

PAPILLON- LEFÈVRE SYNDROME

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Seven cases of Papillon- Lefèvre syndrome from 6 families were studied. Their ages ranged from 3 to 25 years. Falling of deciduous teeth was noticed between 2 1/2 to 7 years. Five patients developed palmoplantar keratoderma at 3 months of age. Two patients showed punctate lesions also. Only 1 patient was born to consanguinous parents.

Key Word: Papillon Lefèvre syndrome

Introduction

Papillon and Lefèvre described the association of palmoplantar keratoderma (PPK) with periodontosis in 1924.¹ Other features of the syndrome are recurrent pyogenic skin infections and calcification of falx cerebri.²⁻⁴ The dental features include frequent periodontal breakdown in both deciduous and permanent dentitions. Loosening of tooth and shedding occurs due to loss of alveolar bone. Gingiva becomes inflamed and bleed easily. Deciduous teeth are lost by the age of 5 years and permanent teeth by 15 years. More than 120 cases have been reported in the world literature upto 1988,^{5, 6} of which about 10 are from our country. PPK may be punctate, striate, or diffuse. This starts between first and fourth years of life.⁷ Keratoderma involves palms, soles, back, heel, external malleolus, tibial tuberosities, elbows, and dorsae of hands and feet. Maximum degree of hyperkeratosis coincides with active periodontal disease.⁶ There may be occasional association with

hypertension, hyperglycemia, systolic murmurs, retardation of skeletal maturation, osteoporosis, and hyperthyroidism.⁸ Etreinate, isotretionin, and acitretin have all been successful in improving the cutaneous eruption, lessening the gingival inflammation, and in saving the teeth in severe cases.⁹

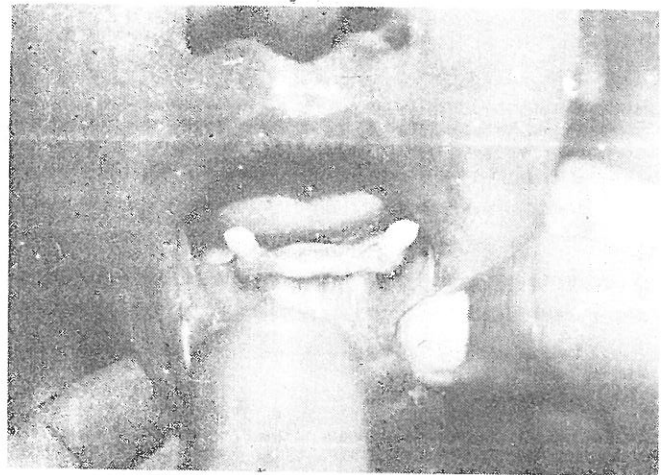


Fig. 1. Loss of teeth
Case Reports

Seven patients with Papillon-Lefevre syndrome were seen. The clinical data of each patient was recorded in a proforma.

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Dental examination was done in 5 patients and dental X-rays were taken wherever feasible.

Among 7 patients, there were 6 males and 1 female (Table I).

teeth which erupted normally, started loosening and falling from 2 1/2 to 7 years of age. One male patient who was seen at the age of 18 years was born to consanguinous parents (Fig.1). All other patients including 2

Table I . Details of the patients

S.No	Age	Sex	PPK (Onset at months)	Falling of teeth at years	Consanguinity	PPK sites	Secondary infection	Dental Formula		Number of affected siblings	Punctate lesions
								ECBA EDCBA	ABCD ABCDE		
1.	3yrs 6mth	M	3	2yrs 6mths	No	Palms and Soles	No	ECBA EDCBA	ABCD ABCDE	No	Yes
2.	9	M	3	6	No	-do-	No	ED ED	DE DE	Yes one (Pt no 3)	Yes
3.	7	M	3	6	No	-do-	No	EC ---	CE BCDE6	Yes one (Pt no 2)	No
4.	8	M	3	4	No	Palms, soles, knees and elbows	No	621 621	126 126	No	No
5.	18	M	12	6	No	Palms and soles	Yes	---		No	No
6.	17	M	3	7	Yes	Palms, soles and elbow	Yes	6 6	6 6	No	No
7.	25	F	6	7	No	Palms and soles	No	-----		No	No

