*British Journal of Dermatology* 1995; 132: 150–171.

# Correspondence

## An unusual case of porokeratosis involving the natal cleft: porokeratosis ptychotropica?

SIR, Porokeratosis is considered to be a disorder in which mutant clones of cells are present in the epidermis.[1] Under the influence of external factors, such as immunosuppression, irradiation and repeated trauma, the dermatosis may become manifest.[2] The presence of abnormal DNA-ploidy has been demonstrated flow cytometrically in the epidermis of several types of porokeratosis, supporting Reed's theory of an expanding clone of mutant keratinocytes, and in keeping with the premalignant nature of porokeratosis.[3] Clinically, six variants are distinguished: porokeratosis of Mibelli, linear porokeratosis, giant porokeratosis, disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis (DSAP), and palmoplantar porokeratosis.[4] An autosomal dominant mode of inheritance has been described for porokeratosis of Mibelli, DSAP and palmoplantar porokeratosis.[4,5] Linear porokeratosis probably represents mosaicism of the responsible gene. The association of linear porokeratosis with DSAP, either in the same patient or in the same family, has tentatively been explained by the genetic mechanism of loss of heterozygosity, indicating somatic crossing-over involving the gene-locus of an individual heterozygous for DSAP, early in embryogenesis, giving rise to a homozygous cell representing a precursor cell of a clone growing out in a linear pattern during embryonic development of the skin.[6] Variable gene expression has been suggested to explain why different members of one family express clinically distinct porokeratotic variants.[7] We report a new and clinically distinct manifestation of porokeratosis, confined to the natal cleft. We propose the designation 'porokeratosis ptychotropica' for this new form of porokeratosis, which, to our knowledge, has not been described previously. The term 'ptychotropica' is derived from the Greek words *ptyché* (fold)

