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SNEDDON'S SYNDROME

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ABSTRACT

Sneddon's syndrome is usually characterized by the association of an ischemic cerebrovascular disease and a widespread livedo reticularis. The incidence of Sneddon syndrome is 4/1000 000. We present 42-year-old woman with livedo reticularis, recurrence ischaemic cerebrovascular accidents, two repetitive miscarriages and positive anti-2GPI antibodies. Skin biopsy specimens reveal inflammatory changes of small- to medium-sized arteries and subendothelial proliferation and fibrosis. The diagnosis Sneddon syndrome is confirmed by skin biopsy, and MR evidence.

We suggest that anti-2GPI antibodies may be pathophysiologically related to the clinical manifestation observed in some patients with Sneddon syndrome.

Key words: Sneddon syndrome, livedo reticularis, ischemic cerebrovascular disease.

INTRODUCTION

Sneddon's syndrome is characterized by livedo reticularis associated with cerebrovascular disease. The condition affects small to medium sized arteries of the dermis-subcutis border. The process begins as an endothelial inflammation followed by occlusion, fibrosis and atrophy of the vessels. Antiphospholipid antibodies were found in some patients with Sneddon's syndrome. The disorder has a slow and progressive clinical course. No effective drug therapy is available.

CASE REPORT

We report a case of Sneddon's syndrome in a 42 year-old woman. She suffered from cerebrovascular accident presented by double vision and lost of consciousness 5 years previously. Because of that she has been treated in the Department of Neurology. In May 2007 a recurrence of her neurological complaints occurred, expressed by dizziness, vomiting, ataxic walking, numbness and muscular debility of the left lower extremity. From medical history she had myocardial infarction three years ago and two repetitive miscarriages. The patient has been suffering from high blood

pressure and migraine for 10 years. She has been having persistent cutaneous lesions on the upper and lower extremities and trunk for the last 20 years.

At physical examination, a slight elevation in pressure levels (150 x 80mm Hg), III degree obesity and slight edema of the lower limbs were found. The neurological examination revealed ataxic walk. Romberg reflex was negative (-) and Babinski was positive (+) in right. The ophthalmologic examination demonstrated an initial angiosclerosis. The dermatological examination showed erythematous violaceous lesions with a reticular pattern, localized in the arms, trunk (Figure 1) thighs and knees (Figure 2, Figure 3). The following exams in the laboratorial evaluation were normal or negative: blood count (including platelet count), glycemia, sodium, potassium, urea, creatinine, creatinine clearance, transaminases, alkaline phosphatase, bilirubin, amylase, cholesterol, triglycerides, PT (prothrombin time), VDRL (venereal disease research laboratory) urinary sediment and anticardiolipin antibodies. Anti- α 2GPI antibody was positive (+).

Regarding the image exams abdomen ultrasound scan was normal. Computerized tomography (CT) of the skull revealed pallium atrophy. The Magnetic nuclear resonance (MNR) of the brain demonstrated old and chronic ischemic areas (Figure 4.1; Figure 4.2). The electrocardiogram revealed evidence for a postinfarction cicatrix.

The immunohistochemical examination with CD 34 of two skin biopsies from the livedo lesions revealed endothelial proliferation and obliteration of the small and middle arteries (Figures 5, 6). The diagnosis of Sneddon syndrome in our patient was made on the basis of anamnesis, clinical features, skin biopsy and MNR. Therapeutics was instituted with 100 mg aspirin (acetylsalicylic acid) once a day. The systemic arterial hypertension was controlled.

DISCUSSION

In 1965, Ian Sneddon was the first author to establish a relationship between livedo reticularis and vascular neurological manifestations in six patients, thereby describing the syndrome that today takes his name (6, 8).