Papillon Lefevre Syndrome

Le syndrome de Papillon-Lefèvre

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An 9 year-old boy, reported to the Department of Pediatric dentistry, in the center of consultations and dental treatments of Rabat, with complaints of loose teeth and discomfort in chewing along with recurrently swollen and friable gums.

Dental history of our patient included premature shedding of his deciduous teeth. Primary incisors were affected first and display marked mobility by the age of three years. By the age of four or five years, all the primary teeth have been exfoliated. Permanent teeth have shown also an early exfoliation (first upper permanent molars and permanent right maxillary central incisor) (fig. a)

The remainder of his past medical history was unremarkable. The family history revealed consanguineous marriage of the parents. The parents and other family members were not affected. Pregnancy and delivery were normal.

The patient also complained of persistent thickening, flaking and scaling of the skin of palms (fig. b), soles, heels and sides of the feet (fig. c). The patient's knees (fig.d) and elbows (fig. e) were also affected, but to a lesser degree than the palmoplantar surfaces.

A panoramic X-ray (fig. f) was made to have an overview of the dental and periodontal structures and a retroalveolar assessment was done. Both the radiographic images

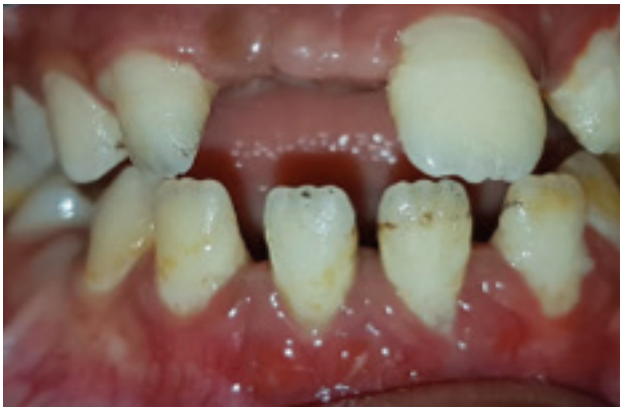

Figure a : Intraoral examination showing missing upper right permanent central incisor, severe gingival inflammation and deep periodontal pockets were noticed. ,heavy deposits of plaque, calculus and halitosis, were also present.

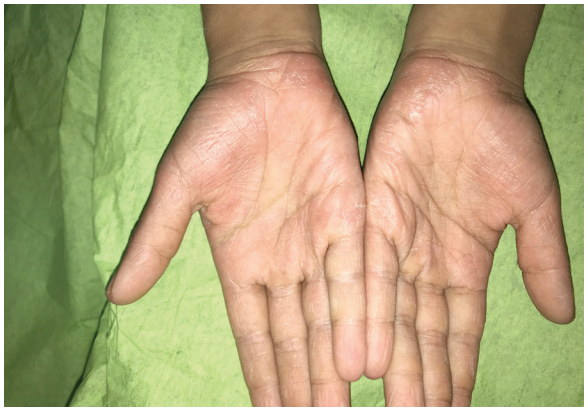

Figure b : keratotic, confluent plaques affecting the skin of palmar surfaces of hands;

