PAPILLON - LEFÉVRE SYNDROME
(A case report)

F. HANDA,* R. R. AGGARWAL † AND ADARSH CHOPRA ‡

Summary

A case of Papillon - Lefévre syndrome is being reported.

Review of Literature

This syndrome was first described by Papillon-Lefévre in 1924[1]. It is characterised by palmo-plantar hyperkeratosis and premature loss of deciduous and permanent teeth. This is a very rare syndrome and upto 1964 only 40 cases were reported in the world literature. Sardari Lal et al[2] reported the first Indian case. Gorlin et al[3] reported that it results from homozygosity for autosomal recessive genes. The frequency of occurrence is estimated to be roughly 1 to 4 per million persons in general population.

Principal manifestation is hyperkeratosis of palms and soles, usually appearing between 1 to 4 years of age. Simultaneously, periodontal involvement of deciduous dentition occurs. Stein[4], Kohler et al[5], have reported that degree of hyperkeratosis is most severe at the time of periodontal involvement. Skin lesions at younger than usual age have been observed by Carson[6] and Garre[7]. Well demarcated erythematous, scaly and hyperkeratotic plaques of palms and soles are present. Similar plaques may involve skin over the tendo-achillis, lateral malleoli, tibial tuberosities, dorsa of fingers and joints. Bataille et al[8ab] and Papillon - Lefevre[1] have reoported that skin lesions are identical to Maleda disease. However, this syndrome is not considered to be a form of Maleda's disease and is a distinct disease as reported by Bellenger E in 1954[9] and Jansen et al[10]. The degree of hyperkeratosis in skin lesions varies intermittently. Lesions are more scaly and fissured during winter as reported by Stein and Hall[11]. Hyperkeratosis may spontaneously and completely disappear in summer. Greither A in 1959[12] reported spontaneous cure in a patient at 14 years of age. His skin lesions regressed and wisdom teeth erupted normally without any pyorrhoea. Usually, hyperkeratosis is moderate but severe cases with thickened and fissured palms and soles are reported by Stein[4] and Garre[7]. Foetid hyperhidrosis of feet may occur. Hyperkeratotic plaques have been reported on eyelids, cheeks and labial commisures by Hawes[13], on legs and thighs by Ziprokowski[14] and on maxillae by Garre[7].

Usually, nails are normal but thumbnail dystrophy has been observed by Garre[7] and Hawes[13]. Gorlin in 1964 reported mild onychogryphosis. Hairs are normal but may be thin in some cases.

* Professor
† Assistant Professor
‡ Registrar
Department of Skin & V. D., Government Medical College, Patiala (Punjab)
Received for publication on 4—3—1977

Other lesions like eczemas and erythema of face, sacral snd gluteal regions have been reported by Jansen[10], Garre[7] and Ziprokowski[14]. In one family with Papillon — Lefévre syndrome, long pointed claw-like fingers were seen.

Dental changes are very characteristic. Deciduous teeth erupt normally but are lost by the age of 4–5 years due to dystrophic changes in periodontal ligaments and alveolar bones with formation of periodontal pockets of suppuration. The permanent teeth erupt normally but are prematurely shed due to formation of periodontal suppuration. Characteristically, patient becomes edentulous by about the 16th year and only the third molars are preserved as reported by Gorlin et al. After shedding of teeth the gingiva almost returns to normal and can tolerate dentures. In only a few instances as reported by Ingle[15], the loss of the teeth has not been accompanied by severe gingival inflammation.

Calcification of dura in the attachment of tentorium and choroid has been observed by Jansen[10]. Age of onset of calcification is not known but in several children it is present at the time of initial examination.

The physical development of recorded cases was considered normal but Ziprokowski et al[14] in 1963 reported obvious retardation of somatic and sketelal development in their case.

High blood pressure, high blood sugar curve, systolic murmur, elevated haemoglobin level, retardation of skeletal maturation, general osteoporosis, diffuse swelling of thyroid, micro-ophthalmia have all been reported but may have been chance associations. These patients are predisposed to infections and regional lymphadenitis occurs in association with periodontal lesions.

