Linear Porokeratosis

Dr. K. Y. Chow

Date:	14 July, 1999
Venue:	Yaumatei Skin Centre
Organizer:	Social Hygiene Service, DH; Clinico-pathological Seminar

CASE SUMMARY

History

A 19-year-old man presented with itchy rash on his right leg. The lesions started from the right groin when he was 13, and over the years slowly extended down the medial aspect of his thigh and leg to the medial malleolus of his right ankle. The lesions were pruritic. The was no similar lesion elsewhere, and none of his relatives was affected.

Physical Examination

The lesions extended from the right groin down the medial aspect of the right leg to the right medial malleolus in a linear fashion (Figure 1). It appeared as dark brownish macules on the right groin (Figure 2), the centre of which appeared atrophic. The edge was slightly raised. The lesions from the knee downwards were hyperkeratotic and excoriated. There were also striae seen around the lesions which could be attributed to prior application of potent topical steroid.

Differential Diagnosis

The differential diagnoses included linear epidermal naevus, lichen striatus and linear porokeratosis.

Investigations

An incisional biopsy was performed at the edge of one lesion on his right thigh. It demonstrated the presence of cornoid lamellation with peri-adnexal lymphocytic infiltration around a hair follicle.

Diagnosis

The diagnosis of linear porokeratosis was made.

Progress

Potent topical steroid and keratolytic agents had been tried but produced no symptomatic relief or reduction of thickness of the keratotic papules. The patient could not tolerate topical 0.025% tretinoin cream

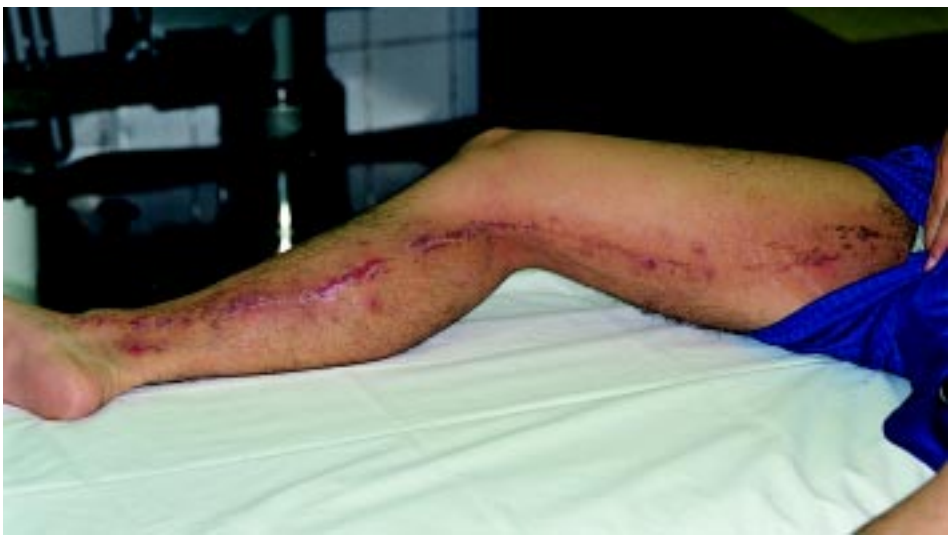

Figure 1: Linear porokeratosis affecting the medial aspect of the right lower limb

