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SHORT COMMUNICATION

Annular Atrophic Plaques on the Face in a Father and a Son: Christianson's Disease, a Real Entity?

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Annular atrophic plaques of the face were first reported by Christianson & Mitchell in 1969 (1). Since then, only a few cases of this condition have been described (2–7). Lesions present as slowly progressive non-inflammatory atrophic plaques surrounded by a narrow sclerotic margin. They occur predominantly on the face, but other sun-exposed areas, such as the hands, forearms and lower legs, may also be affected (1, 2, 5–7).

CASE REPORT

A 58-year-old man presented with asymptomatic alopecic plaques on his face and head that had developed slowly and progressed over the last 8 years. Treatment with topical clobetasol had been ineffective. The patient's medical history was unremarkable, except for arterial hypertension.

Clinical examination revealed 10 non-inflammatory annular alopecic plaques sized 1 × 1 to 4 × 4 cm on the forehead, the scalp, the neck, behind both ears, in the right elbow flexure and on the back of both hands, with depressed, atrophic centres and narrow sclerotic bands at the margins (Fig. 1A). Some plaques had linear sclerotic elements within the centre. Laboratory work-up indicated late-stage borreliosis (previously treated with doxycycline), but no other abnormalities. Antinuclear antibodies and antibodies against extractable nuclear antigens and double-strand DNS were negative. Work-up for porphyria was unremarkable.

Haematoxylin-eosin stained skin biopsies showed actinic elastosis, a focal lymphocytic and plasmacellular infiltrate and thickened collagen bundles in the middle and deep dermis (Fig. 2A–C). In elastica van Gieson stain, elastic fibres appeared well preserved and sometimes thickened. Direct immunofluorescence, performed to test for a lupus band, was negative. Ultrastructural analyses revealed large, compact collagen bundles of variable calibre fibrils (Fig. 2D), degenerative elastic fibres in the papillary dermis, and prominent oblong elastic fibres with focal calcification in the reticular dermis.

In 1970, the patient's 62-year-old father had presented to the Free University of Amsterdam with similar lesions on the face, around the ears, in the neck and on the back of both hands (Fig. 1B). Multiple biopsies had shown epidermal atrophy, basal hyperpigmentation and vacuolar degeneration, along with marked solar elastosis, dermal fibrosis, fibrillar degeneration of collagen, atrophy of the hair follicles, mild dermal lymphocytic infiltrate, and arteriosclerotic changes of the blood vessels (2). The father's case had been published by Kalsbeek (2), with a diagnosis of annular atrophic plaques. Other family members, including 3 younger sisters, 3 younger brothers, 1 daughter, 2 sons, and numerous nieces and nephews of our patient were unaffected.

DISCUSSION

Annular atrophic plaques are characterised by a narrow sclerotic band surrounding a depressed, atrophic centre, reminiscent of head skin grafts (1–7). Since the lesions occur on sun-damaged skin, chronic ultraviolet (UV) light exposure was suggested as triggering factor (1, 6, 7). Microscopic findings vary depending on the age of the plaques. Mild epidermal atrophy and a sparse dermal lymphocytic infiltrate are seen in early lesions; and thickened, sclerotic collagen bundles in late lesions (1). All treatment approaches reported so far, including topical and intralesional steroids, diaminodimethylsulfone, doxycycline, and (hydroxy)chloroquine, were ineffec-

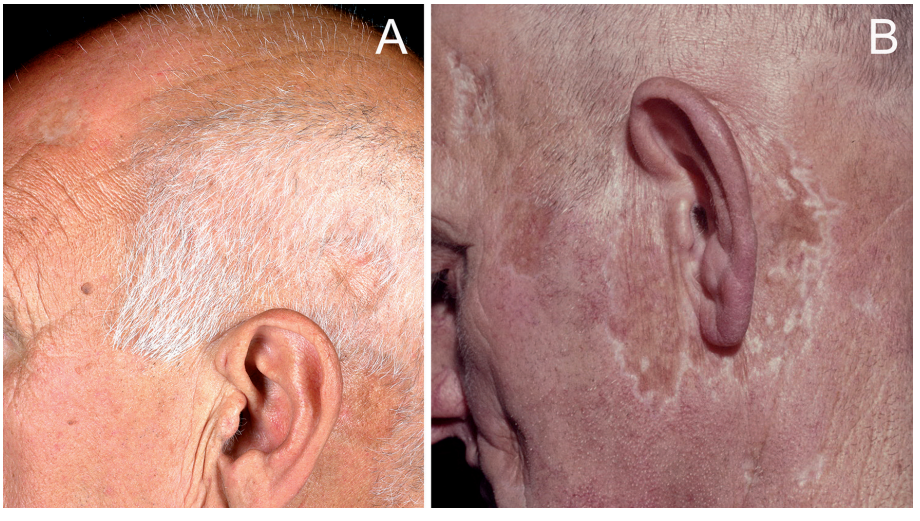

Fig. 1. Clinical features of the patient and his father. (A) The patient presents with non-inflammatory annular atrophic plaques with depressed centre and narrow sclerotic margins on the left forehead and behind the left ear. (B) A photograph of the father from 1970 shows similar lesions in similar locations.