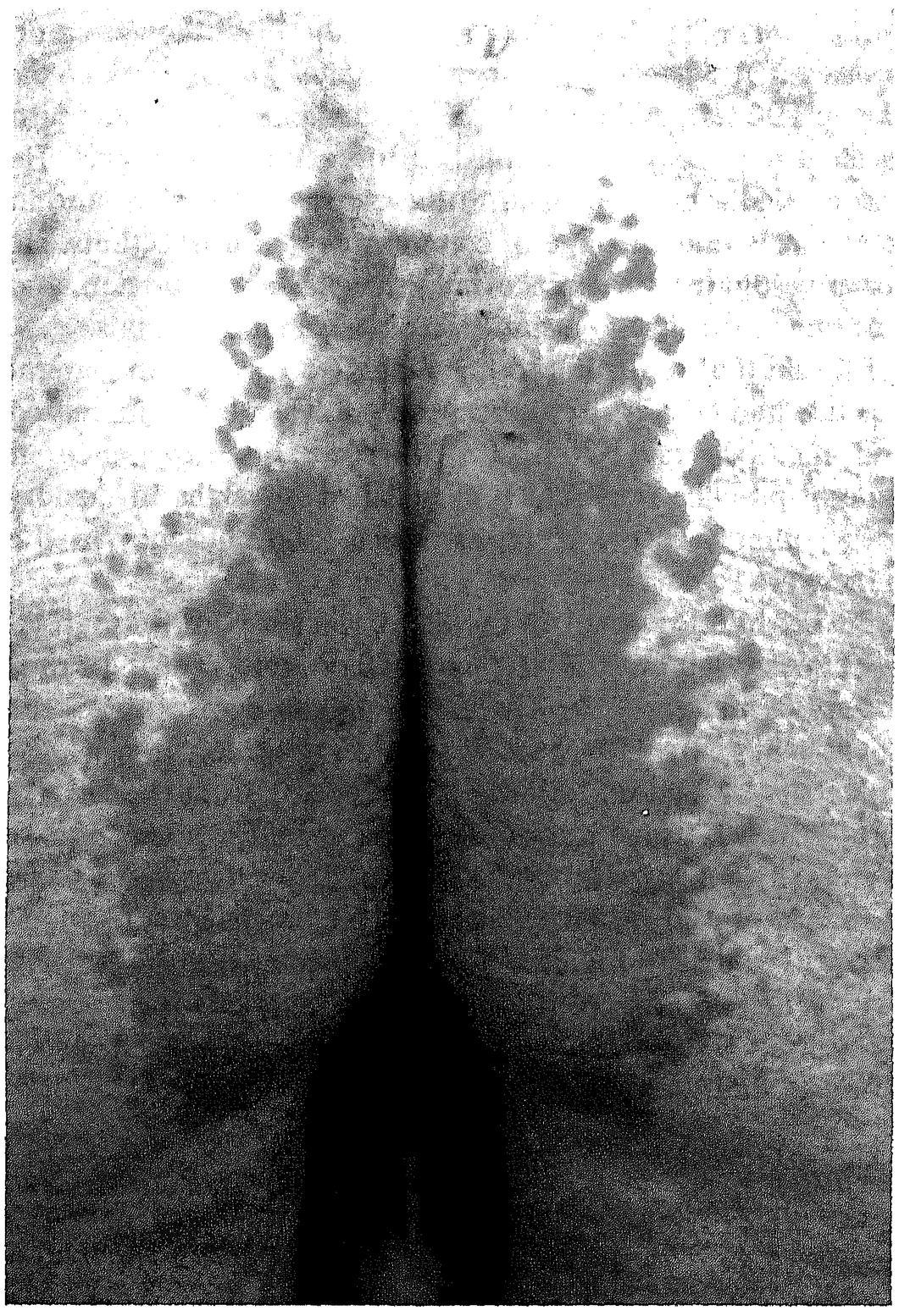

**Figure 1.** Erythematous, keratotic papules in the natal cleft. The lesions are confluent towards the centre of the affected area, forming a plaque.

