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# PAPILLION LEFEVRE SYNDROME
## - *A CASE REPORT*

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**ABSTRACT**
Papillion Lefevre syndrome (PLS) is a rare disease associated with skin lesions, severe destruction of periodontium and in some cases, calcification of dura. Skin lesions consist of hyperkeratosis and icthyosis of localized areas on palms, soles, knees and elbows, multiple functional neutrophil defects, including myeloperoxidase deficiency as well as defective chemotaxis and phagocytosis.

**KEY WORDS:. Papillion Lefevre syndrome (PLS), Hyperkeratosis, Neutrophil, periodontitis, phagocytosis.**

## INTRODUCTION

Papillon-lefevre syndrome is a rare autosomal resessive disorder charecterised by palmar plantar hyperkeratosis and early onset periodontitis[1,2]. PLS is caused by mutations in the cathepsin C gene, located on chromosome[1,3,4]. CathepsinC is a CYSTEINE PROTEASE normally expressed at high levels in various tissues, including epithelium and immune cells such as PMN'S.[5,6] PLS patients who lack almost all cathepsin C activity, are either mutant homozygotes or compound heterozygotes. In some PLS patients, EARLY ONSET PERIODONTITIS is associated with the virulent microorganism Actinobacillus actinomycetemcomitans.[7,8]

Papillion – Lefevre syndrome is inherited and appears to follow an autosomal recessive pattern.[9]. Parents are not affected and both must carry the .autosomal genes for the syndrome to appear in the offspring. It may occur in siblings. Males and females are equally affected. Periodontal involvement consists of early inflammatory changes that lead to bone loss and exfoliation of teeth. Primary teeth are lost by 5 to 6 years of age. The permanent teeth then erupt normally but within a few years the permanent teeth are lost owing to destructive periodontal disease. By the age of 15 yrs, patients are usually edentulous but for the 3rd molars. These are also lost a few years after they erupt. Tooth extraction sites heal uneventfully[10] Estimated frequency is 1 to 4 cases in a million. Rare cases of adult onset of this syndrome albeit with mild periodontal lesions have also been described.

**Case Report:**
Patient aged 15 years approached with a complaint of mobility of teeth since 2 years. He also complained of early loss of deciduous teeth and gaps between the permanent teeth**( Fig.1**). General examination showed extensive thickening of the palms, (both on dorsum **(Fig.2)** and palmar surfaces **(Fig.3).** soles and skin. Patient was always wearing gloves and socks and doesn't like anybody to see his hands and feet. There was generalized coarseness of skin. Patient is very shy and doesn't communicate properly. On examination, the gingiva is red with bleeding on probing. Deep periodontal pockets ranging from 5 to 7mm are present. There is pathologic migration of teeth with GrII mobility [Figno.1] Mild gingival recession [1 to 2mm] is present in some regions.

**Diagnosis:** The patient was diagnosed as having Papillion – Lefevre syndrome with aggressive periodontitis.

**Treatment:** Patient was explained about his condition. Oral prophylasis is done followed by curettage. Flap surgery was planned after one month. By this time all the clinical parameters have improved. A sulcular insicion flap **(Fig.4)** was raised with papilla preservation technique wherever possible. Bone grafting was done in regions of infrabony defects**(Fig.5).** Patient is kept on Chlorhexidine mouthwash. sutures are removed after one week. Heeling is good without signs of inflammation.

**Conclusion:** Today, we have very good treatment options. We are in a stage to save the teeth even in Aggressive periodontal disease. Emphasis should be laid

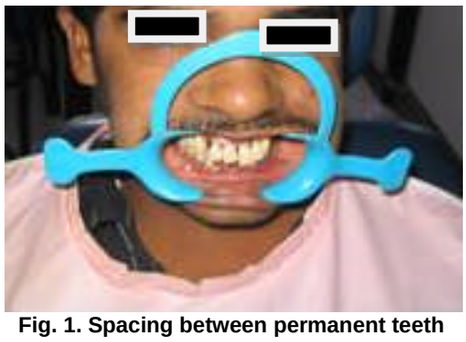

Fig. 1. Spacing between permanent teeth

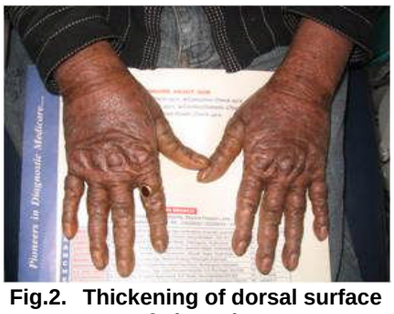

Fig.2. Thickening of dorsal surface of the palm

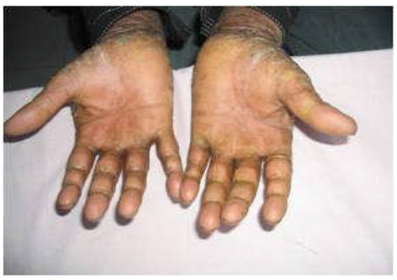

Fig.3. Thickening of Ventral surface of the palm

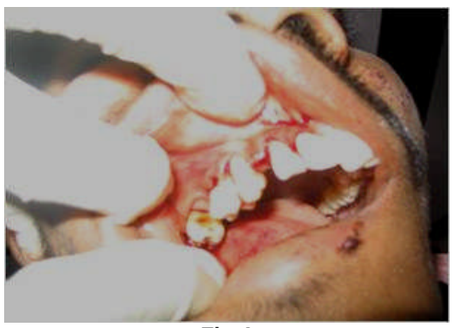

Fig.4

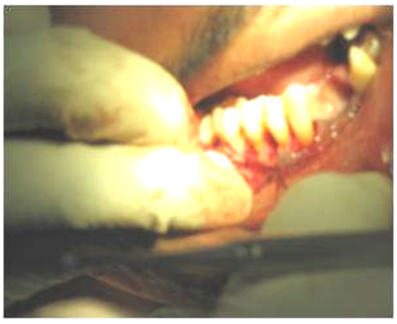

Fig.5

Fig.4. and Fig.5. papillary prservation flap with bone grafting

on early diagnosis so that esthetics and function of the dentition can be protected. Early antibiotic therapy and surgical intervention help in maintaining an intact dentition for longer period of time.

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