

MIXED (LICHENOID AND MACULAR) CUTANEOUS AMYLOIDOSES

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Abstract. Between October 1972 and April 1974 a total of four patients suffering from lichen amyloidosis were admitted to the wards of the Department of Dermatology, University Hospital, Valladolid. All these patients also showed a macular amyloidosis associated with the lichenoid eruption. The authors emphasize that macular amyloidosis is quite commonly found as an associated and closely related condition in lichen amyloidosis.

Key words: Amyloid; Cutaneous amyloidoses; Lichen amyloidosis; Macular amyloidosis

In a recent paper, Brownstein et al. (2) have suggested the term "biphasic" for the existence of both macular and lichenoid amyloidosis in the same patient. Earlier, French authors (3, 4, 6, 7) have called attention to the association of both conditions, and formerly a probably similar case in a 25-year-old woman was registered in 1923 by Gutmann (5).

The purpose of this paper is to report the finding of macular amyloidosis in four patients with lichen amyloidosis seen at the University Hospital, Valladolid, between October 1972 and April 1974.

CASE REPORTS

Case 1

A 78-year-old man complained of an intensely pruritic papular eruption several years ago. He first came under the care of our Department on October 13, 1972. On examination, there were numerous closely grouped papules on the legs (Fig. 1), the extensor aspects of the thighs, the popliteal flexures and the lumbosacral region. These papules, ranging in diameter from 1 to 3 mm, were yellow-brown in colour and showed some scaling. On the back there were several poorly demarcated areas composed of tenuous brown macules, irregularly interspersed with the normal skin and with some hypomelanotic areas.

Two biopsy specimens were taken from the left pretibial area and the upper back.

Case 2

A 58-year-old male was first seen in May 3, 1973, with a 6-year history of pruritus of intense severity. Itching affected initially the back and, 2 years later, also the limbs. On examination, the patient showed a papular eruption on the extensor aspects of the forearms and on external area of the right thigh, clinically diagnosed as lichen amyloidosis. Careful observation of the skin revealed some slightly pigmented macules on the upper back (Fig. 2). As in the

Table 1. Results of microscopic examination for amyloid

Case	Lesion	Site	Conventional methods	Thioflavine T	Electron microscopy
1	Lichenoid	Leg	Positive	Positive	Positive
	Macular	Back	Positive	Positive	Positive
2	Lichenoid	Forearm	Positive	Positive	Positive
	Lichenoid	Thigh	Positive	Positive	Positive
	Macular	Back	Positive	Positive	Positive
3	Lichenoid	Leg	Positive	Positive	Positive
	Lichenoid	Forearm	Doubtful	Doubtful	Positive
	Macular	Back	Doubtful	Doubtful	Positive
	Macular	Back	Negative	Negative	Positive
4	Macular	Back	Negative	Positive	Positive
	Lichenoid	Lumbar	Positive	Positive	Positive
4	Lichenoid	Lumbar	Positive	Positive	Positive
	Macular	Back	Positive	Positive	Positive



Fig. 1. Lichen amyloidosis. External aspect of the left leg (case 1).

preceding case, these pigmented macules appeared interlaced with small whitish patches and areas of normal appearance.

Biopsy specimens were obtained from the right forearm

(lichen amyloidosis), the right thigh (lichen amyloidosis), and the upper back (macular pigmented lesion).

Case 3

A 50-year-old housewife was referred to us on June 8, 1973 with a history of an itching eruption on the limbs and back, about 15 years previously. Examination of this patient revealed a papular and lichenoid eruption on the legs and forearms and pigmented macules on the chest and back.

Five skin biopsies were performed on two lichenoid and three macular lesions.

Case 4

This 45-year-old housewife was seen by us in April 1974 whose chief complaint was generalized pruritus. Itching had been present for more than 10 years. On examination, she showed a widespread papular eruption over the lumbosacral region and the buttocks, the lower back and the extensor surfaces of the forearms. On close inspection, we saw extensive hyperpigmented macular areas on the upper back, along with speckles of whitish discoloration.

Skin biopsies were taken from the lumbar region (lichen amyloidosis) and the upper back (pigmented brown macule).

