Dermal Amyloid Deposits in Disseminated Superficial Porokeratosis

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Sir,

The presence of dermal amyloid deposits in lesions of porokeratosis has rarely been described in the literature (1–11), and the great majority of patients reported have an oriental origin. We present the clinical, histopathological and immunohistochemical findings of two white Spanish patients with secondary localized cutaneous amyloidosis associated with disseminated superficial porokeratosis (DSP). We reviewed the literature with regard to the association of amyloid deposits with the different forms of porokeratosis.

CASE REPORTS

Case 1

A 70-year-old man presented with a 1-year history of multiple asymptomatic annular, slightly pigmented lesions on his back and upper limbs. There was no family background of a similar dermatosis. Physical examination showed multiple small brownish lesions, up to 1.5 cm, with atrophic centres and slightly raised hyperkeratotic borders on the trunk and upper extremities. Two lesional biopsies from distant sites revealed similar findings consisting of an atrophic epidermis with a cornoid lamella in its border. The dermis showed oedema, vascular ectasia and an amorphous eosinophilic material beneath and between the cornoid lamellae (Fig. 1). This material stained with Congo red and tioflavin. Immunohistochemical studies revealed positivity (+++) of this amyloid material with anti-cytokeratin (CK) antibodies CK34βE12 (CK-903), and slight positivity (+) with CK 5/8 and CK 5/6/18. Amyloid deposits were not found in a biopsy from non-lesional skin.

Case 2

A 70-year-old woman complained of multiple small annular pigmented macules on the dorsum of both upper limbs which had persisted over the previous 3 years (Fig. 2). She had received treatment with local 5-fluorouracil with only slight improvement. Two lesional biopsies from distant sites showed cornoid lamellae, some of them located over dilated infundibula, and a thin epidermis between the lamellae. There were extensive amyloid deposits in both biopsies in the upper dermis. No amyloid material was found in a biopsy from non-lesional skin. Immunohistochemical studies were performed showing strong positivity for anti-CK in the epidermis as well as in the subepidermal deposits (Fig. 3).

DISCUSSION

Microscopic amyloid deposits have been described in association with multiple cutaneous pathologies, including seborrhoeic keratosis, intradermal melanocytic naevus, sweat-gland tumours, pilomatrichoma, trichoepithelioma, solar keratosis, Bowen's disease, basal cell carcinoma, dermatofibroma and porokeratosis. Ten of 17 previously reported cases of amyloid deposits

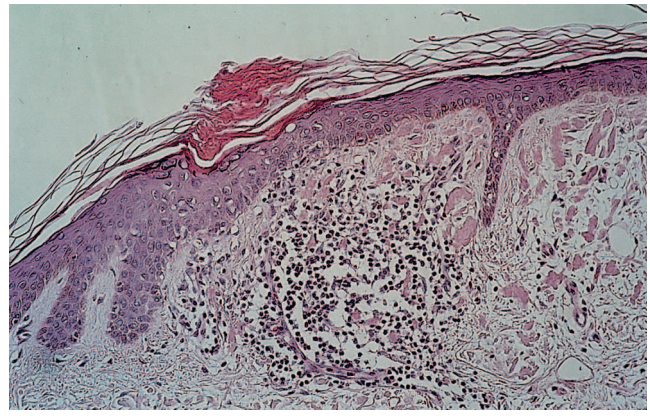

Fig. 1. Biopsy specimen from a lesion on the back in Case 1 showing a cornoid lamella associated with deposits of an amorphous eosinophilic material (haematoxylin-eosin).

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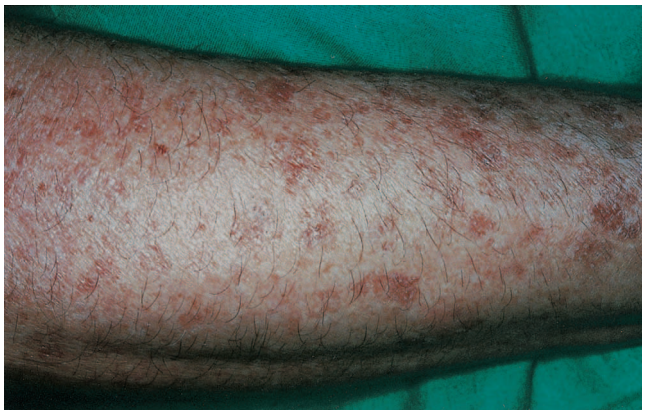

Fig. 2. Lesions of disseminated superficial porokeratosis on the forearm in Case 2.

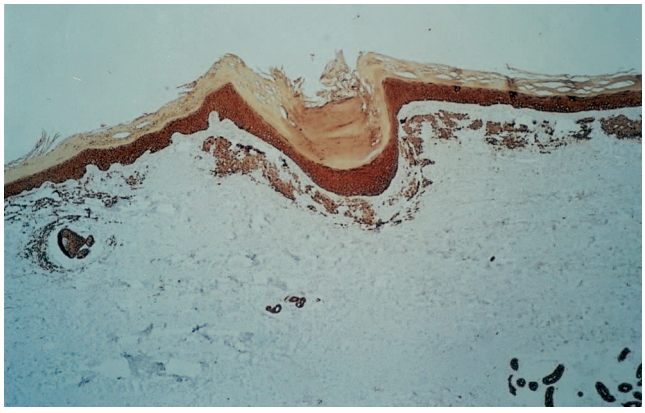

Fig. 3. Positive staining with anti-keratin antibodies CK34βE12 (CK-903) in Case 2.

in porokeratosis were patients with DSP, and 7 had porokeratosis of Mibelli (1–11).

An interesting fact is that almost all the patients described have an oriental origin, mainly Japanese. In our two patients, we found dermal amyloid deposits in association with lesions of porokeratosis, under and between the cornoid lamellae, but we did not find this material in non-lesional skin, which suggests that the association is not causal. In both patients, these amyloid deposits stained with anti-keratin antibodies, as in localized cutaneous amyloidosis, especially in its secondary form, suggesting that the amyloid derives from epidermal keratinocyte degeneration (1, 8–10).

The association of amyloid and porokeratosis is probably understimated. Amantea et al. (8) reviewed 30 cases of DSP with histochemical, immunohistochemical and ultrastructural methods disclosing amyloid deposits in two cases that had been missed in sections stained with haematoxylin-eosin.

The pathogenesis of all types of epidermally derived amyloid deposits may be related to a variable host response directed against keratinocytes in an interface reaction pattern, which might explain why we find these deposits in porokeratosis and in very disparate entities (12–14).

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