Letter to the Editor

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Malignant Hypertension and Antiphospholipid Syndrome

A.E. Sirvent
R. Enriquez
A. Antolin
J.B. Cabezuelo
C. Gonzalez
M.D. Arenas

Nephrology Section, General Hospital, Elche, Spain

Dear Sir,

The kidney is now recognized as another target organ in the antiphospholipid syndrome (AS) [1]. Renal involvement is consistent with thrombosis of the major renal vessels and noninflammatory microvascular disease [2-4]. In the clinical findings renal failure has been emphasized over systemic hypertension.

We report here on a patient with systemic malignant hypertension and thrombotic microangiopathy (TM) with no other occlusive complications.

A 29-year-old man was hospitalized with acute renal failure during a malignant hypertensive episode, after developing hypertension in the 3 months prior to admission. Thrombocytopenia was also detected. Physical examination showed: blood pressure 230/130 mm Hg, grade IV retinal changes, no abdominal bruits were heard; the signs were compatible with congestive heart failure. The most significant laboratory findings were: hemoglobin 10g/dl, WBC 14,000/µl (normal differential count), platelets 47,000/µl, normal coagulation tests, fibrinogen 490 mg/dl and schistocytes on a peripheral blood smear. Urea was 35 mmol/l, creatinine 884 µmol/l, LDH 971 U/l, haptoglobin 11 mg/dl, and urine contained proteinuria 2.1 g/day, 10-15 erythrocytes/hpf. Chest X-ray revealed pulmonary edema and cardiomegaly, and echo-cardiography indicated left ventricular hypertrophy, normal ejection fraction and mild mitral regurgitation. The abdominal ultrasound showed a right kidney of 11.9 cm and a left kidney of 8.7 cm. No evidence of kidney infarction was seen in the renal CT scan. The magnetic resonance angiogram disclosed normal aorta and renal arteries.

The following laboratory data were negative or normal: urine cultures, urine catecholamines and VMA, HBsAg, serology for HCV, cryoglobulins, rheumatoid factor, ANCA, anti-GBM, ANA, anti-DNA, anti-SSA (Ro), anti-Sm, anti-RNP antibodies, C3, C4, VDRL, brain CT, cystography, lupus anticoagulant; anticardiolipin antibodies (aCL) IgG 63.3 GPL/ml (n.v. < 23), IgM 3.1 MPL/ml (n.v. < 11), and repeated after 8 weeks, IgG 40 GPL/ml (ELISA). Antiplatelet antibodies were positive.

The kidney biopsy showed TM; no deposits were noted in the immunofluorescence study. Blood pressure was controlled. An increase in platelets and normalization of LDH was achieved following plasma exchange with FFP as the replacement fluid; upon withdrawal of the plasma exchange, thrombocytopenia recurred. After receiving the aCL determination
results, aspirin (125 mg/day) was prescribed, and the platelet count returned to normal. Meanwhile, there was no improvement in renal function, and the patient was discharged on maintenance hemodialysis. Nine months later, the ANA was 1/640, anti-DNA antibodies negative, aCL levels remained elevated but stable, and there were no new symptoms. This patient could be considered as having AS. He did not meet ARA criteria for systemic lupus erythematosus, and the kidney biopsy did not reveal proliferative changes or immune deposits, thereby excluding an immune-complex glomerulonephritis. The last ANA titer suggests that the most likely diagnosis is lupus-like disease with AS [5].

The renal clinical manifestations of AS are generally proteinuria, acute or chronic renal failure, and arterial hypertension. In some series arterial hypertension is as common as renal failure [2, 3], and its severity varies from mild to accelerated-malignant.

Malignant systemic hypertension in AS is usually related to thrombosis or stenosis of the renal artery, to underlying lupus nephropathy or to the catastrophic form of AS [6, 7]. Malignant hypertension appears less frequently in TM [2, 8-10]. In these cases, in which there are usually other extrarenal thrombotic complications, the pathogenic factor of malignant hypertension might be renal ischemia. Other agents, such as oral contraceptives, can play an additional role [8]. In this case, secondary causes of arterial hypertension were discarded. Renal infarction was not found on the CT scan, and the renal arteries and abdominal aorta were normal on the magnetic resonance angiogram. Therefore, it is likely that this patient’s malignant hypertension was due to TM. However, strictly speaking, we cannot discard the fact that the aCL were the result of vascular wall injury, which was the consequence of malignant essential hypertension. In any case, the good response of thrombopenia to aspirin and the elevated ANA titer seem to suggest some pathogenic role for aCL and the existence of an immunological cause in this patient.

The optimal treatment for patients with AS and TM, especially if there is no other extrarenal thrombosis, is still disputed. Some authors use only antiplatelet agents [11]. In this case, after plasma exchange, we used aspirin with regular monitoring of aCL levels and coagulation parameters, and have had no complications up to the present time. However, some observers, based on recent data [12] and given that TM is a kind of microthrombotic phenomenon may favor anticoagulant therapy. In any case, the therapeutic decision might take into account the clinical form in which AS presents.

KARGH
E-Mail karger@karger.ch Fax + 41 61 306 12 34
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