METHODS

The biopsy specimens were examined with both light and electron microscopes. A part of each specimen was fixed in 10% neutral formalin and processed for routine paraffin-blocked sections. The sections were stained with hematoxylin-eosin, conventional stains for amyloid (crystal violet, methyl violet and congo red), and thioflavine T. The samples for electron microscopy were fixed in a 2% glutaraldehyde solution buffered with sodium cacodylate buffer, and post-fixed in a 1% osmium tetroxide solution. After fixation the blocks were dehydrated with acetone and embedded in an epoxy resin (Araldite). The material was sectioned in an ultramicrotome LKB-111 and stained with uranyl acetate and lead citrate. The sections were observed in a Zeiss EM9A electron microscope.

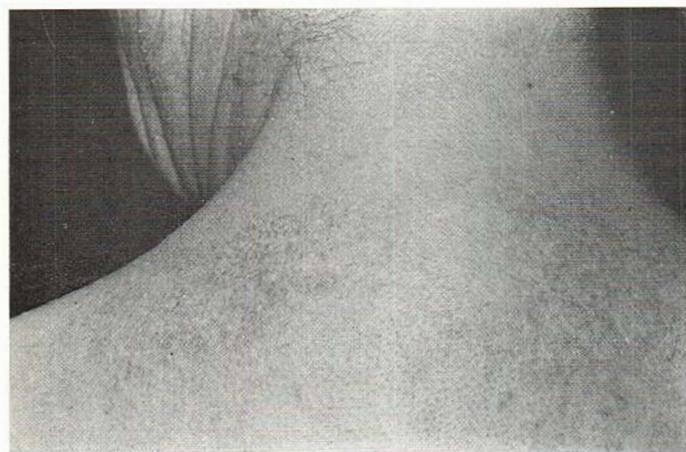


Fig. 2. Macular amyloidosis of the upper back (case 2).

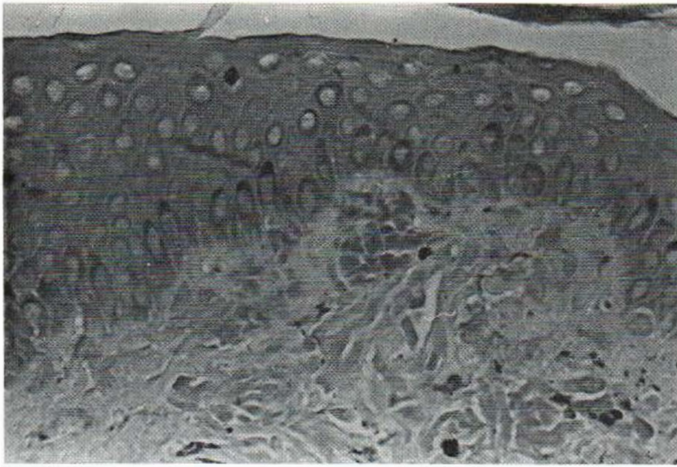


Fig. 3. Macular amyloidosis of the upper back. Deposits of amyloid in papillary dermis. Crystal violet, $\times 185$ (case 2).

RESULTS

The results of the microscopic examination for amyloid are summarized in Table I, and some of them are shown in Figs. 3-7.



Fig. 4. Lichen amyloidosis. Biopsy specimen from the leg. Amyloid deposits in papillary dermis, hyperkeratosis and epidermal hyperplasia. Thioflavine T, $\times 350$ (case 1).

Ultrastructurally, in all the specimens examined, deposits of a distinctly filamentous material were detected in the papillary and upper reticular dermis, showing the characteristics of amyloid filaments. These filaments measured between 70 and 100 Å in diameter and in cross sections they showed a dense central core (Fig. 5).

The electron microscopic picture was nearly the same in lichenoid and macular lesions. However, small isolated lobules of amyloid confined to the papillary bodies could be observed as the most common picture in macular lesions (Figs. 6, 7), whereas lichen amyloidosis frequently showed massive infiltration of the upper dermis by numerous close grouped amyloid islands.

Under light microscopy, hyperkeratosis and epidermal hyperplasia were prominent features in biopsy specimens from lichen amyloidosis. By contrast, the epidermis appeared normal in macular lesions.

