Case Report

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PERIODONTAL MANAGEMENT OF PAPILLON LEFEVRE SYNDROME BY PRF: A CASE REPORT

C.S.Prabakar [1], M. Jeevitha [2]

1. Professor and Head, Department of Periodontics, Best dental science college and hospital, Madurai, India.
2. Post graduate, Department of Periodontics, Best dental science college and hospital, Madurai, India.

ABSTRACT:

Papillon Lefevre syndrome is a rare autosomal recessive disorder (1 in 1-4 million people) characterized by palmoplantar hyperkeratosis and early onset periodontitis. This is one of the monogenic conditions which shows resistance to conventional periodontal treatment and hence periodontal management for these patients remain challenging.

Key words: Syndrome, Autosomal Recessive, Hyperkeratosis, Periodontitis, PRF [platelet rich fibrin]

INTRODUCTION:

Papillon lefevre syndrome is first described by Papillon & Lefevre in 1924 in two siblings, the product of a first cousin mating.[1] The hallmarks of papillon lefevre syndrome are palmoplantar hyperkeratosis and rapid periodontal destruction of both dentitions.[1,2,6,10] Increased susceptibility to infection has been reported in approximately 20% of patients [1,6,9] and some cases have reported intracranial calcifications, retardation of the somatic development, follicular hyperkeratosis and onychogryphosis. [1]

ETIOLOGY:

The etiology of papillon lefevre syndrome is still not confirmed. But various authors have suggested possible etiologies. Laass et al.,1997; Fisher et al.,1997; Toomes et al.,1999 suggested a genetic defect mapped to chromosome 11q14 – q21. [1,2,3,6]. Hart et al 1998; Hart et al.,2000 – put forward that the defect is particularly in the interval between D11S4082 & D11S931, where the gene Cathepsin C (CTSC) lies. Oguzkurt et al., 1996 – advocated that lack of functional Cathepsin C may be associated with a host response against bacteria in dental plaque. [2,3,4,6,7]

A genetic predisposition with greater frequency of occurrence in consanguineous offspring has been reported – 20 – 40% cases according to Papillon MM, Lefevre P. Deux in 1924 [5,6,9]. According to Griffiths WAD, Judge MR et al both the sexes are affected equally. [1,2]

CASE DETAIL:

A Patient by name Sharmila Parveen of 19 years age reported to OPD of Best dental science college and hospital, Madurai with the chief complaint of mobile teeth in the upper front and left back tooth region for the past 2 years. For this reason patient has difficulty

*Corresponding Author Address: Dr. Prabakar Email: cyespee@gmail.com

during mastication and there is no associated pain.

MEDICAL HISTORY:

Patient has consulted dermatologist for dryness of skin in palms and feet and was under medication for the same. At present she is not under any medication.

DENTAL HISTORY:

Patient gives history of early exfoliation of primary dentition and have consulted dentist by age of 7 for the above mentioned problem in primary dentition.

PERSONAL HISTORY:

Brushes once a day with tooth paste and brush. Parents gave a history of consanguineous marriage. She has an elder sister who is apparently normal and an elder brother with history of skin alterations and premature loss of primary & permanent dentition.

PHYSICAL EXAMINATION:

Patient had normal physical and mental development for her age.

Cutaneous examination revealed increased keratinization which shows yellowish keratotic & confluent plaques on the palm and feet (Fig.1,2,3). Mild keratosis are noted on knees bilaterally (Fig.4,5). The skin was dry, scaly & rough on palpation. Dystrophy of nails was noted in legs.

INTRAORAL EXAMINATION:

Intraoral examination revealed generalized inflamed gingiva which is soft and edematous. Generalized exudates were evident with deep periodontal pockets and marked mobility in most of the teeth (Fig.6).

RADIOGRAPHIC INVESTIGATIONS:

OPG showed generalized severe horizontal type of bone loss (Fig.7).

Postero anterior view and lateral view of skull (Fig.8 and 9) revealed no evidence of intracranial calcification.

Hematological investigations were within normal limits.

