**CASE REPORT**

**Prosthodontic Rehabilitation of Papillon Lefevre Syndrome**

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**ABSTRACT**

Papillon Lefevre Syndrome (PLS) is an autosomal recessive (AR) disorder affecting the skin and intra oral soft tissues resulting in palmo plantar hyperkeratosis with premature periodontal problems leading to early tooth loss and associated functional and psychological disturbances. This report presents management of a 17 years old girl suffering from PLS. She was presented with the chief complaint of loss of many permanent teeth and mobility of the remaining teeth. Most of the teeth were lost very early after their eruption due to swollen infected gums. There were no other associated problems except for the mild burning sensation of the palms and soles. A decision was made and remaining teeth were extracted in order to prevent aggressive bone loss. She was then managed using removable complete dentures with bilateral balanced occlusion. The patient is comfortable and functioning well with her new dentition.

Key Words: Papillon Lefevre syndrome. Prosthodontic rehabilitation. Periodontitis.

**INTRODUCTION**

Papillon Lefevre Syndrome (PLS), a rare genodermatosis autosomal recessive (AR) disorder was first reported by Papillon and Lefevre in 1924. The exact etiology is still unknown. However, impaired functions of leukocytes specially the neutrophils-like chemotaxis, phagocytosis have been identified. Severe periodontitis actinobacillus actinocomitans due to micro-organisms is considered very important. There is a loss of functional mutation affecting both the alleles of Cathepsins C gene, located on chromosome 11q14.1-q14.3. The Cathepsin C genes encode a cystine lysosomal protease, known as dipeptidyl peptidase I, which functions to remove dipeptides from the amino-terminus of protein substrate along with endopeptidase activity. Cathepsin C gene usually expresses itself in the epithelial regions affected by the PLS as well as in various immune cells.

The prevalence of PLS has been reported to be 1 - 4 in one million population, with no racial discrimination and affecting both genders equally. Consanguinity has been observed in one-third of the reported cases. Characteristic features of PLS are palmo-plantar hyperkeratosis and a severe periodontitis destroying the alveolar bone and resulting in the early loss of teeth. Both dentitions are usually involved. Associated features may include calcification of falx cerebi and choroid plexus, increased susceptibility to infections, hypohidrosis of palms and soles, and nail dystrophy. These features had been found in isolation as well as in varying relationships.

Periodontal problems associated with PLS usually do not respond to routine conservative and surgical protocols, and ultimately result in total dental exfoliation. However, a multidisciplinary treatment approach may help to rehabilitate the extra and intra oral symptoms of these suffering subjects. Conventional removable prosthodontics with or without osseointegrated dental implants can help to restore an efficient functional dentition. Psychological reassurance and regular follow-up will be the key to success in these complicated scenarios.

This report presents the management of intra oral problems of a patient suffering from Papillon Lefevre syndrome.

**CASE REPORT**

A 17 years old female was presented to our Outpatient Department with the chief complaint of loss of many permanent teeth and mobility of the remaining teeth. Most of the teeth were lost very early after their eruption due to swollen infected gums and excessive mobility. There were no other associated problems except for the burning sensation of the palms and soles. Patient related the dental problem to neglected oral hygiene. She also mentioned about the loss of primary teeth in the same manner. The patient belonged to a low socio-economic class and was a housemaid. She was married for last 3 years but with no kids. There was no relevant drug history and family history was also not significant.

On extra oral examination, skin and hair were normal except mild hypohydrosis like dry areas on palms, soles and even extended on the fingers. Nails were malformed, dystrophied and striated (Figure 1). Patient also reported these thickened dry areas on knee and elbow joints. Patient had already consulted a dermatologist and was using certain biological serum based skin ointments.

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On intra oral examination, gums were swollen and reddish. There was tendency to bleeding as well. All remaining teeth except the most posterior molars were periodontally compromised with grade III mobility. There was excessive bone resorption as well (Figure 2).

Radiographic investigations were done and the orthopantomogram revealed the typical Papillon Lefevre syndrome appearance of floating teeth (Figure 2).

Laboratory investigations included complete blood pictures, serum calcium, serum phosphatase were done. All findings were normal except a slight increase in the alkaline phosphatase which may be age related. Histopathological Investigation was planned but patient was not compliant for that procedure.

After a detailed discussion, the patient was referred to the Department of Periodontology. Their opinion was for the removal of these severely compromised teeth except for the last standing molars which were kept to retain the future prosthesis as long as possible. The patient was referred to Department of Oral Surgery, where all hyper mobile teeth were extracted. The patient was then finally referred to Prosthodontics Department where she was provided with the bilaterally balanced conventional removable dentures with little retentive help from the most posterior molars. This patient had a regular follow-up. She is wearing these dentures, complying with oral hygiene measures and is happy comfortable with her new smile and teeth.

**DISCUSSION**

PLS is characterized by marked destruction of the periodontium (periodontoclasia) of both dentitions with premature loss of teeth, marked palmar and plantar hyperkeratosis. The gene has been mapped to the long arm of chromosome 11. These patients are usually normal at birth with only reddening of palms and soles. Teeth erupt usually in normal sequence, position and time. At around one and an half to two years, marked gingivo-periodontal process develops with edema, bleeding, alveolar bone resorption and teeth mobility with consequent exfoliation. Concurrent with these oral lesions, the palms and soles usually develop a red scaly keratosis that occasionally extends to the dorsal surfaces as well. The oral lesions are complicated by superimposed inflammation and radiographs revealed marked alveolar resorption with vertical pockets of even those teeth whose roots are still incomplete. Teeth are usually lost in the same sequence as of eruption. After exfoliation of all teeth, the soft tissues usually acquire the normal healthy appearance. The permanent dentition usually start to appear at normal time, but just after 2 - 3 years, the gingivo-periodontal condition starts to deteriorate again. All permanent teeth usually exfoliate within a few years except for third molars which usually stay longer. Peripheral blood neutrophil chemotaxis have been reported to be depressed. This decreased chemotaxis suggests that neutrophils may be important factor in periodontal destruction. Current literature suggests involvement of bacterial and viral pathogens as the initiating factors.

No definitive treatment is available for prevention and management of periodontal destruction, although strict oral hygiene maintenance, scaling and root planning along with suitable antibiotic regimen may improve the situation, but in later stages, as with disease progression, all the teeth are usually lost.

The treatment should be planned with a multi-disciplinary team approach involving paediatricians, periodontists, dermatologists, prosthodontists and psychologists. Most therapeutic attempts of the oral periodontal lesions have been unsuccessful. However, edentulous patients can adapt to removable prosthesis very quickly because of young age, better oral stereognostic and oral motor abilities. The osseo-integrated dental implants have revolutionized the possible treatment options, but long-term effects in these syndromic cases are still pending. Treatment of skin lesions with retinoids (oral retinoate) and different nutrient bio-serums have proven to be efficient but must be implemented by the dermatologists. The differential diagnosis should include Ehlers Danlos syndrome and pre-pubertal periodontitis. However, the presenting symptoms were distinguished as there was no joint
mobility or hyperflexed skin and in addition, the aggressive form of generalized periodontitis was not related to the onset of pubertal phase and there were no other systemic complications. There was also history of the periodontal tooth loss of primary dentition.

REFERENCES