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Porokeratosis of Mibelli

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Summary

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Summary : A 72-year-old man had noticed, in his early forties, the appearance of well-defined papulous hyperkeratotic lesions, with increasing growth, located on both sides of his feet. After twenty-five years he consulted a dermatologist for the first time. Physical examination showed annular papules and rose-coloured plaques with atrophic centres, some of them hypopigmented, with higher and irregular borders, separated from the surrounding skin by longitudinal and well-defined furrows. The lesions presented variable sizes and shapes, some of them punctate, involving exclusively and in a bilateral form, both sides, back and sole of the feet (Figs. 1 and 2). The patient did not report any subjective symptoms. He was immunocompetent and did not remember that any relative had the same disease, nor had he been subjected to radiation treatment.

Pictures

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A diagnosis of porokeratosis of Mibelli was made based on the clinical appearance and histological examination of a biopsy specimen. Histologically, the epidermis showed a slight acanthosis with invaginations filled with columns of parakeratosis, forming the cornoid lamella, next to the orthokeratotic stratum corneum of the adjacent epidermis *(Fig. 3)*. Immunohistochemical staining of dermal inflammatory infiltrate below the cornoid lamella (CD 3, CD 4, S-100 protein) showed that most cells were helper T lymphocytes with some intermingled Langerhans cells.

The lesions regressed partially with lubrication and keratolytic treatment but afterwards relapsed. No lesion suggesting malignant degeneration was found.

Porokeratosis is a dermatosis which results from a specific alteration of keratinization [1].

Five clinical types of porokeratosis are known:

1. Classic porokeratosis of Mibelli.

2. Disseminated superficial porokeratosis and disseminated superficial actinic porokeratosis.

3. Porokeratosis palmaris et plantaris disseminata.

4. Linear porokeratosis.

5. Punctate porokeratosis.

It has been postulated that porokeratosis results from proliferation of an abnormal cellular clone to which several trigger factors have been suggested: irradiation, infective agents, trauma and immunosuppression [2]. Mibelli's porokeratosis has occasionally been described following immunosuppression [3]. Nevertheless, none of those factors was apparently implicated in our clinical case. The aetiology is still unknown but it has been suggested that the presence of helper T cells and some Langerhans cells in Mibelli's porokeratosis, as in this present case, is evidence for immunological mechanisms induced by antigen presentation [4].