DISCUSSION

The finding of macular amyloidosis in all our patients suffering from lichen amyloidosis is strongly indicative of the character of this association, and not merely fortuitous. It seems likely that macular amyloidosis is a constant or at least a quite common feature in lichen amyloidosis.

These findings support the previous idea of Brownstein et al. (2) about the existence of a link between macular and lichenoid forms of primary localized cutaneous amyloidosis, and lead us to

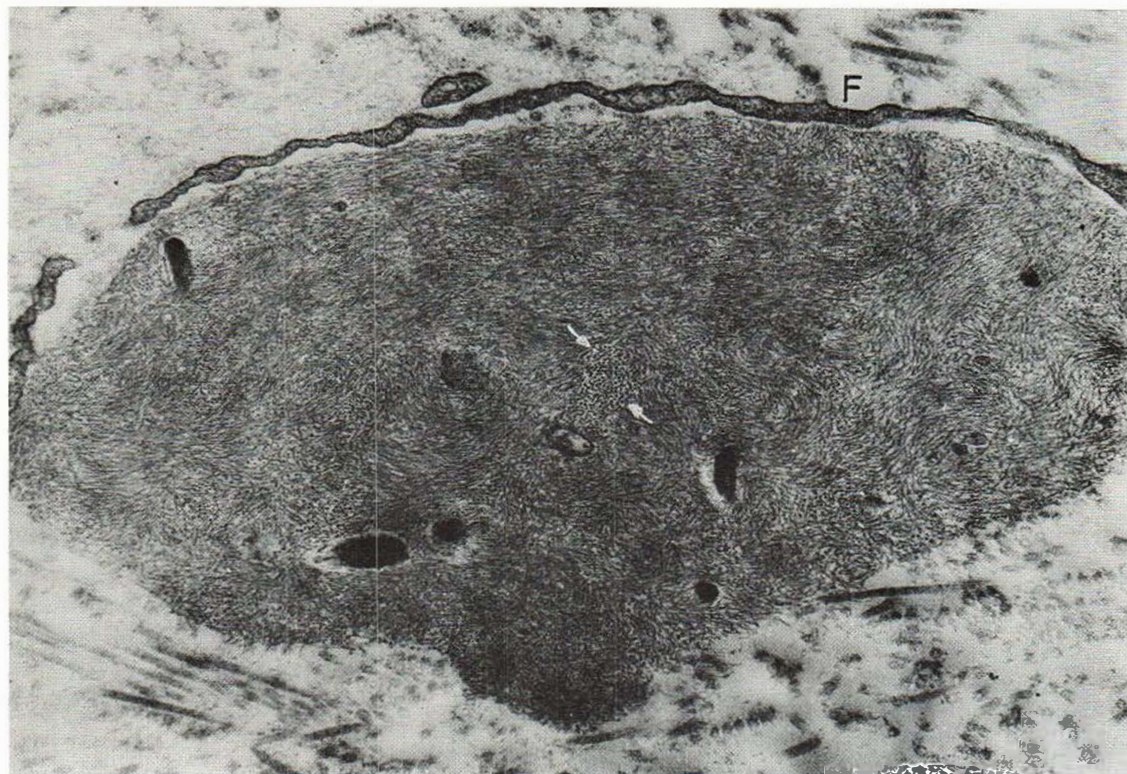


Fig. 5. Lichen amyloidosis of the legs. One amyloid island near to the cytoplasmic process of a fibroblast (F). In cross sections the amyloid filaments show a dense central core (arrows). $\times 26\,300$ (case 3).



Fig. 6. Macular lesion of the back. Amyloid island (A) in a papillary body. Arrows indicate the basal lamina, $\times 18\,200$ (case 3).

