

## LICHEN AMYLOIDOSUS

### *An Unusual Clinical Variant Associated with the Koebner Phenomenon*

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**Abstract.** A patient with an unusual clinical variant of lichen amyloidosis is described in whom the Koebner phenomenon could be elicited. The significance of this finding and its possible relationship to the occurrence of the Koebner phenomenon in lichen planus is discussed.

The Koebner or isomorphic phenomenon has been defined as the induction by and at the site of non-specific trauma of skin changes of a type spontaneously present elsewhere (8). Koebner (5) first described this association in psoriasis, but it is now known that the isomorphic phenomenon can occur in a wide variety of dermatoses. The traumatic effect of light or heat upon the skin can induce an exacerbation in several dermatoses, especially lupus erythematosus, herpes simplex, Darier's disease and pityriasis rubra pilaris. The development of lesions in scratch marks or superficial abrasions is not infrequently found in plane warts and lichen planus. This report describes the development of a Koebner phenomenon in the scratch marks of a patient with an unusual clinical variant of lichen amyloidosis. This association does not appear to have been recorded previously.

### CASE REPORT

A 43-year-old Caucasian female first developed a papular eruption in the cubital fossae about 1 year prior to her first attendance. Since then the eruption had gradually spread to involve the flexor aspect of the lower forearms, extensor aspects of both lower legs and was accompanied by moderately severe pruritus. She had also noticed increasing pigmentation of the lower trunk. Her general health was good and there was nothing significant in her past or family history.

Examination revealed numerous small, flat-topped shiny papules (Fig. 1) in both cubital fossae, and these were also present to a lesser extent on the flexor aspect of the lower forearm. Several papules were arranged in a linear distribution suggestive of a Koebner phenomenon

(Fig. 1). Somewhat larger papules were present on the extensor surface of the lower legs, but these were all hyperpigmented. The skin over the lower back, abdomen and groins showed a diffuse macular hyperpigmentation (Fig 2). No lesions were present on mucous membranes and no abnormalities were detected on general physical examination.



*Fig. 1.* Cubital fossa: clusters of shiny flat-topped papules are present; in the lower part some are arranged in a linear distribution suggestive of a Koebner phenomenon.

