Annular Atrophic Lichen planus and Sneddon's Syndrome

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Key Words

Sneddon's syndrome
Annular atrophic lichen planus

Abstract

We report the case of a patient who had 2 rare diseases, annular atrophic lichen planus (AALP) and Sneddon's syndrome (SNS). This patient had also digital nodules with histological abnormalities suggestive of SNS vasculopathy, which have not been reported so far. AALP is the most rare of all varieties of lichen planus since this case is the third reported to date. The association of livedo racemosa and cerebrovascular disease is the hallmark of SNS, the incidence of which is estimated to be 4 cases per year per million inhabitants. In both diseases, an abnormal production of elastic-tissue-degrading enzymes or a constitutional abnormality of the elastic tissue can be postulated, since SNS is characterized by arteriolar changes with deterioration of the internal elastic lamina and AALP by destruction of the dermal elastic tissue.

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anti-endothelial-cell antibodies, anti- β_2 -glycoprotein-I antibodies, protein C, S, AT III, homocysteine, exploration of fibrinolysis, resistance to activated protein C, Ham-Dacie and sucrose tests and lipoprotein(a) level.

Annular atrophic lichen planus (AALP) has been reported so far only in 2 cases [1,2]. We report an additional patient who associated Sneddon's syndrome (SNS). Such an association may cast some light on the patho-physiology of AALP.

Case Report

A 41-year-old male with a 4-year history of vertigo and with a cerebral CT showing a round hypodensity on the left internal capsule developed annular pruritic lesions on the trunk, livedo and Raynaud's phenomenon. One year later, he had an ischaemic stroke with hemiparesis and dysarthria.

On examination, he exhibited about 20 annular lesions (1-6 cm wide) which were located on the trunk, back and thighs. The lesions had an erythematous papular border and an atrophic centre (fig. 1). A racemosa-type, non-infiltrated, livedo was observed on the trunk, limbs and buttocks. He disclosed also erythematous nodules on the lateral side of some fingers.

Laboratory findings were negative or normal including antinuclear antibodies, rheumatoid factors, cryoproteins, complement values, serum immuno-electrophoresis, lupus anticoagulant, anticardiolipin antibodies,

Fig. 1. Annular lesion with an infiltrated border and an atrophic centre.

There was no α_1 -antitrypsin or α_2 -macroglobulin deficit. Echocardiography was normal.

A biopsy of the annular lesions revealed a lichenoid infiltrate in the border and a com-

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plete disappearance of the elastic fibers in the centre. Biopsies of the livedo, performed both on and between the livid patches, showed no significant abnormality. Examination of a digital nodule showed the typical features of obliterative endo-arteritis.

The patient was given 100 mg acetylsalicylic acid daily and the cerebrovascular diseases had no further episodes within the 2 years of follow-up. Topical steroids had no effect on skin lesions.

Discussion

Our patient had 2 rare diseases, AALP and SNS. AALP is the most unusual variant of lichen planus and is characterized by small violaceous papules slowly enlarging peripherally. The centre becomes atrophic and hypopigmented, while the borders are raised and covered sometimes by Wickham's striae. The plaques can reach up to 10 cm in their largest diameter and may be pruritic [2]. AALP involves the same sites as the common lichen planus, but the trunk, neck and thighs can also bear lesions. The course is very chronic, lasting more than 20 years in the 2 described patients.

While histopathology of the borders shows typical lichen planus features, the atrophic centre reveals a flattened epidermis with loss of rete ridges and numerous thick-walled blood vessels in the papillary dermis. In both sites, the elastic fibres are destroyed in the papillary dermis, both with and without a lymphocytic infiltrate. By electron microscopy, the elastic fibres appear fragmented as in cutis laxa [1]. Topical steroids [1,2] and UV light [2] have been ineffective so far.

SNS includes a racemosa-type livedo and cerebrovascular ischaemic events [3,4]. Digital nodules have not been reported so far and histopathological features of obliterative endoarteritis are rarely observed. Zelger et al. [5] described them in biopsies of livedo, and similar findings have been reported in the antiphospholipid syndrome without SNS [6, 7].

Antiphospholipid antibodies have been found in up to 85% of cases [6], but they seem not to affect the clinical and biological features of the disorder, except for epilepsy and thrombocytopenia which are more frequent in positive patients [8, 9].

The incidence of SNS is estimated to be 4 new cases per million inhabitants per year [3], and our patient is the third to be reported as having AALP. The simultaneous occurrence of 2 extremely rare conditions in the same patient is very unlikely to be coincidental. Furthermore, the arteriolar damage observed in SNS and the destruction of the elastic fibres in AALP recall the cases of anetoderma observed in the antiphospholipid syndrome [10]. Both features might result from a similar pathomechanism, an abnormal production of elastolytic enzymes, a defective protection against them or a constitutional abnormality of the elastic tissue.

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