DIAGNOSIS:

Using the detailed case history, clinical examination & investigation reports, a diagnosis of Papillion Lefevre syndrome was made.

TREATMENT:

Patient education and motivation was done before starting treatment procedures. Teeth with hopeless prognosis were extracted (22, 26). Patient was given Amoxicillin 500mg and Metronidazole 400mg for 8 days. Oral prophylaxis and root planing was done and patient was kept under maintenance phase for one month. Patient oral hygiene maintenance was evaluated using simplified oral hygiene Index. Periodontal flap surgeries were carried out in teeth having deep periodontal pockets in each quadrant with intermittent gap. Bone grafts and PRF was placed wherever necessary. (Fig.10,11,12,13,14,15) Patient was advised for chlorhexidine mouth wash for

2 weeks post operatively and has been asked to maintain meticulous oral hygiene. [11]

DISCUSSION:

Papillon Lefevre Syndrome is a rare autosomal recessive disorder which can be due to genetic mutations of the Cathepsin C gene. This can adversely affect growing children psychologically & esthetically. Hence early diagnosis & parental counseling should be given. As etiopathogenesis of PLS is not clear, a multidisciplinary approach may improve the prognosis of patient. [5,6]

Immunologic, microbiologic & genetic bases should also to be considered. Immunologic studies have shown decreased monocyte chemotaxis, decreased neutrophil chemotaxis [2,6], impaired neutrophil phagocytosis [1,2,3,6], altered superoxide production and decreased response of neutrophils. [2]

Microbiologic studies have shown increased presence of Aggregatibacter actinomycetemcomitans, Haemophilus aphrophilus, Prevotella intermedia, Fusobacterium, Treponema denticola, Porphyromonas gingivalis. [1,6] High antibody titres against A.a were also noted in these patients.[1,8] Genetic studies have shown defect in Cathepsin C expression. [2,3,6,7,9,10]

A definite treatment protocol has not been reported. Hence, to control periodontal disease progression different treatment modalities like conventional periodontal therapy, maintenance of oral hygiene, extraction of teeth having poor prognosis, rehabilitation of missing teeth, systemic antibiotics have been suggested. For added advantage we incorporated PRF which has lot of growth factors and has been proved to have prolonged antibacterial property. In present study placement of PRF and bone grafts were given good results.

CONCLUSION:

Most of the PLS patients were identified and diagnosed by dental professionals. Hence thorough knowledge about this syndrome may help the dentist to make a diagnosis at an early age. If left untreated, PLS may lead to edentulism at an early age which makes early diagnosis & intervention necessary. Proper oral hygiene maintenance and flap surgeries with bone grafts and Platelet Rich Fibrin as early as possible have given better results.

The role of periodontists is more in treating patients with Papillon Lefevre Syndrome, awareness about this syndrome should be made through education and conducting awareness programs.

REFERENCES

1. Hart TC, Shapira L. Papillon-Lefevre syndrome. Periodontology 2000. 1994 Oct 1;6(1):88-100.
2. Kumar TD, Pillai AK, Kulkarni P, Moghe S, Vishnu V, Yadav SD. Papillon-Lefevre Syndrome-A Brief Review of Diagnosis & Management. Journal of Dental and Medical Sciences. 2014 Nov;13(11):92-95. Journal of Dental

