

Management of Gingival Enlargement in Papillon-Lefèvre Syndrome Using Diode Laser-Assisted Gingivectomy: A Case Report

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Abstract

Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive disorder characterized by progressive periodontitis and palmoplantar hyperkeratosis, associated with CTSC gene mutations causing cathepsin C deficiency. This case report presents the diagnosis and management of gingival enlargement in an 18-year-old female with PLS. Clinical examination revealed gingival inflammation, mobile teeth, and periodontal pockets. The patient faced challenges in understanding and maintaining proper oral hygiene. The treatment plan included scaling and root planing, extraction of a hopeless lower central incisor and a laser-assisted gingivectomy to achieve aesthetic and functional outcomes. This case highlights the importance of early diagnosis and a comprehensive treatment approach for managing the oral manifestations of Papillon-Lefèvre syndrome, aiming to improve both oral health and the patient's quality of life.

Keywords: Cathepsin C, Gingival enlargement, Gingivectomy, Oral hygiene, Papillon-Lefèvre syndrome, Periodontitis.

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1. Introduction

Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive disorder characterised by a triad of features: severe, early-onset periodontitis affecting both the deciduous and permanent dentitions, palmoplantar hyperkeratosis, and susceptibility to recurrent infections [1,2]. It has an estimated prevalence of 1 in 4 million individuals, and approximately 2 to 4 out of every 1,000 people are carriers of the condition, with no gender or known racial predisposition.[3] Parental consanguinity is reported in approximately 20% to 40% of cases.[4] The management of Papillon-Lefèvre syndrome remains challenging because of the aggressive periodontal destruction associated with the disorder. Successful treatment requires early diagnosis, meticulous periodontal therapy, rigorous plaque control, and long-term maintenance. Conventional treatment protocols

generally include comprehensive oral hygiene instructions, scaling and root planing, extraction of hopeless teeth to eliminate persistent sources of infection, systemic antibiotic therapy targeting periodontal pathogens, regular periodontal maintenance visits, and prosthetic rehabilitation when required. Dermatological manifestations are commonly managed using topical emollients, keratolytic agents, and systemic retinoids under dermatological supervision. Despite these measures, the long-term prognosis of the dentition often remains guarded because of the progressive nature of the disease.

In recent years, lasers have emerged as valuable adjuncts in periodontal therapy owing to their ability to provide precise soft tissue incision, effective hemostasis, reduced bacterial

contamination, minimal postoperative discomfort, and improved wound healing. Diode lasers are particularly advantageous in gingival surgical procedures because they possess a high affinity for pigmented soft tissues, allowing controlled tissue ablation with minimal collateral thermal damage [3]. Laser-assisted gingivectomy has also been associated with improved patient acceptance, reduced intraoperative bleeding, decreased postoperative edema, and enhanced visualization of the surgical field. These properties make diode lasers particularly suitable for patients requiring extensive soft tissue management while minimizing surgical trauma. Although several reports have described the periodontal manifestations and multidisciplinary management of Papillon-Lefèvre syndrome, published literature describing the use of laser-assisted gingivectomy in these patients remains limited. Most available reports primarily emphasize conventional periodontal therapy combined with antibiotic administration and dermatological management. Consequently, clinical evidence regarding the role of diode laser-assisted gingivectomy in improving gingival architecture and facilitating plaque control in patients with Papillon-Lefèvre syndrome is still scarce [4].

The present case describes the multidisciplinary management of an 18-year-old female diagnosed with Papillon-Lefèvre syndrome in whom laser-assisted gingivectomy was incorporated as part of comprehensive periodontal therapy following initial nonsurgical periodontal treatment. The report emphasizes the rationale for selecting laser-assisted surgery, discusses its potential clinical advantages, and highlights the importance of continuous supportive periodontal care in maintaining periodontal health in this rare hereditary disorder. Although severe periodontitis represents the characteristic oral manifestation of Papillon-Lefèvre syndrome, gingival enlargement has been reported only in a subset of affected patients. The true prevalence of gingival enlargement in PLS has not been established because the syndrome itself is extremely rare and the available evidence is largely limited to isolated case reports and small case series. When present, gingival enlargement is generally inflammatory in nature and is attributed to persistent plaque accumulation, chronic periodontal infection, and compromised oral hygiene resulting from progressive periodontal destruction. The enlarged gingival tissues may further hinder effective plaque control, thereby perpetuating the inflammatory process and contributing to continued periodontal deterioration [5].