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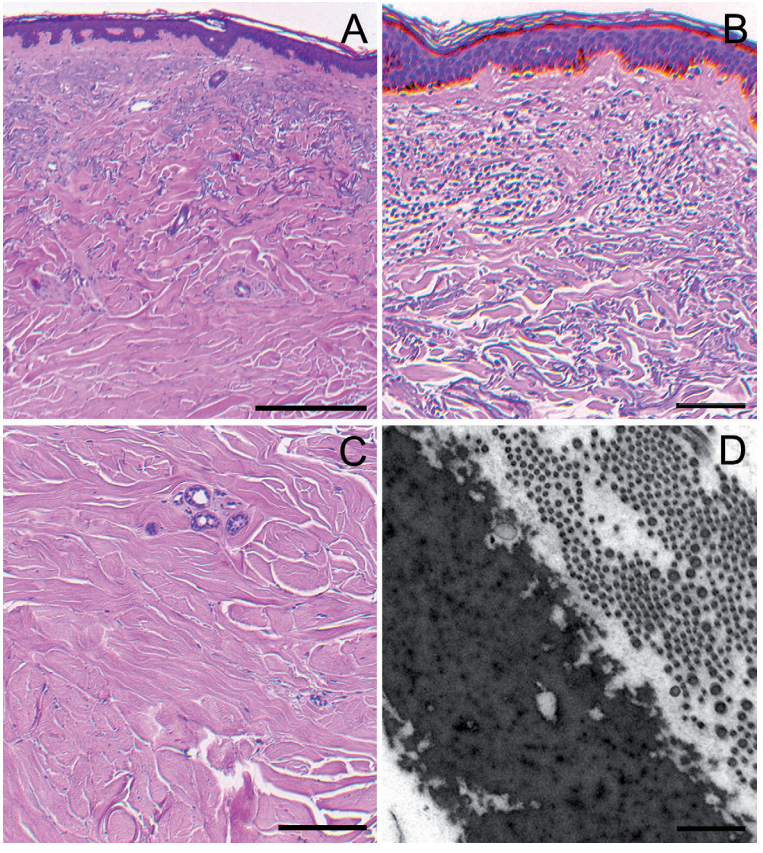

Fig. 2. Microscopic and ultrastructural findings. (A) H&E-stained skin biopsy from the neck showing solar elastosis and dermal fibrosis with thickened collagen bundles. *Bar*: 200 µm. (B) At a higher magnification, basal hyperpigmentation and focal perivascular lymphocytic and plasmacellular infiltrates are seen in the superficial dermis. *Bar*: 50 µm. (C) Detail of the deep dermis, revealing thickened collagen bundles. *Bar*: 50 µm. (D) Electron micrograph presenting a prominent elastic fibre with irregular contours and collagen fibrils with variable diameters in the reticular dermis. *Bar*: 500 nm.

tive (1, 6, 7). Our patient was offered topical tacrolimus off-label, but currently did not desire treatment.

It has been controversially discussed whether annular atrophic plaques represent an entity on its own or an atypical variant of discoid lupus erythematosus (1, 3, 4), lichen sclerosus (1, 5) or morphoea (1), as they share several clinicopathological characteristics with each of these conditions. Features reminiscent of discoid lupus are the location of the plaques in sun-exposed areas, the co-occurrence of atrophy and scarring and several compatible histological findings (1). Features in favour of morphoea are the annular configuration of the plaques, the sclerotic margins and certain histological aspects (1). Features arguing for lichen sclerosus et atrophicus are the white flat appearance of early lesions, which can successively enlarge and coalesce, and histological similarities including epidermal atrophy, focal vacuolar degeneration and upper dermal oedema, followed by hyalinization and pseudosclerosis (1). Christianson & Mitchell (1), who first described annular atrophic plaques, were not convinced that they were dealing with a well-defined clinicopathological entity. They, and most discussants of their paper, alternatively favoured an atypical form of lichen sclerosus, whereas others considered an unusual variant of discoid lupus as the most likely diagnosis (1, 3, 4). Moreover, annular atrophic plaques were discussed as a variant of actinic granuloma, an annular connective tissue disorder of sun-damaged skin characterised by actinic elastosis in the peripheral zone, an inflammatory reaction with foreign-body type giant cells and resorption of elastotic fibres in the marginal zone and, sometimes, mild sclerosis in the centre (8).

It is well known that lupus erythematosus, lichen sclerosus and morphoea can occur with familial accumulation. This is the first case report on annular atrophic plaques in 2 members of a family, suggesting underlying genetic factors; however, this condition might not be a distinct entity.

The authors declare no conflicts of interest.

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