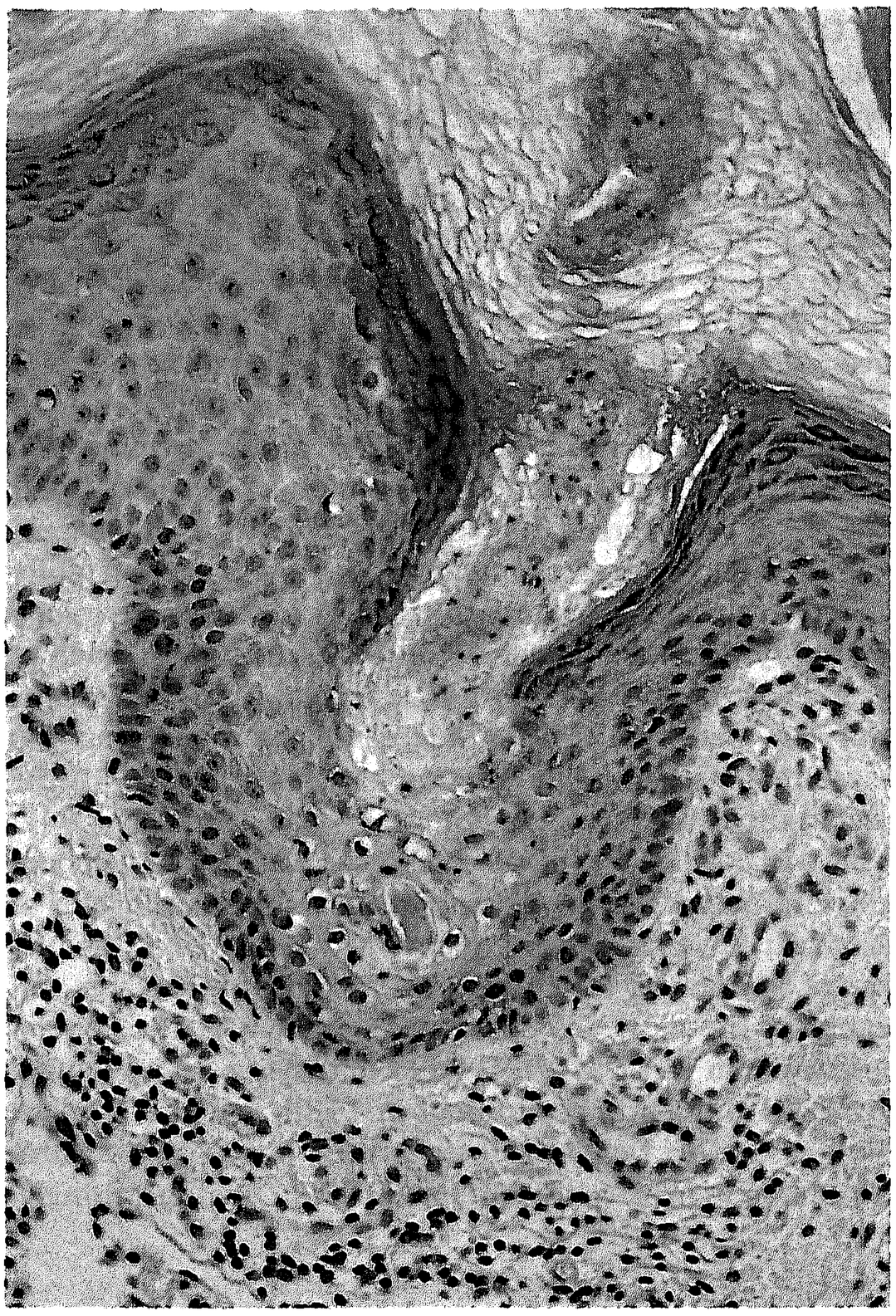

**Figure 2.** Biopsy specimen from the perianal area, showing an invagination of the epidermis, focal loss of the granular layer, mild dyskeratosis, and an overlying cornoid lamella. A mononuclear cell infiltrate is present in the upper dermis (haematoxylin and eosin, ×100).

and *tropé* (a turning), and has been used previously to distinguish the ichthyosiform naevus in the CHILD syndrome, which has a pronounced affinity for the body folds, from all other naevi or naevoid conditions which are not ptychotropic.[8]

A 34-year-old man presented with a pruritic dermatosis confined to the natal cleft. The disease had been present for 9 years, and had gradually become more extensive. Previous treatment had consisted of topical steroids, which relieved the pruritus but did not otherwise influence the lesions. There was no family history of similar skin lesions, or of porokeratosis. On examination, there were multiple erythematous, keratotic papules, which were discrete at the margins of the affected area, but coalesced towards the centre to form a plaque (Fig. 1). The rest of the skin was normal.

Histology of a skin biopsy showed orthohyperkeratosis, acanthosis, and papillomatosis of the epidermis, and typical cornoid lamellae. Beneath the cornoid lamellae, the granular cell layer was absent, and there were numerous dyskeratotic cells. There was a non-specific mononuclear cell infiltrate in the upper dermis (Fig. 2). Focally these lesions were found to extend into the hair follicles.

He was treated with topical all-*trans*-retinoic acid 0·05%, which was applied thinly and evenly once daily. Despite some slight improvement after 1 month, the treatment was eventually discontinued because of unacceptable irritation of the skin.

The characteristic histological feature of all porokeratotic variants is the cornoid lamella. Clinically, most types of porokeratosis consist of lesions characterized by central atrophy and a hyperkeratotic margin. Hyperkeratotic lesions with a centrally keratotic or even verrucous surface may be found in linear prokeratosis and porokeratosis of Mibelli.[9] Porokeratotic lesions usually remain asymptomatic, although intense pruritus has been reported to occur in disseminated superficial porokeratosis.[10]

The morphology and localization of the lesions in our patient differ from the known clinical variants of porokeratosis, and we consider these features may represent a distinctive type of porokeratosis involving the body folds. We propose the designation 'porokeratosis ptychotropica' to distinguish this porokeratotic variant.

