Prosthodontic Rehabilitation of Patients with Papillon - Lefevre Syndrome: A Case Report.

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Abstract

Papillon-Lefevre syndrome (PLS) is an autosomal recessive disorder. The main features of this disease are hyperkeratosis of palms and soles together with premature loss of primary and permanent dentition due to progressive periodontitis. Dental management of patients with PLS is usually challenging because of early excessive loss of alveolar bone support and very few remaining teeth. We describe the rehabilitation of a 18-year-old patient with PLS with severely atrophic mandible with few remaining teeth. Metal suprastructure (modified copings) were fabricated to cover the remaining teeth followed by overdentures. Dentures can be well retained by using support of the remaining teeth. In patients with PLS, this technique can be safely used instead of invasive preprosthetic applications such as bone augmentation, implants or alveolar distraction.

Key Words: *Papillon-Lefevre syndrome, modified copings, overdentures, atrophic mandible.*

Introduction

Papillon-Lefevre syndrome (PLS) is an autosomal recessive disease that was first described by French physicians Papillon and Lefevre. PLS is characterized by hyperkeratosis of the palms and soles, dural calcification, and premature loss of primary and permanent dentitions due to progressive periodontitis.[1] This syndrome is thought to be caused by mutations in the Cathepsin C gene that generates a lysosomal protein, inflammatory mediation, and extracellular matrix deposition which results in hyperkeratosis of the palms soles and gingivae.[2] The prevalence rate of this syndrome is between 1 to 4 per million.[3] No sexual or racial predominance seem to exist.[4] The clinical features of PLS usually become obvious before the age of 4 years. After eruption of primary teeth, the gingivae become inflammed, and is generally followed by a rapid destruction of the periodontium. Premature loss of primary teeth is seen in childern with PLS.[5] Conventional periodontal therapy and systemic administration of various antibiotics usually fail in patients with PLS, and the rapid progression of periodontitis often results in severe loss of the alveolar bone. [6] Early extractions of all permanent teeth is recommended to preserve the remaining supporting bone.[3] Microbiological studies of plaque samples from patients with PLS have implicated *Actinobacillus actinomycetem-comitans, Porphyromonas gingivalis, Fusobacterium nucleatum*, and *Treponema denticola* organisms as examples of the many pathogens that may be involved in this disease.[7] *A. actinomycetemcomitans* is most commonly responsible in the pathogenesis of the rapid periodontal destruction in patients with PLS.[8]

In addition to the dermatologic and oral features, patients may have decreased neutrophil, lymphocyte, or monocyte functions and an increased susceptibility to bacterial infections, leading to persistent pyogenic infections of the skin.[9] These patients have only few remaining teeth. They also have alvelolar bone loss, therefore retention of dentures is a problem. To avoid bone loss and to enhance retention of dentures, copings with metal band casted on it, are provided to teeth. This attachment is preferred as managing patients with Papillon–Lefevre Syndrome with dental implants is troublesome because of the early loss of dentition and severe atrophy of the remaining bone. The aim of this article is to discuss management of this disease as well as review literature available on the subject.

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Case Report

A 18 year old boy who had lost his dentition due to progressive periodontitis and was totally edentulous since the last 5 years, presented with features of Papillon–Lefevre Syndrome. He had hyperkeratosis of both plantar, both knee, and both ankle regions (Figure1). A panoramic radiograph of the patient revealed generalized severe resorption of the alveolar bone, both in the mandible and the maxilla, with very few remaining teeth (Figure 3).

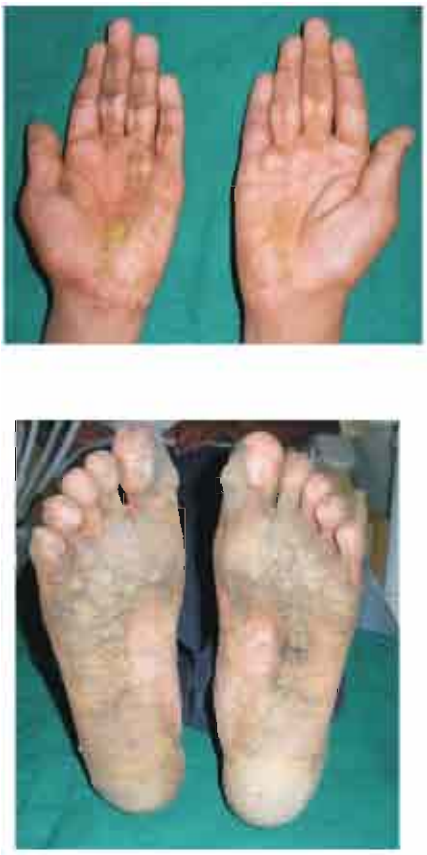

Fig. 1: Hyperkeratotic lesions of the patient at the plantar and palmar surface.

Intraoral and radiological examination

Intraoral examination revealed 2 permanent teeth in the maxillary arch and 2 permanent teeth in the mandibular arch (Figure 2). Radiographic examination revealed that the condition of the remaining teeth were satisfactory and they could be used for overdenture (Figure 3). After carefully studying the casts, radiographs and clinical features, it was decided that all the remaining teeth be retained.

