The antiphospholipid syndrome: a syndrome in evolution

Sirs: I read with interest the leader by Asherson and Cervera1 and agree that disease conditions other than deep vein thrombosis, recurrent fetal loss, or stroke may occur, not only in association with antiphospholipid syndrome: lupus erythematosus (SLE) or ‘lupus-like’ disease, but also with ‘primary’ antiphospholipid syndrome (APS). The authors included Sneddon’s syndrome (cerebrovascular disease with livedo reticularis) in the category of ‘primary’ APS.

Sneddon’s syndrome comprises about 0.26% of total cerebrovascular cases. Clearly, the wide range of testing to detect antiphospholipid antibodies in Sneddon’s original patients was undertaken. However, pathogenesis of Sneddon’s syndrome remains unclear. Sneddon suggested that skin biopsies from future cases would show endarteritis obliterans in dermal arteries. It was thought that intravascular pathology of a similar nature was responsible for cerebrovascular accidents. There is now a large number of cases of cerebrovascular disease with negative tests for antiphospholipid antibodies. Thus, it is possible that some categories of CNS disease may be because of anticoagulation and that these cases belong to the category of ‘primary’ APS, with or without SLE.

A case of Sneddon’s syndrome in association with deep vein thrombosis and pulmonary embolism is presented here; repeated search for antiphospholipid antibodies and lupus anticoagulant proved negative.

In October 1990 a 45-year old man presented with a left hemiparesis, a history of left sided hemiparesis (MRC grade 4) and hemianesthesia. He had had deep vein thrombosis and pulmonary embolism in 1971. A chronic ulcer appeared on the front of his right leg in 1974. Apart from neurological signs, he had left deep vein thrombosis, left sided pleural rub, and a chronic ‘venous’ ulcer on the front of his right leg with surrounding pigmentation. Additionally, he had acrocyanosis of his toes and both dorsalis pedis pulsations were absent. No skin rash or nodules were present. He was in sinus rhythm and normotensive, with no heart murmur or carotid bruit. His laboratory profile was normal.

Laboratory investigations showed an erythrocyte sedimentation rate of 70 mm/h (normal < 10), plasma viscosity 2.08 mPa.s at 25°C (normal ±0.05), C reactive protein 16 mg/l (normal < 5), antiphospholipid antibodies positive at a titre of > 1/1024 (homogeneous pattern), with DNA binding of 13% (normal < 5). Chest radiography showed peripheral consolidation in the left mid-zone, and a venogram of his left leg showed extensive thrombus in the deep venous system. A computed tomographic scan of his brain showed a large wedge-shaped, well defined, hypodense lesion in the parietal lobe.

Results of the following investigations were either normal or negative: haemoglobin, white cell count, platelets, packed cell volume, automated serum chemistry, including cholesterols and triglycerides, creatinin and blood urea nitrogen, liver function tests, prothrombin time, kaolin cephalin coagulation time; plasminogen, protein S, protein C, and antithrombin III levels; lupus anticoagulant, IgG and IgM antiphospholipid antibodies (anticardiolipin or lupus anticoagulant); lupus anti-DNA test, direct Coombs’ test, hepatitis B surface antibody, and protein S, C, and antithrombin III by ELISA.

About two weeks after his initial presentation the patient developed generalised livedo reticu- laris, which continued to increase, and Eighteen months later he still has left sided hemiparesis (MRC grade 4) and hemianesthesia. His skin rash persists. Multiple biopsy specimens of affected skin show essentially normal histology. The patient’s most recent erythrocyte sedimentation rate is normal, but his antinuclear factor remains positive with a low titer of 1/10 (homogeneous pattern). Double stranded DNA antibody is, however, negative. Repeat testing for anticardiolipin antibodies failed to show any abnormalities. In view of his propensity to recurrent thromboembolism, long term anticoagulation treatment with warfarin has been continued.

This case further illustrates that Sneddon’s syndrome may also exist as a distinct entity whose pathogenesis remains to be elucidated.

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Anticardiolipin antibodies and renovascular hypertension

Sirs: Two cases of renovascular hypertension associated with antiphospholipid syndrome have been reported recently, and1 we report a case of renovascular hypertension, without angiographic appearance of thrombus, associated with laboratory features found in primary antiphospholipid syndrome.

A 30 year old woman was admitted to our clinic with hypertension and episodes of fatigue in the previous six months. A systolic blood pressure of 14 a positive Venereal Disease Research Laboratory (VDRL) test with negative fluorescent treponemal antibody (FTA) and a positive antinuclear antibodies (ANA) test. A biopsy of the left renal artery was performed and a section of the renal artery wall was obtained. Histological examination showed a fibromuscular lesion with a thickened intima and subintimal fibrosis. The renal artery was occluded by a fibromuscular lesion and there was a reduction in the diameter of the renal artery. The renal artery was occluded by a fibromuscular lesion and there was a reduction in the diameter of the renal artery. The renal artery was occluded by a fibromuscular lesion and there was a reduction in the diameter of the renal artery.

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