Pseudoherpetic keratitis

Corneal changes in circumscribed palmo-plantar keratoderma

RODNEY WESTMORE AND FRANK A. BILLSON

Division of Ophthalmology, Royal Children's Hospital, Melbourne, Australia

The association of herpetoid corneal changes with circumscribed palmo-plantar keratoderma was first described by Richner (1938). Hanhart (1947) suggested that this represented a recessively inherited ectodermal syndrome, and the condition is sometimes called the Richner-Hanhart syndrome. Subsequent reports have been relatively few: Cremona (1957); Kuske (1959); Franceschetti and Thier (1961); Costa (1962); Grayson (1965). In this case the nature of the "herpetic" corneal lesions was not appreciated until after the appearance of typical skin lesions.

Case report A baby girl, the only child of Italian parents, was first seen in the Eye Clinic at the Royal Children Hospital, at the age of 3 years. The mother stated that, since the age of 3 months, the child had been troubled by recurrent episodes of irritable eyes, characterized by photophobia, lacrimation, and severe pain. These episodes had been treated by idoxuridine and iodization with epithelial debridement on two occasions during the first year of life. At the age of 15 months circumscribed plaques of thickened skin surrounded by narrow margins of erythema appeared on the palmar surface of the hands and, at the age of 24 months, similar changes appeared on the plantar surface of the feet. The affected areas of skin tended to thicken over several weeks and then fall off, after which the process began afresh. During the acute episodes the skin of the palms and soles was noted to sweat more and the feet were tender to walk on.

The exacerbation of eye discomfort paralleled that of the skin lesions and was most distressing in the latter part of the day. The child was otherwise well and appeared to be of normal intelligence. There was no history of past serious illness and no family history of significant ocular or skin disease or of consanguinity in the parents.

Examination

The visual acuity was 6/24 in each eye with -6 D sph. Both corneae showed raised axial epithelial dendritic figures (n a background of hazy epithelial nebulae (Fig. 1).

The lesions showed minimal staining with fluorescein during quiescent periods. At the height of the corneal exacerbations, when the eyes were most painful, there was a breach in the corneal epithelial surface which stained with fluorescein. There was no associated corneal vascularization and corneal sensation was normal. The eyes were otherwise normal apart from mild myopic changes in the fundi. Conjunctival swabs taken during exacerbations were negative for herpes simplex virus on four separate occasions.

The palmar and plantar skin showed the typical lesions of circumscribed palmo-plantar keratoderma (Figs 2 and 3) and skin biopsy further confirmed this diagnosis.



FIG. I Corneal appearances



FIG. 2 Keratoderma on the palms of the handsFIG. 3 Keratoderma on the soles of the feet



(3)

Treatment

An attempt was made to alter the corneal environment with moist chamber glasses, but this had to be discarded because of fogging of the lenses. Methylcellulose drops 0.5 per cent. were found to be the most useful therapeutic agent though relief was incomplete.

Discussion

Herpetoid corneal lesions were regarded by Franceschetti (1968) as pathognomonic of the autosomal recessive form of circumscribed palmo-plantar keratoderma. The histopathological abnormality in the cornea has been revealed by biopsy (Franceschetti and Thier, 1961; Grayson, 1965). The changes are confined to the epithelium and basement membrane. Pseudopod-like extensions of the basement membrane occur between the basal cells of the epithelium and periodic acid-Schiff staining bodies are seen among epithelial cells, some of which show degenerative changes. Bowman's membrane is normal and there is no hyperkeratosis of the epithelium. In our case corneal biopsy was not performed because the appearance of the corneal lesions and typical skin lesions allowed a definite diagnosis. Difficulty in diagnosis may be experienced when corneal lesions are the sole presentation. The condition is likely to be recognized, however, if it is considered in all cases of bilateral axial herpetic keratitis resistant to treatment for herpes simplex and particularly if virus cultures are negative. The corneal lesions appear to be unresponsive to a variety of treatments. Idoxuridine, steroids, epithelial debridement, and iodization appeared to exert little effect on the natural history of the corneal changes in our case. Corneal grafting was not attempted, although it has been reported by Franceschetti (1968) as a form of treatment. It is of interest that the ectodermal skin disturbance has been treated by deep excision and split-skin grafting with some success by Wyn-Williams (1953) and Dencer (1953).

With regard to the genetic implications of this syndrome, the reported families have mostly been Italian or Swiss, and it is of interest that both the parents and all the grandparents in our case were born in northern Italy.

Summary

A case of circumscribed palmo-plantar keratoderma (Richner-Hanhart syndrome) presenting as bilateral pseudoherpetic keratitis is described and its management is discussed. Attention is drawn to the problem of diagnosis where corneal lesions antedate those of the skin. It is suggested that the diagnosis should be entertained in resistant cases of bilateral axial dendritic keratitis in children.

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