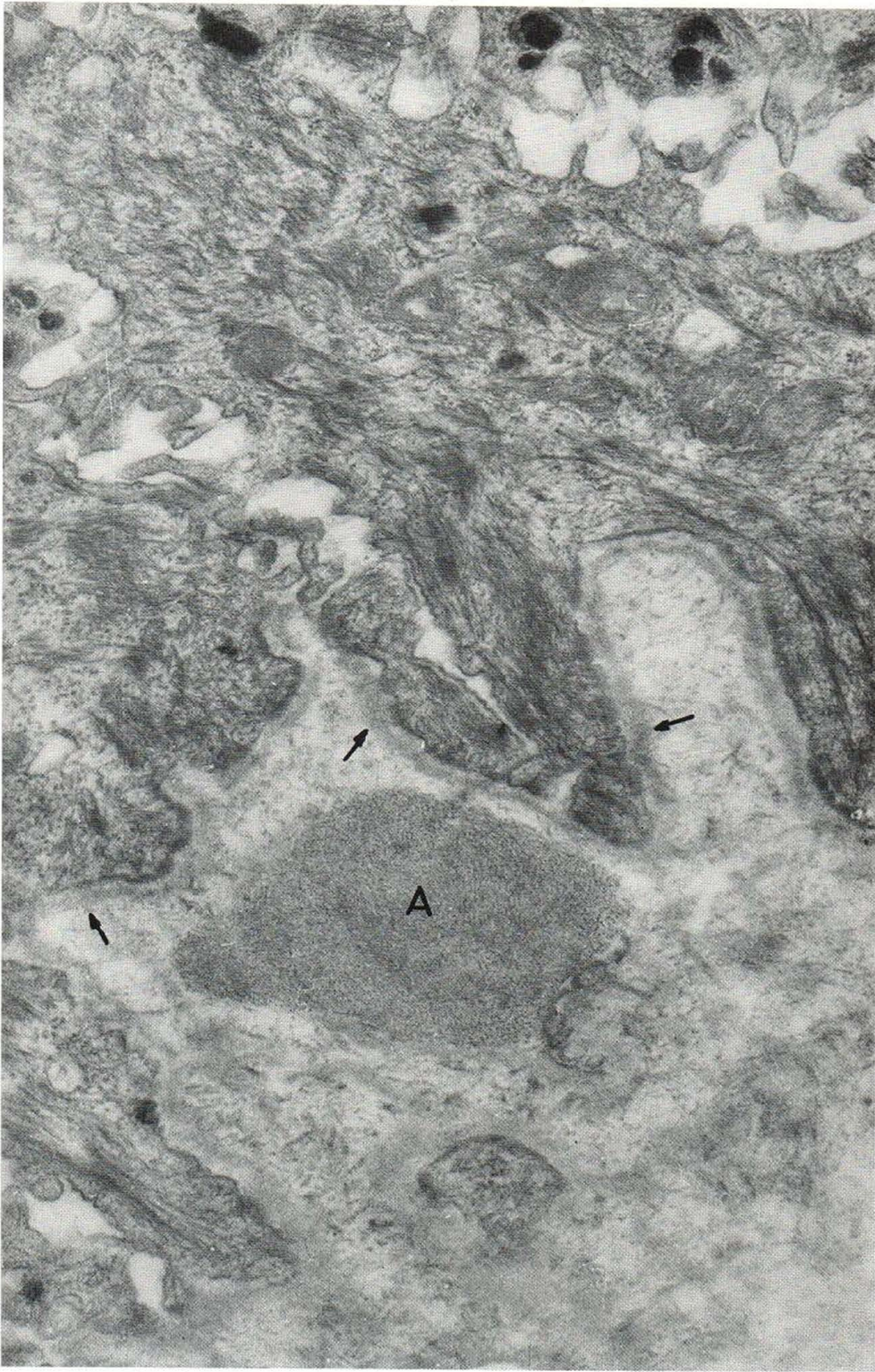


Fig. 7. Macular amyloidosis of the back. Amyloid deposit (A) near to the basal lamina (arrows). $\times 24\,300$ (case 1).

recommend a careful search for associated macular manifestations in patients with lichen amyloidosis.

Our observations also confirm that epidermal changes with hyperkeratosis and hyperplasia of the rete ridges are distinctive features in lichen amyloidosis, as was previously pointed out by Brownstein et al. (1, 2).

Furthermore, our results show that electron microscopy is the most reliable diagnostic method in cutaneous amyloidosis.

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