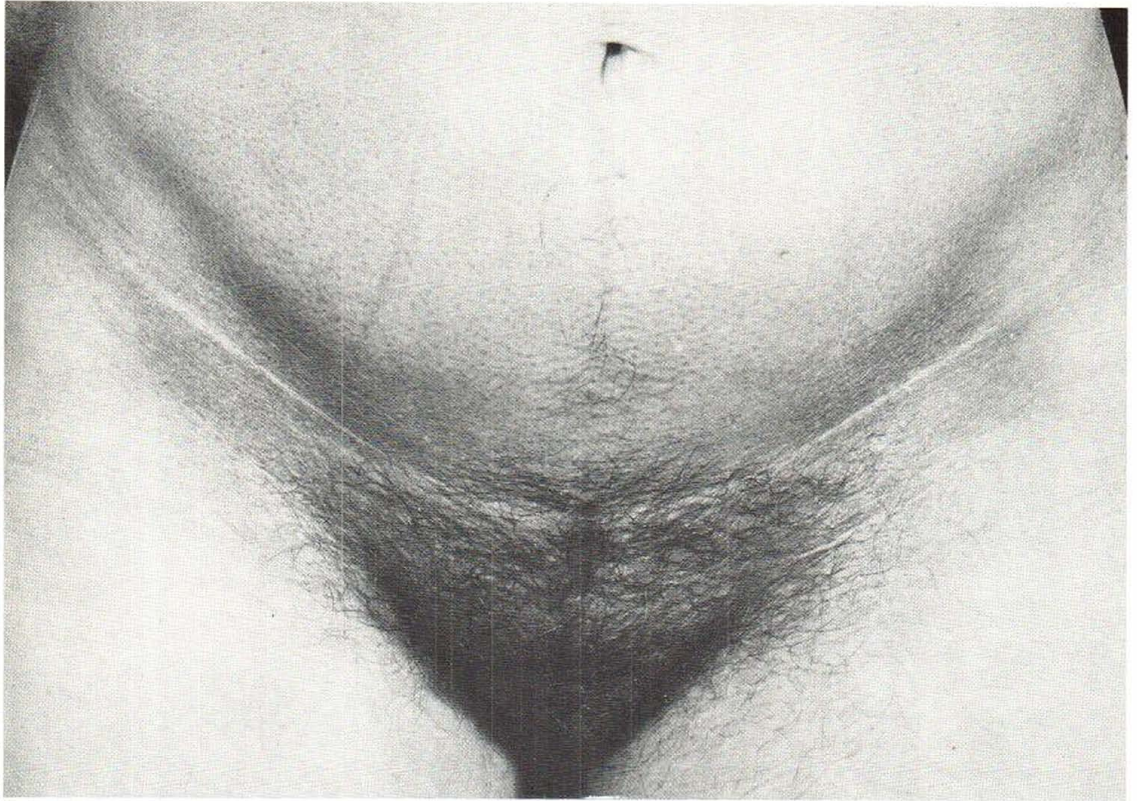


Fig. 2. Diffuse macular hyperpigmentation affecting predominantly the groins and supra-pubic region.

*Investigations*

Haemoglobin: 94% W.B.C. 7 000 (normal differential);  
E.S.R.: 22 mm/1 hour;  
Urine: No albuminuria or Bence Jones protein could be detected;

Blood Urea: 18 mg%, normal electrolytes; W.R. negative;  
A.N.F. negative;  
Serum proteins: 8.1 g/100 ml, electrophoresis revealing a slight increase in  $\alpha_2$  globulin.  
Chest X-ray: Normal.

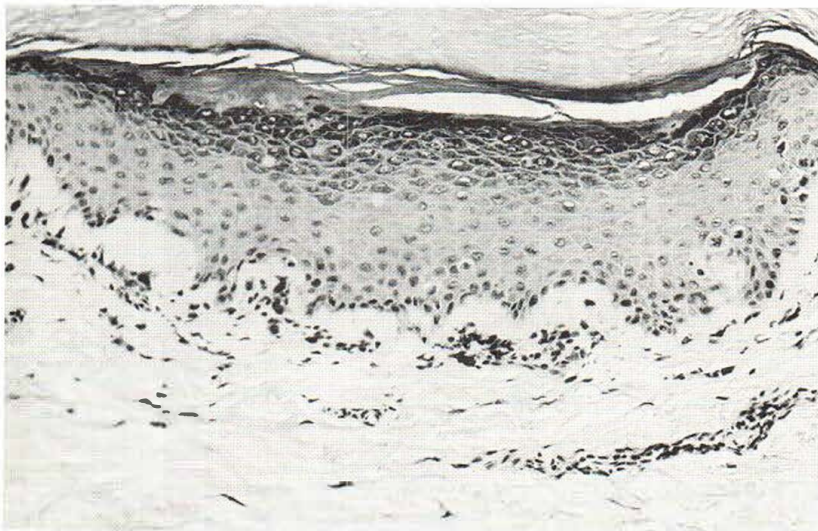


Fig. 3. Biopsy from papule in cubital fossa; H & E,  $\times 80$ . The small amyloid deposits are situated in the dermal papillae beneath the papule. A few hyaline bodies are also present in the lower epidermis. Pigment-containing macrophages are present in the papillary body but the inflammatory infiltrate is sparse.





Fig. 4. H & E,  $\times 430$ . Several hyaline bodies, some containing shrunken nuclei are situated in the lower epidermis, immediately above the amyloid deposits (A).

#### *Histopathology*

Biopsies were obtained from papules in the cubital fossae and lower legs as well as from a macular hyperpigmented area on the lower back. The sections from both papular lesions showed similar histological features. The epidermis showed considerable hyperkeratosis and moderate acanthosis and in places contained a few hyaline bodies in the lower epidermis (Fig. 3). Small hyaline deposits were found in the papillary body and these were present in the majority of dermal papillae. Green birefringence was obtained with polaroid light on sections stained with congo red and an intense fluorescence with thioflavine T, thus confirming that the deposits consisted of amyloid. Some of the hyaline bodies contained shrunken pyknotic nuclei and these were in close proximity to the amyloid

deposits (Fig. 4). A sparse, predominantly lymphocytic infiltrate was situated around blood vessels in the upper dermis, and several melanin-containing macrophages were present within or just below the amyloid deposits.

The biopsy from a macular hyperpigmented area on the lower back showed diffuse pigmentary incontinence but no amyloid could be demonstrated in the papillary body.