and Medical Sciences 2014 Nov;13(11):92-95.
3. Ashri NY. Early diagnosis and treatment options for the periodontal problems in Papillon-Lefevre syndrome: a literature review. Journal of the International Academy of Periodontology. 2008 Jul;10(3):81-6.
4. Kaur S, Verma A, Grover V. Periodontal complications and management of papillon-lefevre syndrome-A case report. Baba Farid University Dental Journal. 2013;4(1):152-5.
5. Kaur B. Papillon Lefevre syndrome: a case report with review. Dentistry. 2013 Aug 6;2013.
6. Khan FY, Jan SM, Mushtaq M. Papillon-Lefèvre syndrome: Case report and review of the literature. Journal of Indian Society of Periodontology. 2012 Apr 1;16(2):261.
7. Ullbro C, Brown A, Twetman S. Preventive periodontal regimen in Papillon-Lefevre syndrome. Pediatric dentistry. 2005 May 1;27(3):226-32.
8. Preus HR. Treatment of rapidly destructive periodontitis in Papillon Lefèvre syndrome. Journal of clinical periodontology. 1988 Nov 1;15(10):639-43.
9. Mercy P, Singh A, Ghorpade AK, Das M, Upadhyay A. Papillon-Lefevre syndrome: two siblings, one developing liver abscess. Indian J Dermatol. 2013 Sep 1;58(5):410.
10. Hart TC, Hart PS, Bowden DW, Michalec MD, Callison SA, Walker SJ, Zhang Y, Firatli E. Mutations of the cathepsin C gene are responsible for Papillon-Lefevre syndrome. Journal of Medical Genetics. 1999 Dec 1;36(12):881-7.
11. Reenesh M, Singh M, Rath SK, Mukherje M. Papillon-Lefèvre syndrome associated with Aggressive periodontitis: A rare case report. Indian Journal of Dental Advancements. 2012 Jul 1;4(3):956-9.

FIGURES:

