Linear Porokeratosis

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**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.

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*Figure 1: Linear porokeratosis affecting the medial aspect of the right lower limb*
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).\

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.\

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.\

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. 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. Cultured fibroblasts taken from the underlying dermis has also been found to have instability of the short arm of chromosome 3. Over-expression of p53 in the nuclei of keratinocytes in the basal layers of the epidermis beneath the cornoid lamella has also been demonstrated. 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.

## 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. 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. 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.

## Treatment

Small lesions may be amenable to excision, cryotherapy or electrosiccation. Topical 5-fluorouracil has been tried with good results in some cases. For extensive lesions, oral retinoid has been reported to be useful. However recurrence occurs soon after stopping the drug. Satisfactory results have also been reported with CO₂ laser, 585 nm pulsed dye laser, and diamond fraise dermabrasion. 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