#### *Induction of the Koebner phenomenon*

A series of linear scratches, sufficient to produce minute bleeding points, were performed on an area of clinically uninvolved skin on the flexor aspect of the upper forearm. After 3 weeks a series of shiny micro-papules developed in the line of scratches which have since persisted.

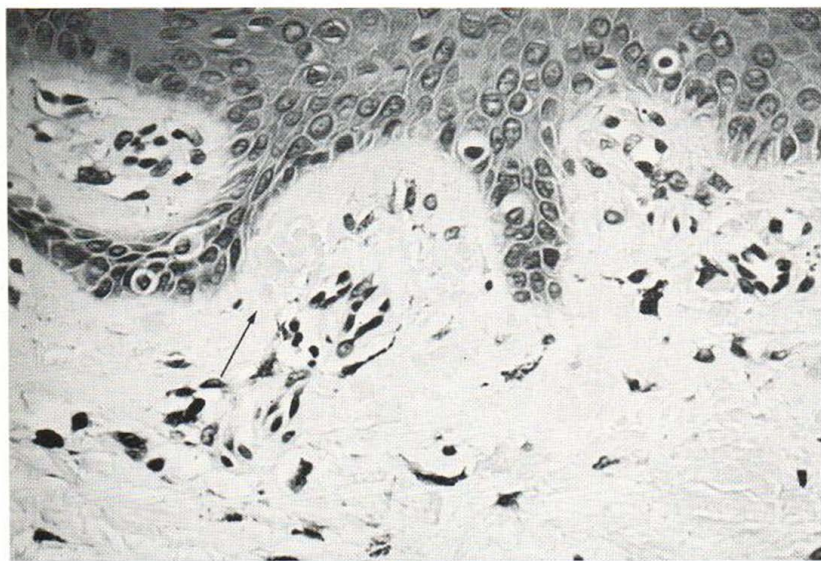


Fig. 5. Biopsy of papule induced by Koebner's phenomenon; H & E,  $\times 190$ . Small hyaline bodies are situated in the dermal papillae which stain positively for amyloid.

A punch biopsy of one of these papules revealed small hyaline bodies in the dermal papillae which stained positively for amyloid (Fig. 5).

### COMMENT

This is believed to be the first recorded association of lichen amyloidosis and the development of the Koebner phenomenon. Dostrovsky & Sagher (3) noted the reappearance of papules at the site of biopsy in a single case of lichen amyloidosis and Morishima (7) observed in a further case the development of papules in scars caused by vaccination and moxibustion.

The presence of shiny flat-topped papules on the flexor aspect of the forearm and linear lesions suggestive of a Koebner phenomenon led to an initial diagnosis of lichen planus. Koenigstein (6) noted that certain types of lichen amyloidosis were clinically very similar to lichen planus. Black & Wilson Jones (1) recently observed certain histological similarities between the two conditions; in particular the small amyloid bodies in the papillary body were similar in size and appearance to the colloid bodies of lichen planus. It was suggested that in lichen amyloidosis degenerating epidermal cells could be converted into amyloid. The Koebner phenomenon is not infrequently observed in lichen planus particularly in scratch marks. In a recent study of this phenomenon in lichen planus (Black, 1970, unpublished date), it was found that lichen planus papules could only be induced in linear scratches which had caused small bleeding points, and after an interval of approximately 3 weeks. Similar findings were found in the present case in that lichen amyloidosis papules were also induced after a period of 3 weeks and only in those scratches which had led to minute bleeding points.

Papular lichen amyloidosis most commonly affects the extensor surface of the lower limbs. The prominent involvement of the cubital fossae in this case appears to be most unusual. Shanon (9) has described papular lichen amyloidosis affecting unusual sites such as the inter-mammary and the

suprapubic areas, but not in the cubital fossae. Morishima (7) has recently drawn attention to a variant of lichen amyloidosis in which extensive macular hyperpigmentation may be found in addition to typical lichen amyloidosis papules. Similar cases have been described by Freudenthal (4); Dostrovsky & Sagher (3) and Bloom (2). Morishima was able to demonstrate small amyloid deposits from the hyperpigmented areas in his cases, and also commented on the widespread pigmentary incontinence. Although no amyloid could be demonstrated in a biopsy from the hyperpigmented area in this case, pigmentary incontinence was a conspicuous feature.

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