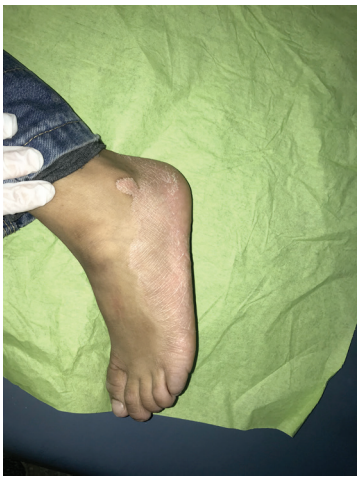

Figure c : Keratotic plaques affecting the dorsal surface of feet

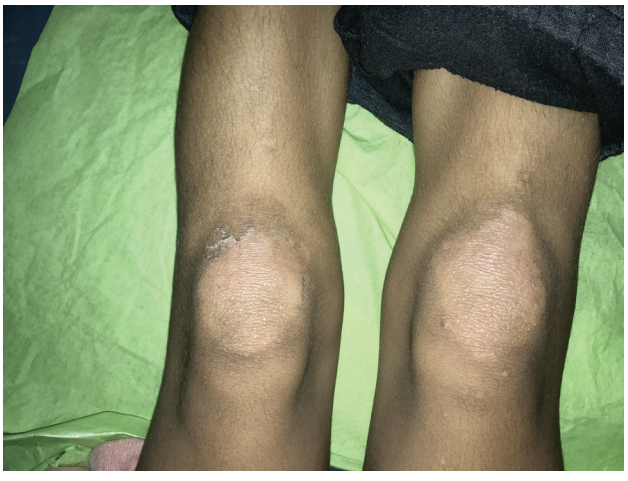

Figure d : well-circumscribed, Keratotic plaques on the knees bilaterally are also noticed.

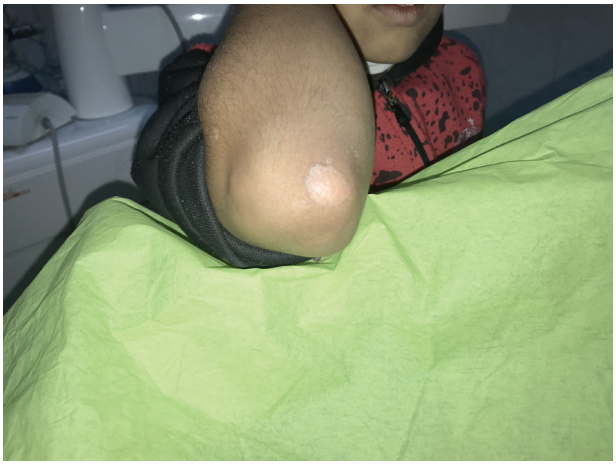

Figure e : keratotic plaques on the elbow

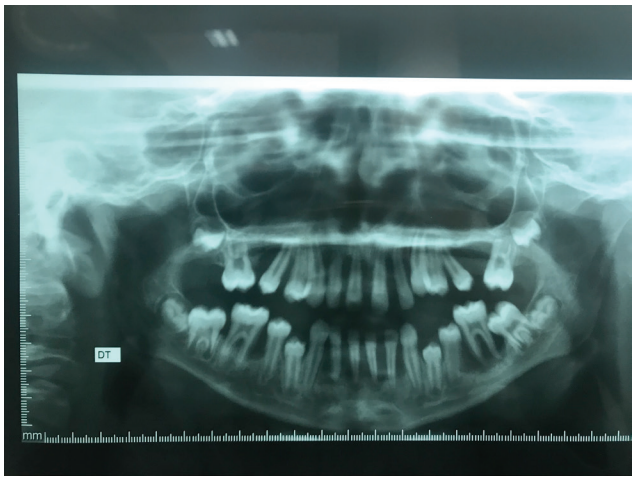

Figure f : f. OPG showing severe generalized destruction of alveolar bone The mandibular left 1st molar was entirely out of its socket .The upper right permanent central incisor has been exfoliated before the scheduled appointment for endobuccal photography.

revealed combination of generalized horizontal bone loss with localized vertical defects in all permanent teeth. the inter-radicular bone underneath the roof of furcation in the mandibular first permanent molars is completely destroyed. The gingival tissue is also receded apically so that the furcation opening is clinically visible.

Based on the patient's history and clinical and radiographic examinations, a diagnosis of Papillon-Lefèvre syndrome (PLS) was made.

The differential diagnosis of this case was made with two rare disorders that are allelic variants of PLS, Haim-Munk syndrome and prepubertal/aggressive periodontitis.

No definitive treatment is available for prevention and management of periodontal destruction, however strict oral hygiene maintenance, scaling and root planning along with suitable antibiotic regimen have been made, but in later stages, as with disease progression, all the teeth will be usually lost.

A follow-up will be instituted for oral rehabilitation ; which includes partial or complete denture prosthetic replacement (according to number of remaining teeth). Osseointegrated implants are an option and can have a great impact psychosocially by restoring esthetics and function.