About two weeks after his initial presentation the patient developed generalised livedo reticularis, a new symptom. 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 proneness 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

SIR: Two cases of renovascular hypertension associated with anticardiolipin syndrome have been reported recently.1,2 We report a case of renovascular hypertension, without angiographic appearance of thrombosis, associated with laboratory features found in primary antiphospholipid syndrome.

A 30 year old woman was admitted to our clinic with hypertension, proteinuria, and anemia. Urine microscopy showed erythroblasts, leucocytes, and casts. Urinary protein excretion was 0.14 g/day (a positive Venereal Disease Research Laboratory (VDRL) test with negative fluorescent treponemal antibody test (FTA) and microhemagglutination assay (MHA) test). The diagnosis of Fabry's disease was considered and screening for Fabry's disease was performed. There was no clinical evidence for progressive systemic fibrosis. Aortography was normal. For the renal failure and hypertension diuretics and angiotensin converting enzyme inhibitors were used.

The patient's blood pressure was 200/130 mmHg and left leg and right ankle edema were present. Echocardiography showed normal cardiac size and function with a normal left ventricular ejection fraction. Renal biopsy showed non-immunologic interstitial nephritis with interstitial fibrosis. At biopsy, glomeruli were normal. Renal function and proteinuria did not improve despite the use of angiotensin converting enzyme inhibitors. Endothelin has been postulated to be an important renal vasoconstrictor.3,4 We detected increased plasma and urine levels of endothelin-1 in this patient.

The patient was treated with a combination of angiotensin converting enzyme inhibitors, angiotensin receptor blockers, and alpha blocking agents and the blood pressure was controlled. The combination of alpha blocker was stopped after six weeks and the blood pressure control was maintained with angiotensin converting enzyme inhibitors alone. The patient's proteinuria normalized after the use of angiotensin converting enzyme inhibitors alone.

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The antiphospholipid syndrome: a syndrome in evolution.

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**Notes**

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