


Papillon-lefèvre syndrome: clinical case of oral rehabilitation with osseointegrated implants

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Abstract:

Papillon-Lefèvre syndrome (PLS) is a rare benign, autosomal recessive condition caused by a mutation in the cathepsin C gene. This alteration results in palmar-plantar hyperkeratosis, or thickening of the soles of the feet and palms of the hands, as well as aggressive periodontal disease resulting in the premature loss of deciduous and permanent teeth. The etiology is multifactorial, and is influenced by immunological, genetic or microbial factors. This case report presents a successful prosthetics oral rehabilitation with osseointegrated implants in a 48-year-old female patient with PLS. Although few reports of dental implants in patients with PLS have been published, it seems to be a possible approach in selected patients.

Keywords: Papillon-Lefèvre syndrome; Dental implants; Rehabilitation.

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INTRODUCTION

PLS is a rare benign, autosomal recessive condition that manifests with palmar-plantar hyperkeratosis, or thickening of the soles of the feet and palms of the hands, and aggressive periodontal disease involving both the deciduous and permanent dentition¹. The syndrome is estimated to have a frequency of 1-4 per million^{2,3}, with no predilection for gender or race³. The etiopathogenesis is uncertain, but it is known to be caused by mutations in the gene encoding cathepsin C located on chromosome 11q14². Cathepsin C plays an important role in epithelial differentiation and desquamation, as well as being fundamental in controlling the immune response of leukocytes². Thus, inactivation of cathepsin C results in a series of clinical symptoms associated with PLS, such as decreased epithelial differentiation leading to hyperkeratosis of the palms and soles, as well as periodontal breakdown and early tooth loss due to the reduced neutrophil response in the fight against oral infection by *Staphylococcus spp.* and *Aggregatibacter actinomycetemcomitans*³.

Treating this condition requires a multidisciplinary team. In the dental field, efforts have been made to seek treatments other than simple tooth extraction⁴. Rehabilitation using dental implants is poorly documented in the literature⁵. Although a recent systematic review with meta-analysis indicates good safety and a low failure rate, few studies and a very restricted sample can be evaluated due to the scarcity of information, since it is a rare condition¹. Therefore, dental implants can be a viable treatment option for these patients, overcoming aesthetic and functional oral problems⁶.

Therefore, this article aims to report a clinical case of a patient with PLS who was rehabilitated with a fixed prosthesis under osseointegrated implants in the mandible, presenting the patient's history, clinical characteristics, surgical management, and the importance of differentiated follow-up.

CASE REPORT

A 48-year-old female patient with PLS came to a private dental service seeking a new oral prosthetic rehabilitation in her lower jaw. The patient reported that she had no systemic involvement and was in good general health. She also related that she was diagnosed with the syndrome in childhood. She has had skin manifestations since the beginning of her life, but it was around the age of 13 years that oral manifestations began to appear,

such as periodontal pockets and tooth mobility. In her dental history, the patient said that supportive therapy was carried out in childhood, with scraping, but the teeth remained mobile. During this period, supportive therapy with splinting was also carried out, to keep the teeth as long as possible. At the age of 20, the teeth with advanced mobility were extracted and she was rehabilitated with a partial prosthesis. However, there was always a lot of concern about the preservation of the prosthesis, which had to be changed periodically because there was bone resorption, which caused discomfort for the patient.

The extraoral clinical examination revealed dry scaling, compatible with diffuse palmoplantar keratoderma, located on the palms of the hands and soles of the feet (Figures 1A and 1B). On intraoral examination, edentulous spaces, occlusal collapse, resorbed ridge, and low level of bone insertion of the remaining dental elements were observed, characteristic of periodontal disease (Figures 2A and 2B).

The patient reported that she had been uncomfortable with her lower removable partial prosthesis since childhood, due to their constant maladjustment, which caused discomfort when chewing and speaking. Due to this complaint, treatment plans were suggested that included extraction of the remaining teeth, regularization of the ridge, installation of implants, and molding for prosthesis on the same day of the surgery. Dexamethasone 4mg and amoxicillin 2g were prescribed 1 hour before surgery. Four implants (Strong SW plus



Figure 1. (A-B) Clinical images of transgressive and diffuse palmar and plantar keratosis.

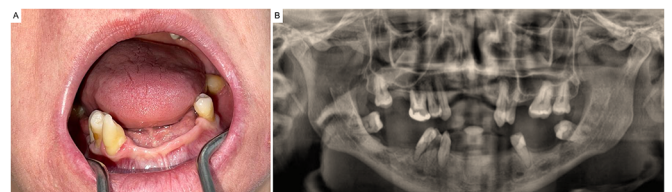


Figure 2. (A) Intraoral clinical image emphasizing the lower jaw. It shows a partially edentulous patient with a thin and atrophic alveolar ridge. (B) Panoramic X-ray image showing missing teeth.

