

# PAPILLON LEFÈVRE SYNDROME: A CASE REPORT WITH A FAMILIAL LINK

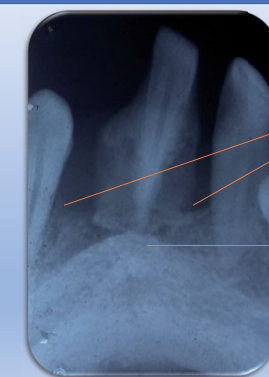
**Papillon Lefèvre Syndrome** : a very rare syndrome of autosomal recessive inheritance.

- Described by 2 French physicians, Papillon and Lefèvre (1942)
- Prevalence : 1-4 / million people in the population
- Etiology : defects in neutrophil function and multiple immune-related mechanisms.
- Genetic studies : loss of function of Cathepsin C gene (chromosome 11q14-21)

✓ **DERMATOLOGIC FEATURE:** Diffuse palmoplantar keratosis.



## RADIOGRAPHIC FEATURES:



- Missing teeth evident: 31,32, & 42
- Complete bone loss irt 41 supported only by a mass of calculus

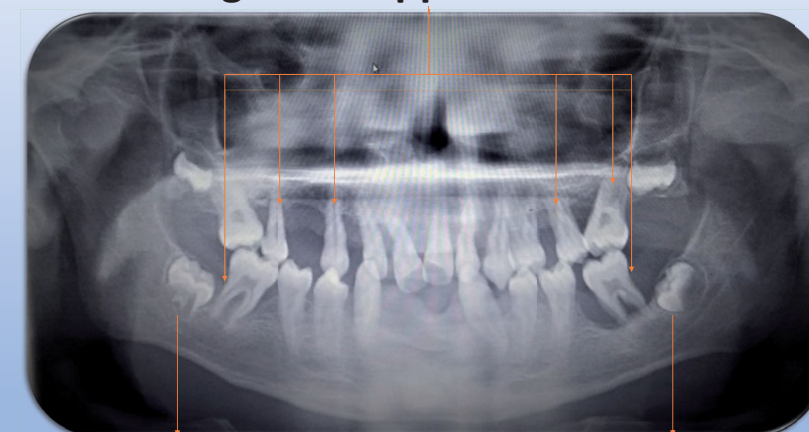
A case of a 14-year-old boy who complained of loose teeth & discomfort in chewing along with recurrently swollen, friable, & receding gums.

**PAST MEDICAL HISTORY:** Patient's parents marriage was non-consanguineous. Elder sibling was also diagnosed with the same disorder and treated previously

**PAST DENTAL HISTORY:** History of early exfoliation of primary and some permanent teeth due to mobility.



✓ Extensive horizontal and vertical bone loss irt 15,13, 25,26, 36, & 46 giving a floating tooth appearance.



✓ Third molar buds present in their bony crypts with normal crown development.

## CONCLUSION:

- Papillon Lefèvre Syndrome can adversely affect the psychological, social, and aesthetic well-being of the patient at an early age
- Devastating disease process associated with cutaneous and oral involvement resulting in partial or complete edentulism.
- Management: multidisciplinary approach: Dentist, Dermatologist, Paediatrician and Psychological Counselling
- Oral retinoids beneficial for both cutaneous and oral lesions along with appropriate periodontal therapy and antibiotics.

## CLINICAL FEATURES:

- ✓ **ORAL:** Advanced periodontitis.
- ✓ Extensive hyperplastic gingivitis.
- ✓ Pathological migration of teeth.
- ✓ Painful mastication.
- ✓ Normal alveolar mucosa in edentulous areas.

**Clinical Differential Diagnosis:** Pre-pubertal periodontitis & Haim-Munk Syndrome (associated with palmoplantar keratosis, severe aggressive periodontitis, arachnodactyly, acro-osteolysis, and onychogryphosis.)

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