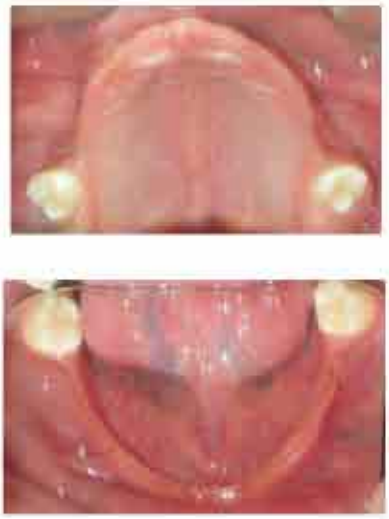

Fig. 2: Preoperative intraoral photograph : Maxillary and Mandibular arch.

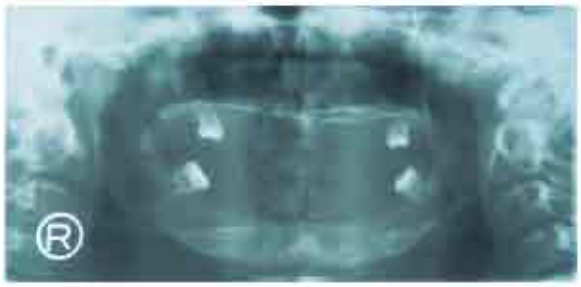

Fig. 3: Orthopantomograph.

Fabrication of Prosthesis

An overdenture was planned on the remaining teeth. The teeth were prepared to receive modified metal copings (Figure 4). The dentures were processed in heat cured acrylic resin. Biofunctional Prosthetic System (BPS) dentures were fabricated (Figure 6). In areas adjacent to copings the dentures were lined with soft liners (Figure 5). Denture were placed in the patient's mouth and occlusion and extensions were checked (Figure 7). Recall appointments were scheduled after one day, one week and two weeks and required minor adjustments made to patient's satisfaction.

Fig. 4: Master casts showing prepared abutments with modified metal copings.

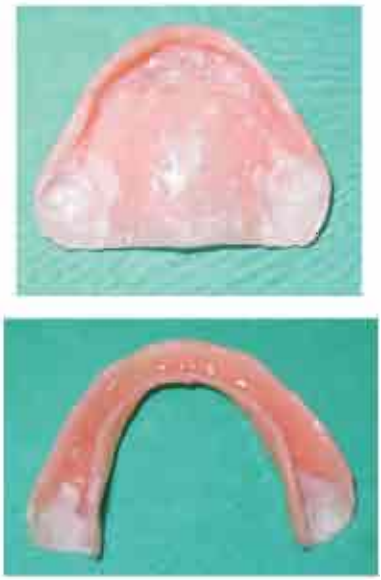

Fig. 5: Denture Lined with Soft liner for retention with modified coping.

Fig. 6: Denture insertion complete.

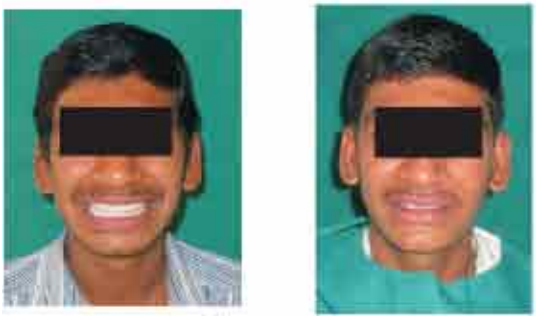

Fig. 7: Pre and Post operative photographs.

Discussion and Summary

Papillon–Lefevre Syndrome is a rare autosomal syndrome characterized by palmoplantar hyperkeratosis and periodontal disease. Although it is unclear, immunologic, genetic, and microbiologic factors are thought to play a role in its etiology.[10] After eruption of primary teeth, the gingivae become inflamed followed by rapid destruction of the periodontium. Both the diagnosis and treatment of the periodontal component of Papillon–Lefevre Syndrome is very difficult. The differential diagnosis include Hiam-Munk syndrome and hypophosphatemia. Hiam-Munk syndrome is characterized by arachnodactyly, atrophy of nails, and deformity of the phalanges in the hands. The patient in this report had no structural deformities in her upper and lower extremities.[7-11]

Psycho-social condition of a patient with Papillon–Lefevre Syndrome tends to be affected because of loss of teeth at a young age, therefore, early dental evaluation is a important step in psychological rehabilitation. In PLS, primary dental care includes treatment of aggressive periodontitis, as conventional periodontal treatment usually fails and the rapid progression of periodontitis often results in a severe loss of alveolar bone.[12] Subsequently, prosthetic management of these patients is difficult due to early excessive vertical bone loss. Early extraction of teeth in place of long-term periodontal treatment can preserve alveolar bone height and facilitate further prosthodontic rehabilitation by use of overdentures. An overdenture is, denture constructed over existing teeth or root structures. The use of overdenture is not a new concept. It has become increasingly popular as the emphasis on preservation of teeth in dentistry has increased.[13]

Overdenture accomplishes three important goals:

1. It maintains teeth as part of the residual ridge.
2. It decreases the rate of alveolar bone resorption.
3. It preserves proprioceptive impulses by utlizing modified copings and increases retention for overdentures.

Conclusion

Papillon-Lefevre syndrome is a genetic disorder resulting in loss of teeth and alveolar bone support. It can easily be treated by overdentures with modified copings on the remaining teeth. This is an extremely cost effective procedure. The other modality of treatment is implant supported prosthesis, which is a more expensive procedure.

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