Fig.1: Keratotic plaques on the palm

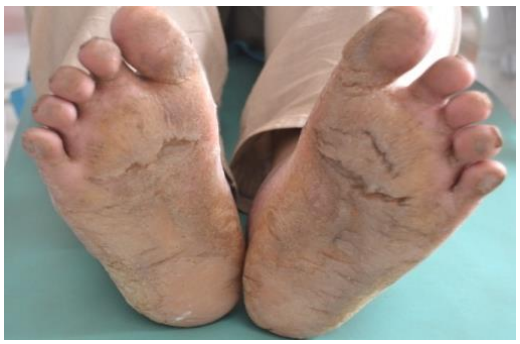


Fig 2: Keratotic plaques on feet

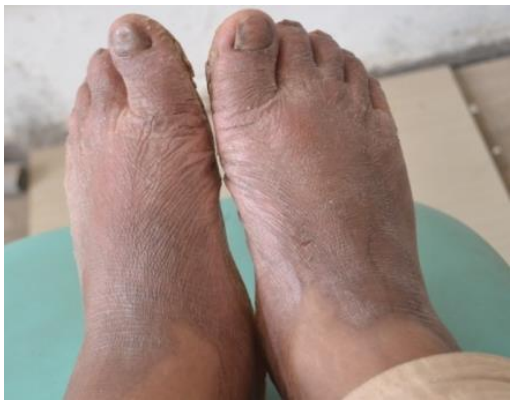


Fig 3: Keratotic plaques on feet

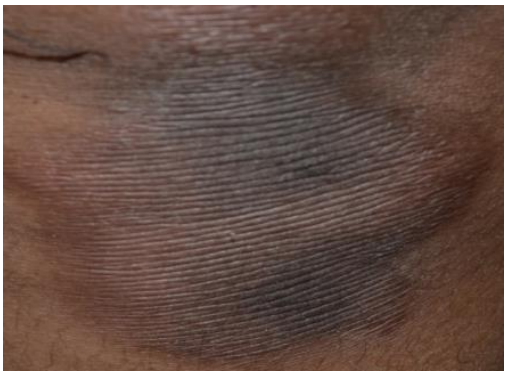


Fig 5: Mild keratosis on knees

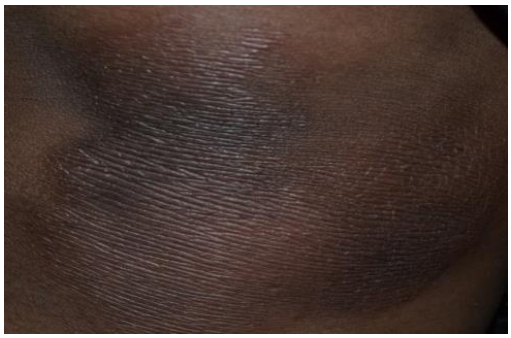


Fig 4: Mild keratosis on knees

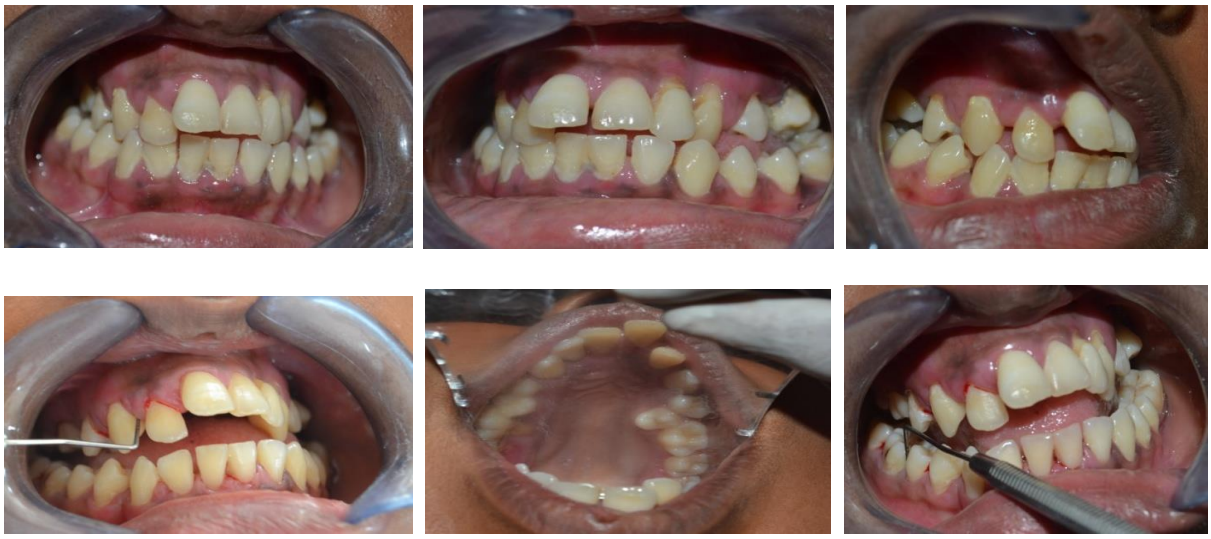


Fig.6: Intra oral photographs

Fig.7: OPG

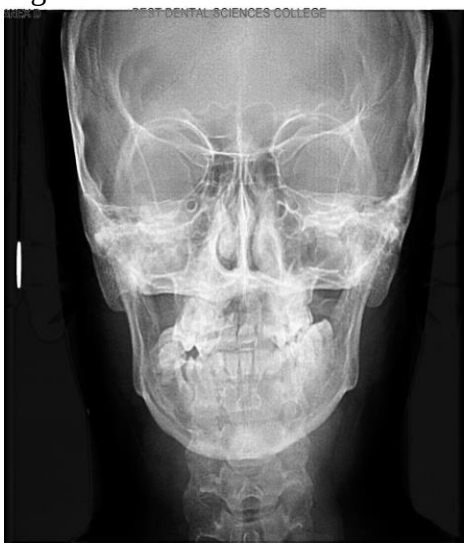


Fig.8: PA view of skull

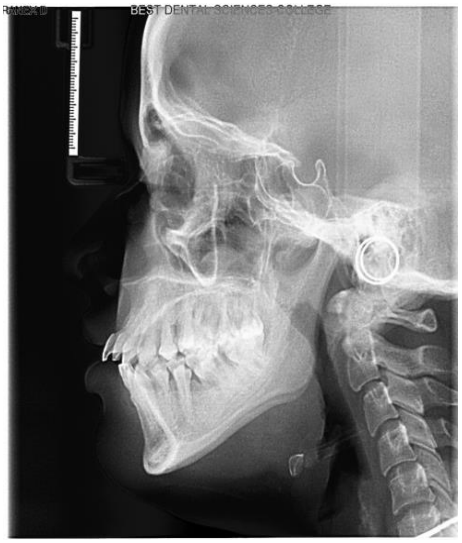
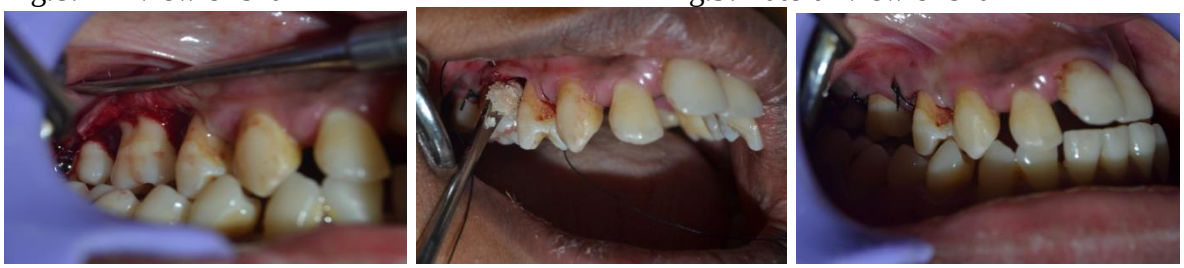


Fig.9: Lateral view of skull