38x15.Sin®) were installed between the mental foramen, two vertical anterior and two angled posterior implants, all of which achieved a torque of over 45N, making immediate loading possible. The prosthesis has an internal welded titanium bar, which generates better force dispersion, and is encased in acrylic (Figures 3A and 3B). Three days after the surgery, the prosthesis was placed (Figure 3C).

After 3 months of follow-up, the patient reported no pain or discomfort, and oral hygiene, and implant maintenance were satisfactory (Figure 4A and 4B). No implants have been lost or are mobile and there was no bone loss around the implants. The patient was advised to follow up every six months for implant health assessment appointments.

DISCUSSION

PLS is an extremely rare genodermmatosis that often goes undiagnosed until the patient develops an affected periodontium, thereby creating a significant psychological and social impact on the person diagnosed with the syndrome^{1,7}. The prevalence is between 1 and 4 cases per million people, with no predominance by gender or race and an uncertain cause, demonstrating a complex etiology, including genetic, immunological and microbiological factors⁸.

The diagnosis of this syndrome is purely clinical and shows both extraoral and intraoral manifestations^{1,9}. Both situations were evident in this case. The first, keratoderma, was more discreet on the palms of the hands

compared to the soles of the feet. As for the intraoral manifestation, the patient had few remaining teeth, since the syndrome directly affects the periodontal tissues. It is well known that palmar-plantar keratosis coexists with large gingivitis of primary teeth together with rapid periodontal deterioration and premature exfoliation of primary teeth at around four to five years of age, a fact that distinguishes PLS from other conditions^{1,10}. It is worth noting that the same sequence of events also recurs when the permanent dentition erupts¹.

Studies show that patients with PLS may have deficiencies in the function of neutrophils, lymphocytes and monocytes, thereby increasing their vulnerability to bacterial infections and the risk of developing recurrent pyogenic diseases, including liver abscesses^{1,3,11}. Almuneef et al (2003)¹², showed that pyogenic liver abscess can be recognized as a complication of PLS due to impaired immune system function. The patient in the present case did not report altered liver function or other systemic involvement.

The oral rehabilitation of PLS patients with dental implants is a challenge particularly because the scarce amount of bone^{1,13}. In the current case, only 4 implants (two vertical anterior implants and two angled posterior implants) were used, all in the anterior region of the mandible allowing immediate loading. A systematic review on the success rates of dental implants in individuals with PLS concluded that there is limited evidence available to suggest that the survival rate of dental implants in patients with PLS is similar to that of healthy individuals¹. However, no strict contraindications for implant-supported prosthesis were justified in these groups of patients in the 10 studies included in the review. Atarbashi-Moghadam et al (2020)¹, reported that the implant failure rate in the maxilla (9%) was higher than in the mandible (2%) in PLS patients. Therefore, a delayed loading protocol was adopted to prevent complications and future implant failures.

Although the success rates of oral rehabilitation with dental implants in patients with PLS seems to be lower compared to those without the disease, most previous studies have reported that it can still be a viable treatment option with a good prognosis and excellent durability^{1,13-15}.

Our PLS patient was successfully rehabilitated with osseointegrated implants and no implant failed in the mandibular arch, all implants remaining fully functional during the 3-month follow-up period. According to the patient, the rehabilitation has improved



Figure 3. (A) Intraoral clinical image showing the 3-day postoperative period after implant installation. (B) Image of the prosthesis before installation in the oral cavity. (C) Image of the prosthesis installed 3 days after surgery.

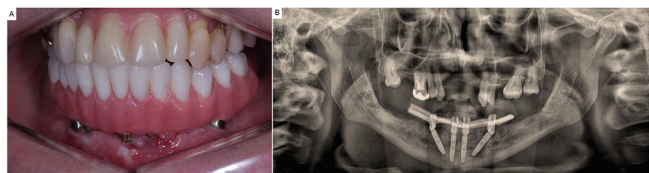


Figure 4. (A) Clinical image showing centralization, together with the upper removable partial prosthesis, 3 months after installation of the osseointegrated implants. (B) Panoramic X-ray image showing follow-up after 3 months. The implants are well adjusted to the lower dental arch.

her functional well-being, as well as her psychological and aesthetic well-being.

CONCLUSION

Early detection of this syndrome is essential to improve the patient's general well-being. It is common for patients with PLS to be characterized due to their early edentulism status. Therefore, at a certain point, the only solution is oral rehabilitation, and the dentist is responsible for individualized planning for each patient, including long-term follow-up. Rehabilitation using osseointegrated implants for PLS patients is a viable treatment option, as implantation can help to preserve the alveolar bone and prolong the maintenance program, if they are maintained.

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