Case History

An 18 years old male patient attended skin OPD with the complaints of : red scaly lesions on palms and soles since the age of 4–5 years and premature loss of temporary and permanent teeth.

At the age of 6 years, he developed scaly erythematous lesions on extensor surfaces of hands. At the age of 8 years, patient developed similar lesions on extensor surfaces of elbow and knee joints. Erythema and scaling were worse during the winter at which time the lesions became thickened and fissured as well. This used to cause pain and burning sensation when hands and feet were immersed in water. During summer, at times, lesions regressed and disappeared for a few days. With appearance of skin lesions, gums used to swell up and within a few days, all deciduous teeth were shed. Patient suffered from pyorrhoea and pain at that time. Permanent teeth started erupting at 6–7 years of age but all teeth were lost prematurely except the three molar teeth which erupted at the age of 12 years. Patient had no associated problems when the permanent teeth were shed. Wisdom teeth had not erupted and patient had worn dentures for 4 years. Hyperhidrosis of whole body, specially of palms and soles was noticed from the age of about 12 years.

Past history was not significant. Family history revealed that an elder sister had erythematous scaly plaques on her body. She had died of jaundice at the age of 16 years.

Systemic examination showed no abnormality.

Local examination revealed diffuse erythematous scaly hyperkeratotic lesions on palms and soles which extended towards the dorsa and stopped abruptly showing well defined margins. Similar

plaques were present over metacarpophalangeal and interphalangeal joints, lateral and medial malleoli, extensor surfaces of elbow and knee joints. Soles had thick scaly lesions with superficial fissures. Hand nails were normal but last two toe nails on both feet showed yellowish discolouration, subungual hyperkeratosis and slight onychogryphosis. All teeth were absent except 3 molars.

RIGHT	LEFT
7	7

Investigations—Routine blood tests, including Hb, WBC T & D, & ESR were normal. Urine examination blood urea, Serum, Ca > P were also normal. X-Ray of teeth showed 3 buds. X-Ray skull was normal. Urine nitrogen including (NH_3-N) was - 0.456 gms per 24 hrs.

Histopathology showed marked hyperkeratosis and no parakeratosis marked acanthosis, a thin corium and marked perivascular inflammatory infiltration.

Discussion

This patient who is a typical case of Papillon — Lefévre syndrome manifested few interesting features which have been observed by earlier workers also. These were as follows :-

At the time of shedding of permanent teeth, this patient showed no gingival swelling or periodontitis. Only few toe nails showed dystrophy. Hyperhidrosis was present all over the body. At the age of 18 years, wisdom teeth had not erupted. Skin lesions and periodontitis began simultaneously. Lesions which were severe in winter used to regress sometimes spontaneously during summer. Physical development was normal.