Management of gingival enlargement in patients with Papillon-Lefèvre syndrome should be individualized according to the severity of periodontal destruction and the extent of soft tissue involvement. Initial treatment primarily consists of comprehensive oral hygiene instruction, scaling and root planing, professional plaque control, and appropriate antimicrobial therapy to reduce the inflammatory burden. Surgical intervention is considered when gingival enlargement persists despite nonsurgical therapy or interferes with oral hygiene, mastication, speech, or esthetics. Conventional scalpel gingivectomy, periodontal flap surgery, electrosurgery, and laser-assisted gingivectomy have all been described as treatment options. However, because of the aggressive nature of the underlying periodontal disease, all surgical procedures should be combined with meticulous supportive periodontal therapy and regular maintenance visits to minimize recurrence.

2. Case report

An 18-year-old female patient was referred to the Department of Periodontics, with the chief complaint of loose upper and lower front teeth and bleeding gums for the past year. Her medical history indicated the presence of recurring skin rashes and thickened, keratotic patches on the elbows, knees, and soles, since age four. Past dental history of the patient revealed premature loss of primary teeth, and family history revealed parental consanguinity, with no other affected family members.

Extraoral assessment of the patient revealed distinct, yellowish keratotic plaques on the palms as shown in Figure 1 and soles as shown in Figure 2, extending onto the dorsal aspects of both regions. The affected skin appeared to be peeling, exposing a shiny, reddish underlying surface. Comparable hyperkeratotic lesions were also observed bilaterally on the knees.

On intraoral examination, the teeth exhibited Grade I mobility and Grade II mobility, along with class II Recession in the mandibular right central incisor. The gingiva appeared erythematous, edematous and enlarged with a tendency to bleed spontaneously, as shown in Figure 3.

Treatment protocols included standard periodontal therapy comprising oral hygiene instructions, professional scaling and root planing, regular recall visits, and antibiotic administration: Cap Amoxicillin – 500mg twice a day for 5 days, and Tab Metrogyl (FLAGYL) – 400mg twice a day for 5 days.

However, due to poor oral hygiene maintenance, the hopeless right mandibular central incisor has been extracted, as shown in Figure 4.



Figure 1. Thickened, keratotic lesions observed on the dorsal aspects of both palms.



Figure 2. Desquamation of the epidermis on the feet..



Figure 3. Pre-operative image showing erythematous and enlarged gingiva.



Figure 4. Pre-operative image showing erythematous and enlarged gingiva.



Figure 5. Laser-assisted gingivectomy using diode laser.

To correct gingival overgrowth and improve esthetics, Laser-assisted gingivectomy was performed under local anaesthesia using a Clean-Cut Dental Diode Laser operating at a wavelength of 980 nm (Figure 5). The laser was used in pulse mode with a power output of 2.5 W. Pulse settings consisted of an ON time (T_{on}) of 900ms and an OFF time (T_{off}) of 700ms using the pre-programmed gingivectomy setting. The enlarged gingival tissue was excised using controlled sweeping movements while maintaining the physiological gingival contour. Simultaneous coagulation during laser application provided excellent hemostasis, improved visualization of the surgical field, and minimized intraoperative bleeding. Standard laser safety protocols, including the use of protective eyewear by the patient and all operating personnel, were strictly followed throughout the procedure. The Orthopantomogram revealed generalized horizontal bone loss as shown in Figure 7. The missing teeth were replaced, and the patient was referred to the Dermatologist for management of cutaneous lesions. All routine clinical investigations were within the normal limits. A thorough clinical assessment, accompanied by laboratory and histopathological investigations, confirmed the diagnosis of Papillon-Lefèvre Syndrome.



Figure 6. Post-operative (80Days).

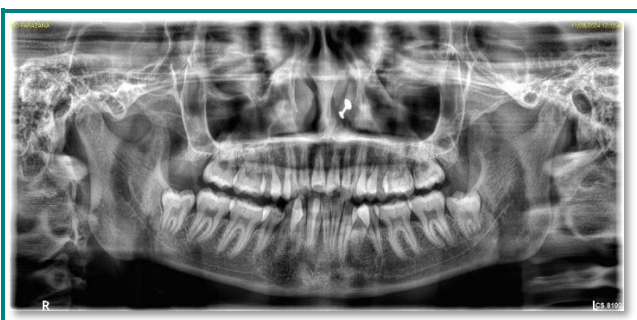


Figure 7. Orthopantomogram showing generalized bone loss.