Their ages ranged from 3 to 25 years. Five patients developed rough palmar and plantar skin 3 months after birth, the other 2 at 6 months and 1 year respectively. The

siblings were children of nonconsanguinous parents. Two male patients were affected with recurrent pyoderma on the lower limbs since childhood. Hyperhidrosis was noticed

in 2 patients. None of our patients had any systemic abnormality. Dental examination done in 5 patients revealed less number of teeth than their age, and most of the teeth present were in grade I or grade II mobility. One adult patient seen at the age of 17 years had lost most of his permanent teeth. Two other adult patients who were examined at their residence also lost most of their permanent teeth. Dental X-rays revealed the features of missing teeth and loss of alveolar bone support. Routine and additional investigations like serum calcium, phosphorous, and alkaline phosphatase levels did not reveal any abnormality.

Case no.1 at the age of 3 1/2 years showed premature exfoliation of tooth numbers 54 & 65. The other teeth were in grade I to grade II mobility. In the second patient 4 upper and 4 lower incisors, and 4 first molars should have erupted along with deciduous canine, first and second molars. All these were missing along with permanent incisors, permanent first molars, and all deciduous canines. The other teeth which were present were mobile. The third patient had no teeth in the right mandibular region which were extracted because of grade III mobility. In the fourth patient all deciduous canines and first molars which should have been present were not seen. In the sixth patient a full complement of permanent teeth excluding the third molars should have been present whereas he had only 4 first molars which were also in grade I mobility. One patient showed early loss of tooth by the age of 2 1/2 years which was very early for his age group.

Dental X-rays showed complete resorption of alveolar bone giving the teeth

"floating-in-air" appearance (Fig.2).⁶ Our patients did not show dural calcification or any other rarer associations like hypertension or hyperglycemia.

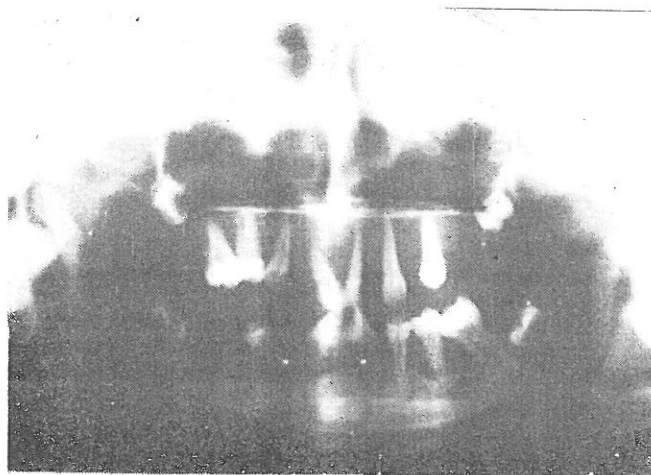


Fig. 2. Dental X-Ray showing "Floating-in-air" appearance.

Comments

PPK has been reported to occur between 1-5 years of life whereas majority of our patients (5 out of 7) developed this at 3 months of age.⁷ The skin lesions of PPK might extend beyond the palms and soles which was seen in 2 of our patients.⁵

Papillon-Lefèvre syndrome is probably inherited as an autosomal recessive disease but only 1 patient in our study (case no.6) was born to consanguineous parents. Two of our patients had history of recurrent pyoderma on the legs since childhood. This phenomenon is usually seen due to disordered leukocyte function or decreased neutrophil phagocytosis.³⁻⁵

The interesting feature noticed was that 6 out of 7 patients belonged to one village. Detailed epidemiological study could not pinpoint any particular reason for this.

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