References

1. Papillon MM, Lefevre P : Deux Cas de keratodermie palmaire et plantaire symetrique familiale (Maladie de Meleda) Chez le frere et la soer. Coexistance dans le deux cas dalterations dentaires graves, Soc Franc Dermat et Syph, 31 : 82, 1924.
2. Sardari Lal, Venkatpathy L, Bhargava I, et al : Papillon Lefevre Syndrome. Case report with cytogenic study, Indian J Derm Vener, 3 : 83, 1971.
3. Gorlin RJ, Sedano H and Anderson VE : The syndrome of palmo-plantar hyperkeratosis and premature periodontal destruction of teeth, J Paediatrics, 65 : 895, 1964.
4. Stein P : Papillon Lefevre Inaugural Dissertation Hautklinik, University of Freiburg Germany, 1960.
5. Kohler JA : Paradentopathie Fugendlicher and Keratoma palmare et plantare Deutsche Zahnarzt, Ztschr, 8 : 885, 1953.
6. Corson EF : Keratosis palmaris et plantaris with dental alterations, Arch Derm Syph, 40 : 639, 1939.
7. Garre A : Periodontitis og tiddlig tap av on temporaere eg permanente tenner hos barn med keratosis palmaris et plantaris To kasus, Norske Tan Tid, 68 : 297, 1958.
8. (a) Bataille R and Durperrat Ms : La dentition dans Les Keratodermies palmo-plantaires congenitales, Soc Franc Dermat et Syph, 59 : 121, 1952.
   (b) Bataille MR : Les parodontoses des keratodermies palmoplantaires La maladie de Meleda, Rev Stomat, 54 : 139, 1953.
9. Bellenger E : Chute precoce des dents temporaires et permanentes (Alveolyse Infantile) et keratose palmoplantair : Un Syndrome recessif, Thesis Paris, 1954.
10. Jansen LH and Dekker G : Hyperkeratosis palmo-plantaris with periodontosis (Papilon Lefevre), Dermatologica, 113 : 207, 1956.
11. Hall RK : Papillon Lefevre Syndrome, Australian Dent J, 8 : 185, 1963.
12. Greither A : Keratosis palmo-plantaris mit periodentopathe (Papillon Lefevre) Dermatologica, 119 : 248, 1959.
13. Hawes RR : Report of 3 patients experiencing Juvenile periodontosis and early loss of teeth, J Dent Child, 27 : 169, 1960.
14. Ziprkowski L, Yochanan R and Brish M : Hyperkeratosis palmo-plantaris with periodontosis (Papillon Lefevre), Arch Derm, 88 : 207, 1963.
15. Ingle JI : Papillon Lefevre Syndrome prococious periodontosis with associated epidermal lesions, J Perio, 30 : 230, 1959.

SYMMETRICAL PROGRESSIVE ERYTHROKERATODERMA
(A case report)

T. R. KAPUR,* M. A. ANSARI † AND O. P. SINGH ‡

Summary

A case of symmetrical progressive erythrokeratoderma in a 32 year old male patient noticed at the age of 27 years, has been described.

Symmetrical progressive erythrokeratoderma is a rare condition characterised by asymptomatic, hyperkeratotic lesions with an erythematous base, distributed bilaterally and symmetrically on the dorsal aspects of hands, forearms, feet and legs. In extensive cases the lesions may also be present on the upper arm, shoulders, neck and face[1]. The lesions usually start appearing during infancy but may be delayed until adult life[1]. It is suggested that the condition is transmitted as an autosomal dominant character[2]. To the best of our knowledge, no such case has been reported from India.

Case Report

A 32 years old man noticed asymptomatic, well demarcated, erythematous and hyperkeratotic plaques on the dorsal aspects of both hands 5 years earlier. These gradually extend to involve the extensor aspects of forearms and the skin over the dorsal aspects of proximal phalanges. The lesions were bilaterally symmetrical. (Fig. page No. 170) Other members of the family upto 3 generations did not show any evidence of similar disease. Other parts of the body including the palms, soles and nails were not involved. Repeated scrapings for fungus and the Auspitz sign were negative. The patient was prescribed topical corticosteroid with salicylic acid ointment, but there was little improvement over a period of six months.

Discussion

Asymptomatic hyperkeratotic plaques on an erythematous background, distributed bilaterally, limited to the dorsal aspects of hands and forearms are quite suggestive of symmetrical progressive erythrokeratoderma, even though the disease started late in life and there is no evidence of familial transmission. Sparing of the palms and soles excludes the recessive form of palmo-plantar keratoderma ; Mal de Malda[1]. It is also unlikely to be symptomatic erythrokeratoderma which may occur at the site of chronic inflammatory changes or repeated trauma in subjects who do not produce the more common lichenifying response to such

* Classified Specialist (Derm and Ven)
Command Hospital (W.C.)
Chandigarh
† Medical Officer
W. B. Workshop (GREF), Pathankot
‡ Lecturer (Derm and Ven)
All India Institute of Medical Sciences
New Delhi-16

Received for publication on 2—5—1977