*Department of Dermatology,*
*University Hospital Nijmegen,*
*PO Box 9191,*
*6500 HB Nijmegen, the Netherlands*
G.P.H.Lucker
R.Happle*
P.M.Steijlen
**Department of Dermatology,*
*University Hospital Marburg,*
*Deutschhausstr. 9,*
*35033 Marburg, Germany*

## References

1 Reed RJ, Leone P. Porokeratosis—a mutant clonal keratosis of the epidermis. *Arch Dermatol* 1970; 101; 340–7.
2 Coskey RJ, Mehregan A. Bowen disease associated with porokeratosis of Mibelli. *Arch Dermatol* 1975; 111: 1480–1.
3 Beers B, Jaszcz W, Sheetz K *et al.* Porokeratosis palmaris et plantaris disseminata: report of a case with abnormal DNA ploidy in lesional epidermis. *Arch Dermatol* 1992; 128: 236–9.
4 Griffiths WAD, Leigh IM, Marks R. Disorders of keratinization. In: *Textbook of Dermatology* (Champion RH, Burton JL, Ebling FJG, eds). Oxford: Blackwell Scientific Publications, 1992; 1325–90.
5 McKusick VA, Francomano CA, Antonarakis SE. *Mendelian Inheritance in Man.* Baltimore: Johns Hopkins University Press, 1992; 902–3.
6 Happle R. Somatic recombination may explain linear porokeratosis associated with disseminated superficial actinic porokeratosis. *Am J Med Genet* 1991; 39: 237.
7 Moreland ME, Wyre HW. Porokeratosis: two morphological forms within a family. *Arch Dermatol* 1981; 117: 245–6.
8 Happle R. Ptychotropism as a cutaneous feature of the CHILD syndrome. *J Am Acad Dermatol* 1990 23: 763–6.
9 Marghescu S, Anton-Lamprecht I, Melz-Rothfuss B. Disseminated bilateral hyperkeratotic variant of porokeratosis Mibelli. *Arch Dermatol Res* 1987; 279 (Suppl.): 38–47.
10 Kanzaki T, Miwa N, Kobayashi T, Ogawa S. Eruptive pruritic papular porokeratosis. *J Dermatol* 1992; 19: 109–12.

## More than 1000 J/cm$^2$ of UVA for PUVA treatment of psoriasis

Sir, The guidelines for management of patients with psoriasis produced by the Workshop of the Research Unit of the Royal College of Physicians of London, the Department of Dermatology, University of Glasgow and the British Association of Dermatologists[1] is a useful document. In the section on PUVA therapy it is stated that the total lifetime exposure to UVA should (if possible) be a maximum of 1000 J/cm$^2$. We were aware that we had many patients who had exceeded this limit and therefore felt it an important subject for audit.

We have identified 37 patients with psoriasis who have received over 1000 J/cm$^2$ of UVA since PUVA was started in Devon in 1980 (Table 1). The population served is about 1·1 million, and the total number of psoriasis patients treated with PUVA to date has been approximately 300.

Our policy is to use a clearing regimen of PUVA, and then stop therapy, or sometimes to continue for 2–3 months before stopping. Most patients therefore have repeated clearing courses. The majority of high-dose patients are those who relapse promptly if PUVA is stopped, so many are on virtually continuous therapy. All patients in this group have severe psoriasis, and many have had other systemic treatments for their psoriasis (Table 2).

The main reason for continuing PUVA has been patient preference, although we periodically review all psoriasis patients on systemic treatment, and discuss alternative therapy with them. Squamous cell carcinoma has occurred in seven patients, to date, in this group of 37 high-dose UVA patients (Table 1). Most patients who receive PUVA in our area have failed to clear with dithranol and UVB or, having cleared, flared too quickly to make it a practical treatment (this study presents our findings prior to the introduction of calcipotriol; Dovonex®). All systemic agents for the treatment of psoriasis have side-effects, and it is often considered that