Acute Renal Failure as First Clinical Manifestation of the Primary Antiphospholipid Syndrome

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Dear Sir,

The primary antiphospholipid syndrome (PAS) is an entity defined by thrombotic phenomena and antiphospholipid antibodies in the absence of an underlying cause [1]. Renal involvement has scarcely been covered in the literature as compared to other manifestations. We describe 1 case of this syndrome which was explosively introduced as thrombotic microangiopathy and acute renal failure.

A 33-year-old woman was admitted with oliguria renal insufficiency. Family history was of no interest. In the personal history we must point out 2 pregnancies which were carried to the end, with no fetal loss or drug intake. In the physical examination we detected: blood pressure 180/100 mm Hg, P 37.6°C, jugular veins distended, tachycardia with an S3 sound, livedo reticularis on the legs and ankle edema. Funduscopy: exudates and retinal hemorrhage in the right eye. Initial complementary values: leukocytes 19,800/ μ l (88% neutrophils), hematocrit 26%, hemoglobin 8.8 g/dl, ESR 109, reticulocytes 5%, the platelet count was 66,000/ μ l, prothrombin time 19 s, partial thromboplastin time 64 s (with a control of 39), fibrinogen 649 mg/dl, fibrin degradation products 0.5 mg/l. The peripheral blood smear showed few schisto-cytes, the monospecific direct Coombs was negative. Urea was 33 mmol/l, creatinine 540 μ mol/l, LDH 2,099 U/l, haptoglobin 528 mg/dl. Urine: proteinuria 500-700 mg/day, 4-5 erythrocytes/hpf. Chest x-ray showed bilateral alveolar edema. ECG: sinus tachycardia. Echocardiography revealed no abnormalities of the heart valves. Normal kidneys on sonography.

The hemogram showed a decrease of the hematocrit (21%) and platelet count (12,000/ μ l). The clinical picture was interpreted as acute renal failure due to thrombotic angiopathy. Eight plasma exchanges (40 ml/kg each one) were carried out with fresh frozen plasma as replacement fluid. The evolution was satisfactory with an increase in hematocrit and platelet, normalization of the LDH, recovery of diuresis and improvement of the renal function. The following tests were negative or normal: blood and urine cultures, gastroesoph-ageal study, barium enema, abdominal CT, bone marrow examination, HBsAg, cryoglobulins, rheumatoid factor, ANCA, anti-GBM, ANA, anti-DNA, anti-SSA, anti-SSB, anti-Sm and

anti-RNP antibodies. C3 was 74 mg/dl, C4 11 mg/dl (normal 12-54 mg/dl). CT brain scan disclosed a right frontal infarct. VDRL was positive with negative antitreponemal antibodies. Lupus anticoagulant was detected by the test of Exner. Anticardiolipin antibodies (ELISA) in a first determination were: IgG 29.7 GPL/ml (n.v. < 23); repeated after 8 weeks: IgG 50.2 GPL/ml and IgM 15.1 MPL/ml (n.v. < 11). A renal biopsy was carried out 1 month after her admission and it identified ischemic lesions with collapse of the glomerular tufts; the capillary walls were thickening with double contour images, microthrombi and fragmented erythrocytes were occasionally observed in the glomerular capillary lumens (fig. 1); there was a heavy interstitial lymphoplasmocytic infiltrate and the medium-size arteries showed subintimal proliferation; immunofluorescence was positive for fibrinogen at the glomerular level, no deposits of immunoglobulin or complement were seen. We started treatment with aspirin (125 mg/day) and prednisone (1 mg/kg/day) with stabilization of creatinine clearance at 35-40 ml/min and normalization of partial thromboplastin time.

Our patient was admitted with a clinical picture of acute renal failure due to thrombotic microangiopathy, livedo reticularis and a brain infarct. Anticardiolipin antibodies were positive in two determinations. We reasonably discarded a base process, and for this reason it can be labelled as a PAS [2]. The renal pathology in the PAS has vascular predominance, where arterial and glomerular thrombosis with or without necrosis, thrombosis of the renal vein, stenosis of the renal artery, cortical infarcts and renal ischemia stand out [3-7]. The insidious loss of the renal function after chronic ischemic lesions without inflammatory vascular pathology has also been described [8]. It is probable that the renal involvement in the PAS represents a continuous spectrum which would go from the slow and progressive deterioration of the renal function to acute clinical pictures such as the one we have described. For this reason the nephropathy must progressively achieve an important position in the clinical chapter of PAS.

The response to plasma exchange in our patient was good. Although the role of plasma exchange in the PAS is not well defined, it seems reasonable to use when it manifests itself as a thrombotic microangiopathy [3]. The controversies with respect to the long-term treatment of the PAS are many;

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Fig. 1. Light microscopy photomicrograph of a glomerulus demonstrating thickening of capillary walls and fragmented red blood cells in glomerular capillary lumens. HE. × 400.

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from the use of antiplatelet agents or anticoagulants alone, to the association with steroids or immunosuppressors [9, 10]. Perhaps, the therapeutic decision depends on the way and severity with which it presents itself.

As in other autoimmune diseases, it is difficult to evaluate the degree of activity of the PAS by basing exclusively on the antibodies. In some cases partial thromboplastin time could be a useful parameter.

In summary, plasma exchange can be a good therapeutical option in thrombotic microangiopathy due to PAS, and we must also include this syndrome in the differential diagnosis of renal thrombotic microangiopathy.

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