Flap elevation Placement of bone graft Sutures placed

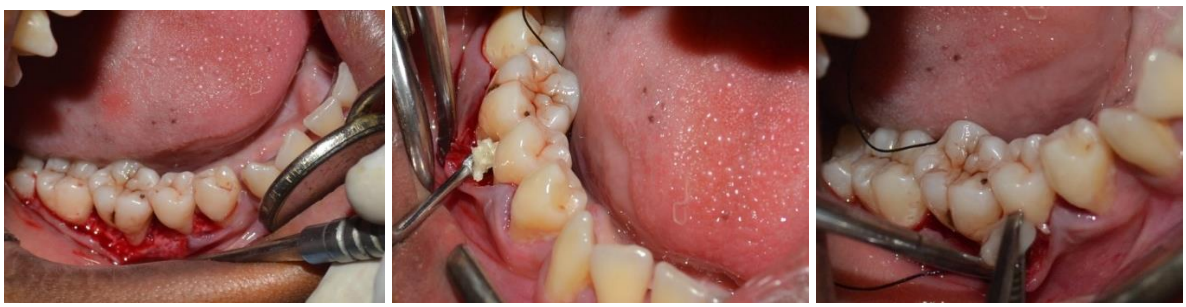
Fig.10 Flap surgery in I quadrant

Flap elevation Placement of Platelet Rich Fibrin Sutures placed

Fig.11 Flap surgery in II quadrant

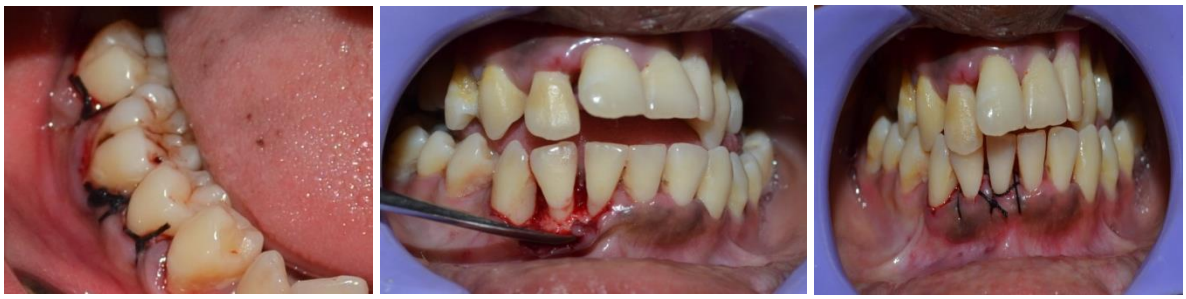
Flap elevation Sutures placed

Fig.12 Flap surgery in lower anterior region



Flap elevation Placement of Bone graft Placement of Platelet Rich Fibrin

Fig.13 Flap surgery in III quadrant



Sutures placed Flap elevation Sutures placed

Fig.14 Flap surgery in lower anterior region

Papilla Preservation Flap Flap elevation Sutures placed

Fig.15 Flap surgery in upper anterior region