Papillon-Lefevre Syndrome: Periodontist's Perspective

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Abstract:

Papillon- Lefevre Syndrome (PLS) is a rare autosomal recessive disorder characterized by palmoplantar hyperkeratosis and severe periodontitis affecting both primary and permanent dentitions. This condition is caused by mutations in the Cathepsin C gene on chromosome 11q14-q21, leading to impaired neutrophil function and increased susceptibility to periodontal destruction. This case report describes a 13-year-old female who present with bleeding gums, mobile teeth, and hyperkeratotic lesions on her palms and soles. Clinical and radiographic examinations revealed advanced periodontal boneless around multiple teeth. The patient underwent comprehensive periodontal therapy, including scaling and root planning, followed by regenerative procedures using Osseo grafts and systemic antibiotic therapy. Dermatological management with oral retinoids was also initiated to address cutaneous manifestations. Follow-up evaluations demonstrated significant improvement in gingival health, reduced probing depths, and stabilization of tooth mobility. This case highlights the importance of early diagnosis, a multidisciplinary management approach involving periodontists and dermatologists, and strict maintenance therapy to preserve oral function and improve overall prognosis in PLS patients. Multidisciplinary care and patient education remain critical for long-term outcomes.

Keywords:

Papillon- Lefevre Syndrome, periodontitis, palmoplantar hyperkeratosis, Cathepsin C, regenerative periodontal therapy, multidisciplinary management.