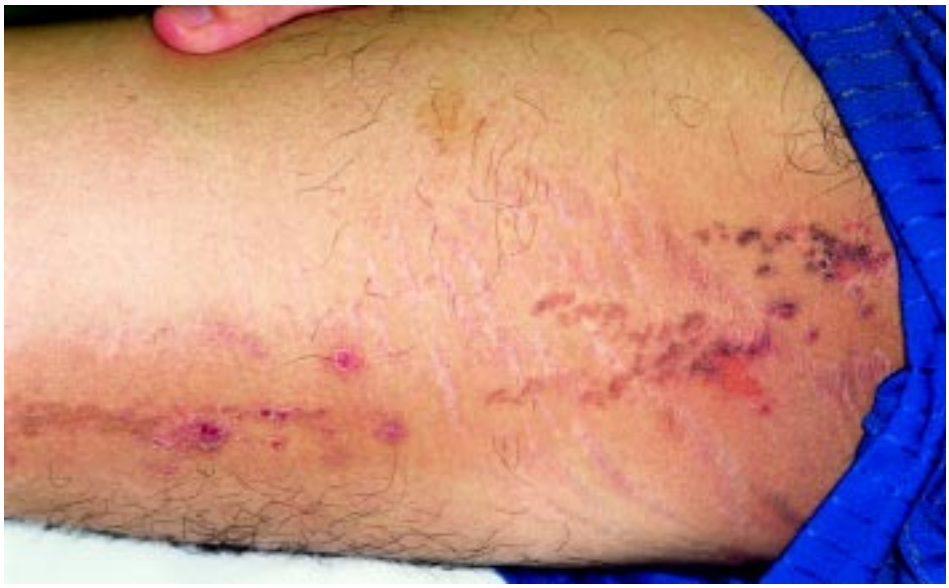

Figure 2: Brownish macules of linear porokeratosis over the right groin with surrounding steroid induced striae

because of local irritation. A few sessions of cryotherapy had been carried out but were again not beneficial. The patient did find topical calcipotriol ointment helpful in partially relieving the itch and the thickness of the keratotic ridge.

Discussion

Most of the cases of porokeratosis seen locally are sporadic rather than familial. Topical calcipotriol, which acts by promoting cellular differentiation, may be useful as a topical treatment for porokeratosis.

REVIEW ON LINEAR POROKERATOSIS

Porokeratosis is a disorder of keratinization. Clinically the lesion is sharply demarcated and hyperkeratotic. Five clinical variants have been described: (1) classical porokeratosis of Mibelli (PM); (2) disseminated superficial actinic porokeratosis (DSAP); (3) linear porokeratosis (LP); (4) porokeratosis plantaris, palmaris et disseminata (PPPD); (5) porokeratosis punctata palmaris et plantaris (PPPP).[1]

Clinical features

The incidence of linear porokeratosis is not known, but is believed to be rare. Linear porokeratosis comprises 3.5-16.7% of the total number of reported porokeratosis in various reviews.[2]

Linear porokeratosis may occur in a unilateral, linear or systematized form. The lesions are typically small and annular, with central atrophy and a raised edge. They are grouped and linearly arranged, more commonly affecting the extremities than the trunk. Onset is usually in infancy and childhood, but no definite inheritance pattern has been established. Association with other clinical variants of porokeratosis has been reported.[3]

Etiology

The etiology is unknown, but an autosomal dominant mode of inheritance has been reasonably well established for PM, PPPD and DSAP. However, numerous non-familial cases have also been reported. It has been suggested that perhaps it is the tendency to develop abnormal clones that is inherited, and additional triggering factors are needed before the lesions become manifested. For example, in DSAP, lesions occurred mainly on sun-exposed area, and some patients may note exacerbation in summer. Porokeratosis has also been reported in immunosuppressed patients, as seen in those following liver or renal transplantation.[4] Because of the similarities of clinical and histological appearances of the various types of porokeratosis, as well as the fact that different variants of porokeratosis may coexist in one patient or in different members of

an affected family, it has been suggested that the various types of porokeratosis may represent different phenotypic expressions of a common genetic aberration.

Histopathology

The characteristic cornoid lamella lies in the peripheral rim of the lesion. It is a thin column of parakeratotic cells with an absent or decreased underlying granular zone. There may be vacuolated or dyskeratotic cells in the spinous layer. Studies have demonstrated the presence of abnormal DNA ploidy and abnormalities in keratinocyte maturation beneath the parakeratotic column.[5] Cultured fibroblasts taken from the underlying dermis has also been found to have instability of the short arm of chromosome 3.[5] Over-expression of p53 in the nuclei of keratinocytes in the basal layers of the epidermis beneath the cornoid lamella has also been demonstrated.[6] All of these may explain the malignant potential of the porokeratotic lesion. It has to be noted that cornoid lamella is not diagnostic of porokeratosis. It may also be seen in other conditions like seborrhoeic keratosis, solar keratosis, squamous cell carcinoma, basal cell carcinoma, verruca vulgaris, scar and milia.[7]

