

## SHORT COMMUNICATION

### Localized Cutaneous Amyloidosis Associated with Poikilodermatous Mycosis Fungoides

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Secondary localized cutaneous amyloidosis (LCA) occurs in association with skin inflammatory disorders or around various epithelial tumours. We describe here a case of LCA in association with generalized, patch-stage mycosis fungoides (MF).

#### CASE REPORT

A 68-year-old man had had pruritic scaly eruptions on the trunk and extremities for 54 years. He had not received any medical treatment for his eruptions before he visited our department. He had no history of serious illness. Initial examination revealed diffuse atrophic erythema with pityriatic scales on the trunk and extremities (Fig. 1). Hyperpigmentation, hypopigmentation and telangiectasia were also intermingled with the erythema. A biopsy specimen from his left thigh revealed band-like lymphocytic infiltration in the papillary dermis (Fig. 2a). Epidermotropism and Pautrier's microabscesses were also observed (data not shown). In some areas, focal interface vacuolar changes were evident, with associated pigment incontinence in the biopsy specimen (Fig. 2b). The infiltrating lymphocytes were positive for CD3 (Fig. 2c), CD4 (Fig. 2d), and CD8 (Fig. 2e). The population of CD8<sup>+</sup> cells predominated over that of CD4<sup>+</sup> cells.

In addition, amorphous eosinophilic deposits were present in the papillary dermis and around the axnaxal, surrounded by lymphocytes infiltration (Fig. 2a; *asterisk*). The deposits were positive for direct fast scarlet (Fig. 2f). We also performed Congo red staining, and the deposits were negative for this staining (data not shown). Therefore, we checked the polarizing

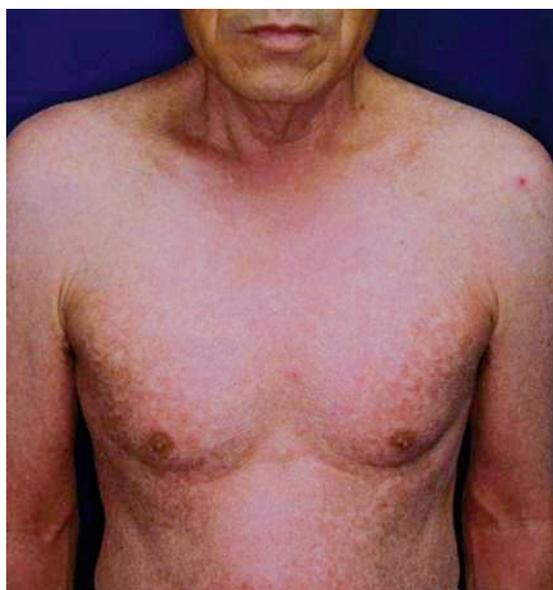


Fig. 1. Diffuse atrophic erythema, hypopigmentation, hyperpigmentation and telangiectasia on the patient's trunk.

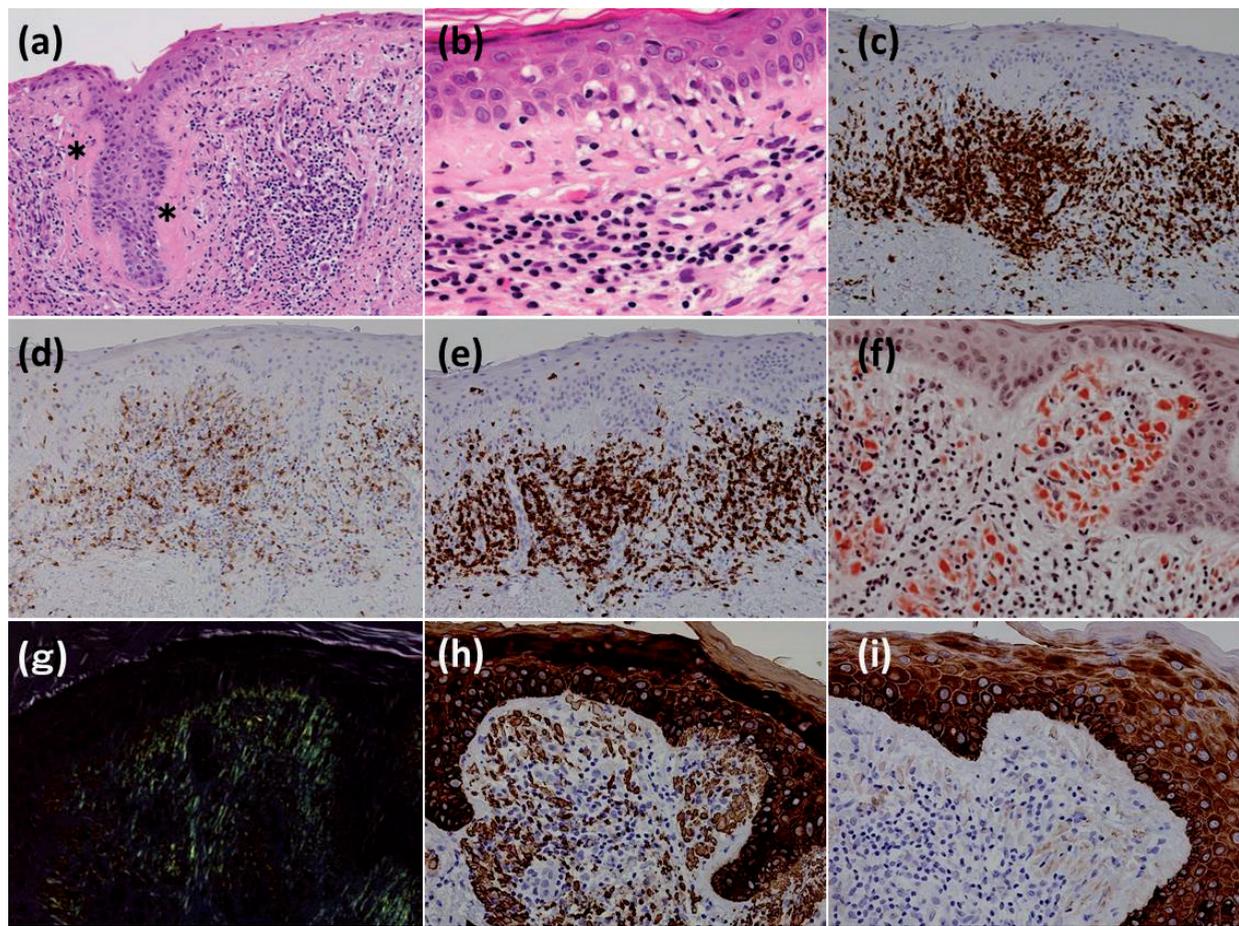
microscopic findings with the specimens stained by direct fast scarlet, and this staining revealed green birefringence (Fig. 2g), therefore we finally considered these deposits as amyloid-like materials. Immunohistochemical staining for keratin antibody (34 $\beta$ E12 (which reacts with cytokeratin (CK) 1, 5, 10, 14), CK5/6, CK7, CK8, CK13, CK14, CK18, CK19 and CK20) was performed, and positive staining was observed for 34 $\beta$ E12 (Fig. 2h) and CK5/6 (Fig. 2i).

