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Actinic Granulomas and Relapsing Polychondritis

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Abstract. A patient developed concomitantly chondritis of the two auricles, diffuse cutaneous vasculitis and actinic granulomas. Alterations in skin and cartilage were prominent in the elastic tissue. Anticollagen type II antibodies were absent from the serum and there was no deposit of immunoreactants in cartilages. In this form of relapsing polychondritis, the pathomechanism resembles that of diffuse actinic arteritis as proposed by O'Brien. It is concluded that relapsing polychondritis may represent a heterogeneous syndrome with regard to its pathogenesis.

Key words: Elastic fibres; Collagen type II; Elastolysis

Immunologic abnormalities are important in the pathogenesis of relapsing polychondritis. Cell-mediated responses to cartilage proteoglycans were reported to be active at the time of disease activity (2, 6). Recent developments in immunopathology have also demonstrated the existence of circulating antibodies to type II collagen and of circulating immune complexes during the acute phase of general involvement (1). In vivo deposits of immunoglobulins and complement in cartilage were sporadically observed.

These results do not prove that some of the described immunological alterations are responsible for the initiation of the disease. They could be related phenomena participating in the generalization of the disease. They could represent a consequence rather than the cause of the initial alteration at one site in the body.

We report here the clinical and histological presentation of a patient who experienced an acute chondritis of both ears in association with actinic granulomas (3, 4, 5) and discrete vasculitis.

CASE REPORT

A 57-year-old man was referred to us for treatment of a widespread erythematous dermatitis associated with papular lesions on the arms and with an inflammatory edema of both ear pinnas. No previous similar dermatitis was reported by the patient, except for edema of the ears. Cartilages other than those of the ears were apparently uninvolved.

Pertinent laboratory data included the following: normal blood cells counts and ESR, fibrinogen slightly increased: 5 to 7 g/l (N: 4), α_2-globulin over 10.5% (N: 9.5%), C1q-binding activity 9.4% ($N = 5.6 \pm 3.8\%$). Joints were found normal at X-ray examination.

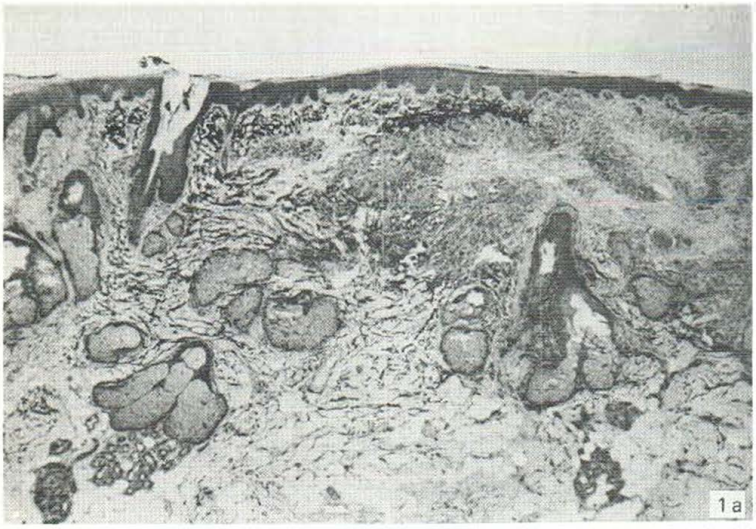
After 3 weeks of topical corticotherapy, papular lesions resolved leaving atrophic macules, and inflammation of the ears faded, though leaving a permanent alteration of the aspect of the pinna.

We studied by optical microscopy five biopsies taken from the ears, face, arms, and buttock. Antibodies to type II collagen were searched by direct and indirect immunoflourescence using the technique of Foidart et al. (1).

RESULTS

A lesion biopsied on the buttock corresponded to a sample of the diffuse erythematous dermatitis. The lesions were interpreted as a lymphocytic vasculitis.

Lesions collected on the arm and on the face corresponded to isolated erythematous papules. Histologically, tiny granulomas were organized around vessels localized inside a thick band of actinic elastosis. Small mononuclear cells and multinucleated giant cells predominated in the infiltrate



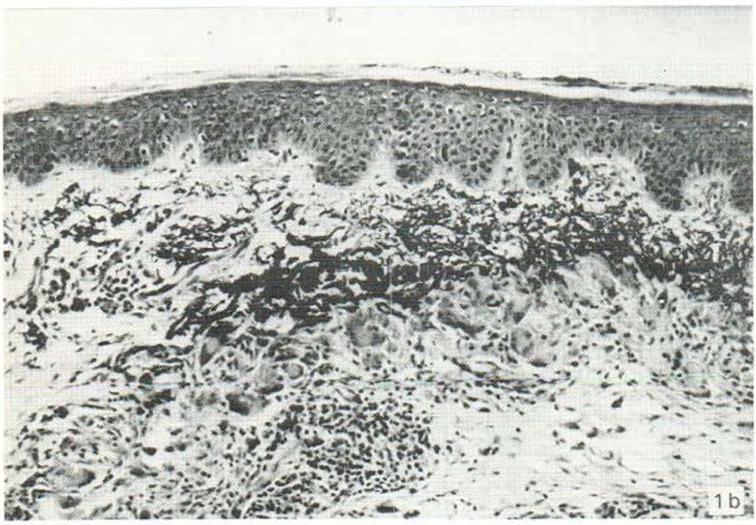


Fig. 1 a, b. Biopsy of the forearm skin. Granuloma disrupting solar elastosis; giant cells are numerous and phagocyte elastotic fibres. Orcein Giemsa.