Malignant potential

From various retrospective analyses, it is found that between 7-11.6% of patients suffering from porokeratosis will develop malignant or premalignant change in the lesion.[2] However, this malignant potential does not occur to the same degree in all types of porokeratosis. From a review of Japanese case reports, Otsuka et al concluded that malignant transformation was absent in DSAP but present in other types.[8] In a survey of English language reports in the period 1964-1994, Sasson and Krain found that large lesions, those of long-standing duration and the linear type were at greatest risk.[9]

Treatment

Small lesions may be amenable to excision, cryotherapy or electrodesiccation. Topical 5-fluorouracil has been tried with good results in some cases.[10] For extensive lesions, oral retinoid has been reported to be useful.[11] However recurrence occurs soon after stopping the drug. Satisfactory results have also been reported with CO_2 laser,[12] 585 nm pulsed dye laser,[13] and diamond fraise dermabrasion.[14] Therapeutic measures which might increase the malignant potential of the porokeratotic lesion, such as irradiation, immunosuppression or excessive ultraviolet exposure should be avoided.

Learning points:

Cornoid lamella is not diagnostic of porokeratosis. It may be seen in seborrhoeic keratosis, solar keratosis, squamous cell carcinoma, basal cell carcinoma, verruca vulgaris, scar and milia.

References

1. Griffiths WAD, Leigh IM, Marks R. Porokeratosis, in Champion RH, Burton JL, Ebling FJG (eds): Textbook of Dermatology, 5th ed, Blackwell Co, Oxford, 1992, p1388.
2. Taniguchi Y, Yuasa T, Shimizu M. Linear Porokeratosis, J Dermatol 1993;20(8):489-92.
3. Dover JS, Phillips TJ, Burns DA, et al. Disseminated superficial actinic porokeratosis. Coexistence with other porokeratotic variants, Arch Dermatol 1986;122(8):887-9.
4. Hunt SJ, Sharra WG, Abell E. Linear and punctate porokeratosis associated with end-stage liver disease. J Am Acad Dermatol 1991;25(5 Pt 2):937-9.
5. Imakado S, Otsuka F, Ishibashi Y, et al. Abnormal DNA ploidy in cells of the epidermis in a case of porokeratosis. Arch Dermatol 1988;124:331-2.
6. Magee JW, McCalmont TH, LeBoit PE. Over-expression of p53 tumour suppressor protein in porokeratosis. Arch Dermatol 1994;130:187-90.
7. Wade TR, Ackerman AB. Cornoid lamellation- A histologic reaction pattern. Am J Dermatopathol 1980;2:5-15.
8. Otsuka F, Huang J, Sawara K, et al. Disseminated porokeratosis accompanying multicentric Bowen's disease: characterization of porokeratotic lesions progressing to Bowen's disease. J Am Acad Dermatol 1990;23:355-9.
9. Sasson M, Krain AD. Porokeratosis and cutaneous malignancy. A review. Dermatol-Surg 1996;22(4):339-42.
10. McDonald SG, Peterka ES. Porokeratosis (Mibelli): Treatment with topical 5-fluorouracil. J Am Acad Dermatol 1983;8:107.
11. Danno K. Etretinate treatment in disseminated porokeratosis. J Dermatol 1988;15:440.
12. Barnett JH. Linear porokeratosis: treatment with the carbon dioxide laser. J Am Acad Dermatol 1986;14(5 Pt 2):902-4.
13. Alster TS, Nanni CA. Successful treatment of porokeratosis with 585nm pulsed dye laser irradiation. Cutis 1999;63(5):265-6.
14. Cohen PR, Held JL, Katz BE. Linear porokeratosis: successful treatment with diamond fraise dermabrasion. J Am Acad Dermatol 1990;23(5 Pt 2):975-7.