

PAPILLON-LEFÉVRE SYNDROME (A case report)*

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SUMMARY

Papillon-Lefèvre syndrome refers to palmar-plantar hyperkeratosis associated with severe periodontopathy. A 13-and-one-half-year-old girl presented to our clinic with complaints of teeth mobility and exfoliation with pain on chewing. In physical examination she had a bilateral and symmetrical hyperkeratosis of the face, palms, soles, elbows, knuckles and knees. Severe gingivitis and periodontitis, plaque, spontaneous hemorrhage, exudate and suppuration were observed. In this article a case with Papillon-Lefèvre Syndrome was described and compared with similar cases reported in the world literature. Key words: Papillon-Lefèvre syndrome, gingivitis, periodontitis.

(Figs. 1, 2 and 3). The skin of the palms and soles became red, thickened, and scaly. The skin lesions on the knees were well-demarcated plaques or patches, while the lesions on the palms were of a diffused pattern. In some involved areas the keratotic lesions were slightly raised above the surrounding skin. Urinalysis, complete blood count, differential blood count, serum calcium, phosphate and alkaline phosphatase values were normal. Computerized tomography disclosed no evidence of ectopic intracranial calcification of the falx cerebri and choroid plexus. Her mental abilities and intelligence were normal.

The Papillon-Lefèvre syndrome (PLS) is manifested by redness and diffuse sometimes localized hyperkeratosis of the palms and soles, which usually extends to the dorsal aspects of the hands and feet. Gingivitis and periosteal changes of the alveolar bone (juvenile periodontitis) result in loss of both deciduous and permanent teeth.¹

A 13-and-one-half-year-old girl with Papillon-Lefèvre Syndrome was described and compared with similar cases reported in the world literature.

CASE REPORT

B.A., a 13-and-one-half-year-old girl presented to Atatürk University, Dentistry Faculty with complaints of teeth mobility and exfoliation with pain on chewing. Her mother stated that condition was first noticed when her primary teeth exfoliated at 5 years of age and that oral manifestations appeared almost simultaneously with the onset of palmar-plantar hyperkeratosis. There was no history of other serious illness or recurrent infection. The patient's health history was unremarkable. There was no familial history of skin rash and premature loss of teeth due to periodontitis.

Physical examination was unremarkable except for the integument which revealed a bilateral and symmetrical hyperkeratosis of the face, palms, soles, elbows, knuckles and knees



Figure 1: Appearance of her face.

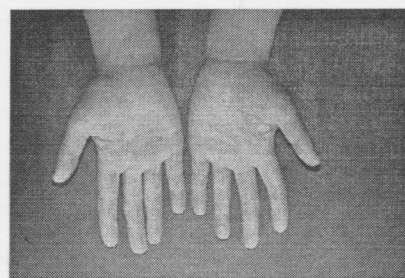


Figure 2: Palms of hand showing diffused hyperkeratotic erythematous lesions.

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The oral mucosa was within normal limits except for severe erythematous and edematous gingiva. Severe gingivitis and periodontitis, plaque, spontaneous hemorrhage, exudate and suppuration were observed. Abscess on the facial of 16, 15, 24, 26, 33, 43, 46. The gingiva was bright red, severely inflamed, swollen, and tender. Pus exuded from the periodontal pockets and gingival abscesses upon slightest pressure. The teeth were extremely mobile and gingival recession was evident (Figs. 4a, 4b, 4c). The oral mucosa, including that covering the edentulous area, was normal in color and consistency. The submaxillary lymph nodes were not palpable. There was strong oral malodor. Eleven of these teeth had mobility scores (Mühleman 1960) of +3.²

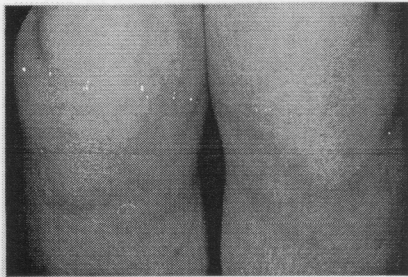
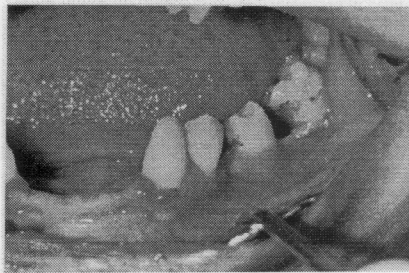
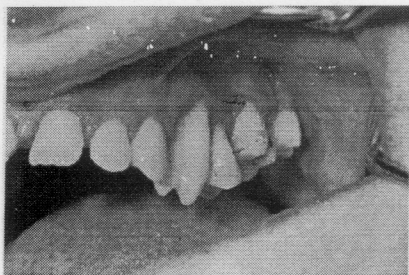


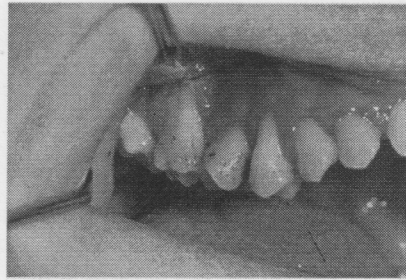
Figure 3: Knees showing well-demarcated erythematous scaly hyperkeratotic lesion.