3. Discussion

Papillon-Lefèvre Syndrome is believed to involve a complex interaction of genetic susceptibility, immunological impairment, and microbial colonization [6,7], though the exact pathogenesis remains uncertain [1]. Recent Advancements in genomic research have highlighted Cathepsin C protein as a major contributor to the pathophysiology of Papillon-Lefèvre Syndrome. The increased risk of periodontitis in individuals with Papillon-Lefèvre Syndrome may be related to Cathepsin C gene mutations [6,8,9] that disrupt normal epithelial differentiation or desquamation [10]. In addition to genetic and immunologic factors, a bacterial origin has been proposed, involving key periodontal pathogens like *A. actinomycetemcomitans*, *P. gingivalis*, *F. nucleatum*, and *P. intermedia* in the progression of periodontal disease in PLS [11]. The differential diagnosis includes Haim-Munk Syndrome and Hypophosphatasia. Haim-Munk Syndrome shares similar features with Papillon-Lefèvre Syndrome, but the patients also present with arachnodactyly of the hands which is a typical feature of Haim-Munk Syndrome.

Hypophosphatasia also shows palmoplantar keratodermitis along with progressive

periodontitis. But hypophosphatasia also appears with deficiency of alkaline phosphatase, the values of which are within normal limits for our patient [1]. The use of diode lasers in periodontal soft tissue surgery has gained increasing attention because of their favourable interaction with haemoglobin and melanin, enabling precise soft tissue excision with simultaneous coagulation. Diode lasers have been widely employed for gingivectomy, gingivoplasty, frenectomy, operculectomy, depigmentation, and other minor periodontal surgical procedures. Compared with conventional scalpel surgery, diode laser-assisted gingivectomy has been associated with reduced intraoperative bleeding, improved visualization of the surgical field, decreased postoperative pain and edema, minimal need for sutures, and favourable wound healing. These characteristics make diode lasers particularly useful in the management of inflammatory gingival enlargement and other soft tissue conditions requiring precise tissue recontouring. A recent systematic review evaluating diode laser gingivectomy reported that patients treated with diode lasers experienced significantly lower intraoperative and postoperative bleeding and reduced postoperative pain compared with conventional scalpel gingivectomy and nonsurgical periodontal therapy. However, the authors also emphasized that the available evidence remains limited because of the small number of randomized clinical trials and methodological heterogeneity among studies [12].

The management of gingival enlargement associated with Papillon-Lefèvre syndrome remains challenging because the inflammatory enlargement frequently coexists with extensive periodontal attachment loss. Published reports have described a multidisciplinary treatment strategy incorporating mechanical periodontal debridement, systemic antibiotics directed against periodontal pathogens, extraction of teeth with hopeless prognosis, conventional gingivectomy, periodontal flap procedures, prosthetic rehabilitation, and dermatological management with systemic retinoids when indicated. More recently, diode laser-assisted gingivectomy has been explored as an adjunctive surgical modality because of its excellent hemostatic properties, precise soft tissue ablation, improved visibility of the operative field, and favorable postoperative healing. Although the current evidence is largely confined to case reports and small case series, laser-assisted surgery appears to be a promising alternative for the management of inflammatory gingival enlargement when integrated into a

comprehensive periodontal treatment plan [1,13-16].

4. Limitations

Papillon-Lefèvre Syndrome is known to have a high risk of recurrence of periodontal destruction and gingival enlargement. Accordingly, the patient has been enrolled in a strict supportive periodontal maintenance program with regular recall visits for professional periodontal care, reinforcement of oral hygiene, and early detection and management of recurrence. The patient was followed for approximately 80 days following surgery, during which satisfactory healing and improvement in periodontal condition were observed. We agree that longer follow-up would provide additional information. No genetic testing for CTSC gene mutation or immunological investigations were performed due to the limited availability of these diagnostic facilities.

5. Conclusion

In conclusion, the management of PLS is challenging and a multifactorial approach should be undertaken. Novel approaches like specific antibiotic regimens and retinoid usage can be implemented. Pyogenic liver abscess is now being increasingly reported as a systemic complication of Papillon-Lefèvre syndrome, primarily attributed to underlying immune dysfunction associated with the disorder. However, with the identification of underlying genetic defects of the Cathepsin C gene and better treatments targeted at the chromosomal defect, stem cell therapy should be developed, which may open new horizons for treatment of PLS patients.

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