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Catastrophic antiphospholipid syndrome presenting with renal thrombotic microangiopathy and diffuse proliferative glomerulonephritis

Sirs,

The catastrophic variant of the antiphospholipid syndrome (APS) is an unusual but often lethal form of presentation of this syndrome characterized by a rapid development of multiorganic failure, mainly due to thrombotic microangiopathy in several organs (1). Since the early description of the catastrophic APS (1), more than 300 cases have been collected, being the kidney one of the more commonly affected organs (70%) (2). However, there are no previous reports of the simultaneous presence of diffuse proliferative lupus glomerulonephritis and renal thrombotic microangiopathy (TMA) as the first manifestation of catastrophic APS.

A 29-year-old Caucasian man was admitted at the Emergency Department in June 2004 due to the appearance of generalized oedema in the last 4 weeks accompanied by decrease in urine output. He had been diagnosed as having systemic lupus erythematosus (SLE) in 2002 due to a history of Evans' syndrome, recurrent leg ulcers, presence of antinuclear antibodies (ANA) (1/160), anti dsDNA antibodies (42 U/mL [normal < 7 U/mL]), and lupus anticoagulant (LA), and was on treatment with aspirin alone at the time of admission. Physical examination revealed marked *livedo reticularis* in the lower extremities and a generalized oedema. During the first hours of admission, he presented seizures with a cerebral computed tomography (CT) scan that showed a cortico-subcortical ischaemic lesion and a lacunar infarct in the right semioval region. Transthoracic echocardiography disclosed severe decrease in left ventricular ejection fraction (LVEF) (35%), mild aortic and mitral regurgitation, and a moderate pericardial effusion. Laboratory tests at admission showed microangiopathic haemolytic anaemia (Hb 8.5 g/dL) with schistocytes, elevated serum creatinine (5.6 mg/dL), and prolonged activated partial thromboplastin time. LA was positive, whilst IgG and IgM anticardiolipin antibodies (aCL) were negative. Anti ds-DNA antibodies were positive (> 200 U/mL) and C3, C4 and CH50 complement levels were low. He was admitted at the Intensive Care Unit (ICU) where i.v. methylprednisolone (1 g per day for 5 days) and i.v. cyclophosphamide (1,250 mg) were started. One week later, percutaneous renal biopsy was performed disclosing the presence of diffuse proliferative lupus glomerulonephritis and TMA (Figure 1). A diagnosis of definite catastrophic APS was made (3) and anticoagulation and plasma exchange (PE) sessions were started. One month later, he was discharged of the ICU because of progressive improvement of his clinical condition, including the heart involvement (LVEF > 60%). However, 4 months later, he was admitted again because of fulminant hepatic failure. The patient's clinical condition progressively deteriorated in the following days and died due to multiorgan failure. Autopsy showed multiple liver infarcts, inferior vena cava thrombosis (6.0 x 0.4 cm) and signs of bilateral pneumonia, as well as persistence of the renal TMA previously described.

In the present case, a "double" renal injury was produced probably due to an immune-complex glomerular deposition (SLE nephritis) and an ischaemic glomerular damage (TMA induced by APS) and this was the first clinical manifestation of a catastrophic APS in a patient with SLE, a combination that has not been previously described. Although there are few reports describing the simultaneous presence of proliferative glomerulonephritis and renal TMA in SLE patients (4-7), none of them fulfil the recently proposed criteria for the classification of definite catastrophic APS (3).

This variant of the APS is a life-threatening condition with an elevated mortality rate (around 50%) that requires high clinical awareness. Therefore, it is essential that it should be diagnosed early and treated aggressively. The combination of high doses of heparin plus steroids plus PE and/or intravenous gammaglobulins is the treatment of choice in patients with catastrophic APS (2).

J.A. GÓMEZ-PUERTA
E. SALGADO
R. CERVERA
S. AGUILÓ
M. RAMOS-CASALS
M. SOLER[1]
A. TORRAS[2]
J. FONT

Department of Autoimmune Diseases, Institut Clínic de Medicina i Dermatologia; [1]Department of Pathology, Centre de Diagnòstic Biomèdic Clínic, [2]Department of Nephrology, Institut Clínic de Nefrologia i Urologia, Hospital Clínic, Barcelona, Catalonia, Spain.
Address correspondence to: Ricard Cervera, MD, PhD, FRCP, Servei de Malalties Autoimmunes, Hospital Clínic, Villarroel 170, 08036-Barcelona, Catalonia, Spain.
E-mail: rcervera@clinic.ub.es

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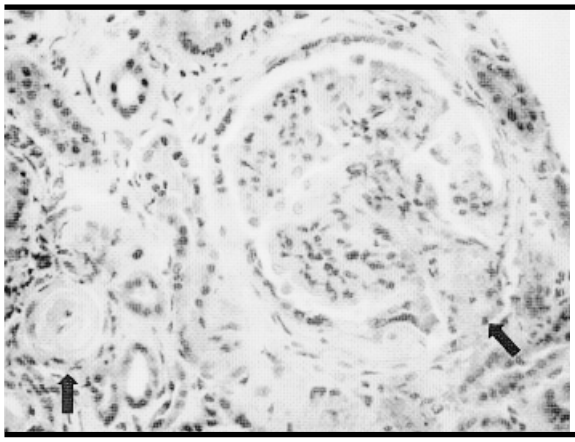

Fig. 1. Percutaneous renal biopsy specimen showing prominent diffuse endocapillar hypercelularity (type IV [WHO classification] lupus glomerulonephritis). Luminar thrombi can be seen in arterioles (**arrows**), small arteries, arterioles and glomerular capillaries. (Hematoxylin & eosin, original magnification x 400).