4a



4b



4c

Figure 4a, 4b, and 4c: Intraoral photographs revealed

Radiographic examination (panoramic and periapical views) revealed that the alveolar bone surrounding the mobile teeth lacked definable lamina dura and had widened periodontal membrane spaces (Figs. 5, 6).

Subgingival plaque was removed from mesial sites on teeth 16, 15, 24, 26, 33, 43, and 46 by means of one sterile paper point per site. Subgingival plaque samples, plated aerobically and anaerobically on blood agar, revealed predominance of *Actinobacillus actinomycetemcomitans*, *Capnocytophaga*, *F. nucleatum* and *E. corrodens* species.

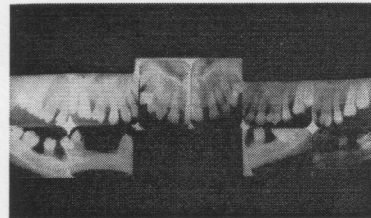


Figure 5: Panoramic radiograph revealing generalized severe alveolar bone loss with no evidence of root resorption.

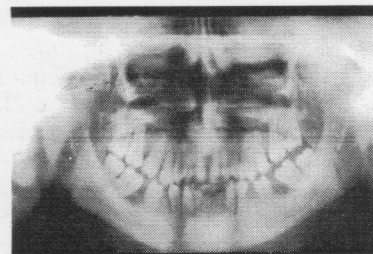


Figure 6: Periapical radiograph revealing generalized severe alveolar bone loss and widened periodontal membrane spaces.

DISCUSSION

Papillon-Lefèvre syndrome, first described in 1924, refers to palmar-plantar hyperkeratosis associated with severe periodontopathy.³ The syndrome appears in childhood and adolescence; however, a late onset variation of the disease has been reported.^{4,5} PLS is characterized by diffuse or localized hyperkeratosis of the palms and soles and extensive loss of periodontal attachment accompanied by generalized, severe, and rapid destruction of the alveolar bone around the primary and permanent teeth, causing premature loss of both dentitions.¹ Although the etiology is unknown, the disease is considered to be transmitted as an gene frequency of 0.001:⁶ A prevalence of PLS in general population of 1 to 4 per million. Males and females are equally effected.¹

The severity of keratoderma is variable, with the soles being prominently more involved. The margins of the keratoderma are well defined and erythematic, frequently extending to the thenar and hypothenar eminences of the palms. Other sites of involvement include the dorsal surface of the fingers and toes, elbows, legs, thighs, and, rarely the trunk.^{1,7} Skin lesions may vary in color, texture, and manifestation. The lesions appear as white, light yellow, brown or red plaques and patches.⁷

The primary teeth erupt at the expected age. The deciduous teeth are lost prematurely in most cases. The permanent teeth are always periodontally involved.⁸ The pathognomic dental features of PLS are the looseness, hypermobility, drifting, migration, and exfoliation of teeth without signs of root resorption.

In recent years, three main factors have been suggested to be responsible for the initiation and progression of PLS. First, the presence of virulent gram-negative anaerobic pathogens at the site of the lesion (plaque/periodontal pockets) such as *Bacteroides gingivalis*, *Capnocytophaga*, *spirochetes*, and *A.a.*⁹⁻¹² Of the pathogens, *A.a* constituted more than 50 % of the total colony-forming units. There are also numerous virulence factors such as leukotoxin, collagenase, endotoxin, epitheliotoxin, and fibroblast-inhibiting factor 13 suggested that PLS is bacteriologically mediated and therefore could be treated with antibiotics.¹⁴ Second, impairment of neutrophil chemotactic, phagocytic, and bactericidal activities accompanied by a decrease in the cells migration.^{9,14} Third, defect of immune-mediated mechanism including reduced

lymphocyte response to pathogens, depression of helper-to-suppressor T cells ratio, deficient monocytic function, elevation of serum immunoglobulin to *A.a*, and degenerative changes of plasma cells.¹⁵⁻¹⁷ The elevated antibody titers to *A.a* support the microbial findings of the involvement of this bacteria in the periodontopathy associated with PLS.

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