Whole-body computed tomography (CT) scanning and Ga-67 scintigraphy did not reveal the involvement of MF in the lymph nodes or internal organs, nor any abnormalities suggesting systemic amyloidosis. The results of blood tests for cell counts and chemistry panel were within normal ranges, and M-protein was not detected. We diagnosed this case as LCA associated with poikilodermatous MF. The patient was treated with topical steroid and psoralen plus ultraviolet A (PUVA), and his skin manifestations improved gradually.

#### DISCUSSION

LCA can be observed both primarily and secondarily. Secondary LCA occurs in various inflammatory skin diseases and epithelial skin tumours. The amyloid in the LCA is mainly derived from epidermal keratins. The keratin from damaged keratinocytes drops off to the dermis and is degenerated to form amyloid materials. Amyloid deposits are usually positive with Congo red staining and green birefringence by polarizing microscopy. In polarized light the deposits of our case revealed green birefringence after staining with direct fast scarlet, but the deposits were negative for Congo red staining. This discrepancy between direct fast scarlet and Congo red staining may result from an amount of amyloid deposits, because Congo red staining may be equivocal and inadequate for detecting small deposits of amyloid, such as LCA (1). Chang et al. (2) investigated the cytokeratin profiles in LCA in detail. The amyloid material in secondary LCA positively stained in 34 $\beta$ E12 and CK5 in 100% of cases analysed in the research. The data support the theory that amyloid materials are generated by epidermal damage.

There have been only 2 cases of LCA associated with MF (3, 4). One was the patch stage and the other was the plaque stage of MF, and both cases had pruritus, similar to our case. The former case showed positive immunohistochemical staining for keratin antibody (AE1/AE3 (reacts with CK1-8, 10, 14–16, 19)) on the amyloid deposition (3). In latter case, staining of lymphocytic markers was performed and the infiltrating lymphocytes were positive for CD3 and CD4 (4).



**Fig. 2.** A skin biopsy specimen from the left thigh shows band-like lymphocytic infiltration in the superficial dermis. (a) Amorphous eosinophilic deposits (asterisks) are observed in the superficial dermis. Haematoxylin-eosin stain, original magnification  $\times 40$ . (b) In some areas, focal interface vacuolar changes associated with pigment incontinence are observed. Haematoxylin-eosin stain, original magnification  $\times 200$ . (c–e) Infiltrating lymphocytes are positive for (c) CD3, (d) CD4, and (e) CD8 (original magnification  $\times 40$ ). (f) The amorphous deposits are positive for direct fast scarlet, and (g) polarizing microscopic findings of these deposits reveal green birefringence (original magnification  $\times 100$ ). The amorphous deposits are positive for (h) 34 $\beta$ E12 and (i) CK5/6 (original magnification  $\times 100$ ).

In our case, the clinical and histological features were consistent with poikilodermatous MF. Poikilodermatous MF is a clinical variant of MF that is characterized by earlier age of onset, longer duration of clinical symptoms before diagnosis and predominance of the CD8<sup>+</sup> phenotype (5). Vacuolar degeneration, interface dermatitis and pigment incontinence are commonly seen in poikilodermatous MF. CD8<sup>+</sup> cells are cytotoxic in nature; therefore, the infiltration of the many CD8<sup>+</sup> cells in our case might be attributable to the formation of amyloid material from attacks on epidermal keratinocytes. A possible alternative mechanism of amyloid formation in this case is amyloid formation through prolonged scratching and rubbing, like that of friction amyloidosis. However, the amyloid deposition was seen around the hair follicles, suggesting that the amyloid formation was not solely the result of external stimulus. We performed detailed immunostaining of keratins, and positivity was seen in 34 $\beta$ E12 and CK5/6. This suggests that the mechanism of amyloid generation in MF may be similar to those of the other skin conditions that are

associated with secondary LCA. This is the first report to demonstrate that the amyloid seen in MF lesions is positive for CK5/6.

*The authors declare no conflicts of interest.*

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