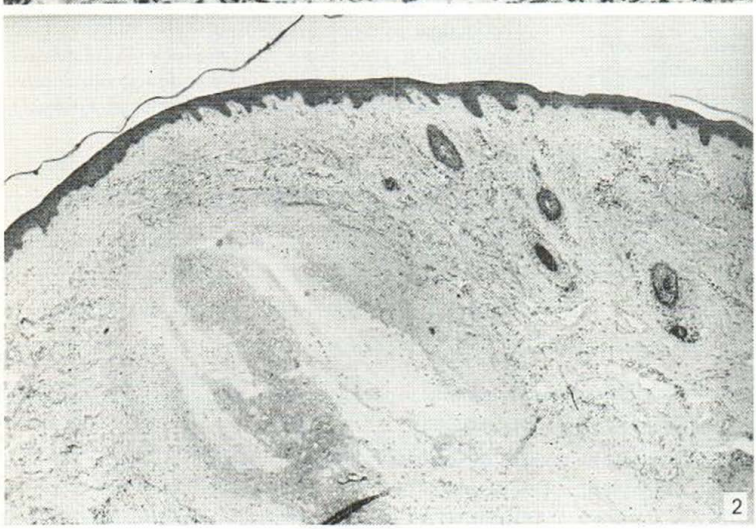


Fig. 2. Biopsy of the ear. Cartilage is partly destroyed and its limits are obscured. In the cartilage, elastic fibres are disrupted and in the skin solar elastosis is remodelled by an inflammatory reaction. A granulation tissue is present in the cartilage. Orcein Giemsa.

(Fig. 1*a*, *b*). The multinucleated histiocytes contained elastotic fibres and asteroid bodies. In the centre of the granuloma, elastotic material could no longer be detected, there was no excess of proteoglycans, collagen bundles appeared normal and fibroblast-like cells were increased in number.

The two biopsies collected from the ears and containing cartilage revealed similar changes. There was a marked solar elastosis disrupted by foci of granulomatous inflammation (Fig. 2). Edema and widely dilated vessels were present underneath. The inflammatory reaction extended to the cartilage whose contour was disrupted. Granulation tissue was replacing the destroyed cartilage but did not extend into the dermis (Fig. 2). At the junction between granulation tissue and normal-looking cartilage, changes variously affected the cartilage components. The elastic fibres were altered, while proteoglycans and collagen were preserved. The epithelial structures of the ear were normal.

Direct and indirect immunofluorescence searching in serum for antibodies to collagen type II was fruitless. Ear cartilage did not reveal any deposits of immunoglobulins or C3.

DISCUSSION

The diagnosis in this patient was and remains a puzzling problem because several conceptual questions are unsolved. We therefore preferred to define it in the form of a descriptive title associating actinic granulomas with relapsing polychondritis.

We observed a lympho-histiocytic vasculitis with formation of granulomas that could fit the spectrum(s) of granuloma annulare, Miescher's granulomatosis disciformis, actinic granulomas and diffuse actinic arteritis (3, 4, 5).

Electromagnetic radiation is probably involved in the pathogenesis of the disease described here. Changes are predominant in light-exposed areas of the body and granulomas are restricted to the elastotic zone. The mechanism of cartilage destruction could therefore be related to a specific immune reaction directed against an element of elastic tissue present in elastotic material and elastic cartilage, and to the release of enzymes from the granulation tissue as in the rheumatoid pannus. The concept of diffuse actinic arteritis proposed by O'Brien (3, 4) to group actinic granuloma, temporal giant cell arteritis and polymyalgia rheumatica could be further extended to include the presently described pathology.

Relapsing polychondritis can be considered as the result of an immune reaction directed against one of the various components of cartilage, namely chondrocytes, proteoglycans, collagen type II, elastic fibres, and other glycoproteins, resulting in an enzymatic breakdown of the tissue mediated by the release of lysosomal enzymes from chondrocytes and histiocytes. The immunological disorder could be primarily directed against one specific component of cartilage and followed by secondary enzymatic breakdown. The reverse condition could also occur, in which the inflammatory disease would be responsible for a local degradation of the cartilage, followed by specific immunization against one of its components and generalization of the disease. Relapsing polychondritis may therefore represent a heterogeneous syndrome with regard to its pathogenesis.

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