Annular Atrophic Lichen planus and Sneddon’s Syndrome

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Key Words
Sneddon’s syndrome
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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-ß,-gly-coprotein-I antibodies, protein C, S, AT III, homocysteine, exploration of fibrinolysis, resistance to activated protein C, Ham-Dacie and sucrose tests and lipopotein(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,-antitrypsin or ou-macro-globulin deficit. Echocardiography was normal.
A biopsy of the annular lesions revealed a lichenoid infiltrate in the border and a com-
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 acetylsali-cylic 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, A ALP 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 hy-perpigmented, while the borders are raised and covered
sometimes by Wickam’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 commonplace
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 en-doarteritis
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.
References
392-394.


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