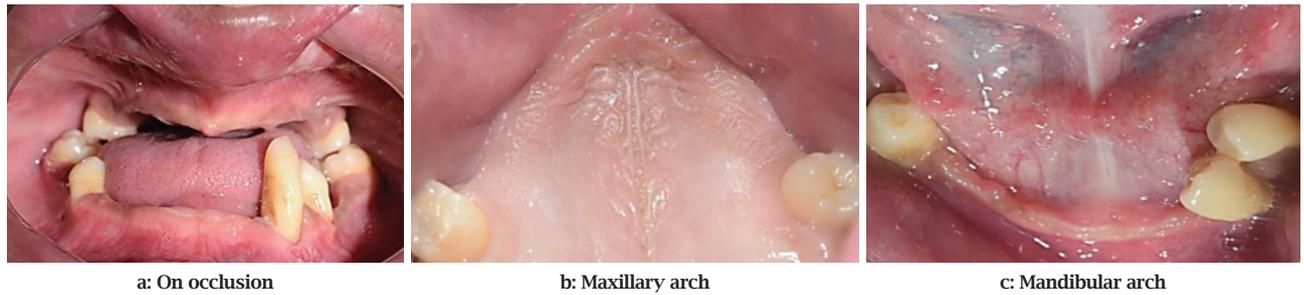


Figure 3: Intraoral findings clinically.



Figure 4: Orthopantomogram.

Figure 5: Histologic picture.

chronic inflammatory infiltration in connective tissue (Figure 5).

Based on clinical findings, radiographic and histologic reports, we came across the diagnosis of Papillon-Lefevre syndrome. We planned cast partial denture on maxilla and implant supported overdenture on mandible after extraction of 33, 34 and 44 in mandible. We did cone beam computed tomography of mandible but revealed extension of mandibular canal anteriorly up to canine region and extension of lingual vessels at midline region of mandible and very thin alveolar ridge in between mental foramina which contraindicated the possibility of implant placement. So we sent the patient for possible prosthodontic rehabilitation.

## DISCUSSION

Papillon-Lefevre syndrome is challenging for periodontal management. Treatment modalities include combined mechanical and antibiotic periodontal treatment, extraction of severely diseased teeth, oral hygiene instruction, intensive maintenance therapy and Microbiological monitoring and treatment of infection with *A. actinomycetemcomitans*. Yacoub AA and Hattab FN in 2008 treated the case of Papillon-Lefevre syndrome using amoxicillin and metronidazole three times a day for seven days every two to three months combined with mechanical and chemical periodontal therapy. Only short-term improvement was noticed but recurrence of disease was reported soon after the completion of therapy.<sup>5</sup>

In a study, Kinaia et al. in 2017 did a complete rehabilitation of maxilla and mandible using a cranium graft for a dental implant supported prosthesis. One year follow up showed well osseointegrated implants with good function with esthetic smile.<sup>6</sup>

Nickles et al. in 2013 did a retrospective evaluation of periodontal status in 8 patients with Papillon-Lefevre syndrome observed for  $\geq 10$  years. After comprehensive periodontal therapy, teeth were retained in only two patients, in six patients, all teeth were extracted almost entirely due to periodontal reasons and in four patients, implants were placed but three patients showed peri-implantitis already.<sup>7</sup> This shows how challenging is the case of Papillon-Lefevre syndrome for periodontal management.

However, if early diagnosis and comprehensive periodontal therapy and proper maintenance is rendered, teeth can be retained for longer time and improve the quality of life of patient (Tinanoff et al. 1995, Wiebe et al. 2001, Lux et al. 2005). For this, parents are suggested to show to clinicians if their children have early loss of teeth and if hyperkeratotic lesions are seen on skin to diagnose the case early and anticipate prompt treatment.

Periodontist should do thorough intraoral and extraoral examination and proper medical history to prevent miss out of case. In this case, patient presented late at age of 25 years with loss of many teeth and counselled properly for the condition and sent for possible prosthodontic rehabilitation but patient lost follow up without fabrication of prosthesis.

**Conflict of Interest:** None.

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