Case Report

Acute renal failure due to bilateral renal artery thrombosis associated with primary antiphospholipid syndrome

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Introduction

The antiphospholipid antibody syndrome (APS) is characterized by antibodies directed against either phospholipids or plasma proteins bound to anionic phospholipids. Three types of antiphospholipid antibodies have been characterized: anticardiolipin antibodies (ACLAs), lupus anticoagulant and false positive serologic test for syphilis.

The APS can lead to a variety of clinical manifestations, including venous and arterial thromboses, recurrent spontaneous miscarriages and thrombocytopenia. Other possible findings include livedo reticularis, migraine, Raynaud’s disease, haemolytic anaemia, neurologic dysfunction, renal disease, pulmonary hypertension, etc. [1,2].

This disorder is referred to as the primary APS when it occurs as a single entity, however, it can also be found in association with systemic lupus erythematosus (SLE) and other autoimmune diseases [3].

We report a patient with bilateral renal artery thrombosis due to the primary APS and rapidly progressive renal failure successfully resolved by interventional dissolution of the thrombi and subsequent long-term anticoagulation.

Case

A 34-year-old female had no history of abortion and two uncomplicated pregnancies with spontaneous deliveries. Subsequently she took hormonal contraception for 10 years. In 1996 she was repeatedly admitted to a local hospital for erythema nodosum, arterial hypertension and a high erythrocyte sedimentation rate. She had false positive serology for syphilis at that time.

In January 1997 she complained of increased fatigue, lumbar pain, blurred vision and progressive dyspnea. Arterial hypertension, and left ventricle dilatation with pleural exudate on the right side was disclosed by chest X-ray and confirmed by echocardiography. Ophthalmoscopy demonstrated hypertensive neuroretinopathy. The patient was treated with corticosteroids and anti-hypertensives. The dyspnea disappeared and systemic hypertension subsequently improved. However, she developed progressive nonoliguric renal failure and was transferred to our hospital. Laboratory examination disclosed high levels of serum creatinine (580 μmol/l), urea (49 mmol/l), anaemia (haemoglobin 108 g/l), increased white cell count (16 × 10⁹/ml), but normal platelet count (149 × 10⁹/ml).

Elevated ACLA were of the IgG isotype (10.6, normal up to 2.8, the number indicates multiples of standard deviations above the mean of normals). Titres of the IgM isotype were normal (1.4). Other antibodies (antinuclear antibodies, antidualle stranded DNA, extractable nuclear antigen, antineutrophil cytoplasmatic autoantibody) and lupus anticoagulant were negative. By ultrasonography the kidneys were of normal density and size without any signs of urinary obstruction. However, renal arteriography disclosed complete amputation of both renal arteries due to obliterating bilateral thromboses. Only an accessory branch to the lower pole of the left kidney was patent. This vessel was derived directly from the aorta (Figure 1).

Intervention

A diagnostic catheter was successfully placed through the thrombosed right renal artery to the kidney hilus. The main renal artery and some peripheral renal arteries were occluded by thrombi. Percutaneous angioplasty (PTA) of the main renal artery was performed in combination with thrombolysis (direct infusion into the thrombus of 20 mg of rt-PA over 15 min). Ostial dissection was seen immediately after PTA and a
Palmaz stent (dilated to 7 mm) had to be implanted. Patency of the arterial bed of the right kidney was 60–65% after this intervention. Subsequent heparin was administered (25 000 units/day, intravenously). Unfortunately, severe bleeding from the puncture site at the right femoral artery started 12 h after the intervention and surgical suture was necessary. Heparin treatment had to be interrupted. Subsequently renal function improved and serum creatinine decreased to 132 µmol/l.

After 2 weeks control angiography was performed and the main right renal artery was partially, and peripheral arteries completely, patent (Figure 2). Subsequently recanalization of the left renal artery was achieved by aspiration of the thrombotic mass (thrombolysis was not attempted because of the bleeding problems on the first occasion). This was followed by PTA and successful placement of a Palmaz stent (because of ostial dissection). Subsequently, renal arteriography showed patency of the left main renal artery and of ~50–60% of the peripheral branches. Long-term anticoagulation with warfarin (6–9 mg/day) and aspirin (100 mg/day) was administered (INR 2.5–3.0).

Ten weeks later dynamic renal scintigraphy with 99mTc-DTPA was performed. Activity projecting upon the lower poles was comparable on both sides; only 28% activity, however, it was registered in projection upon the upper half and 72% upon the lower half of the left kidney.

One year later, the patient is normotensive, has no complaints and is in excellent condition. Serum creatinine (103 µmol/l) and a creatinine clearance (0.95 ml/s) are impaired only slightly. Titres of ACLAs are still elevated (IgG isotype 4.2 and IgM isotype 3.1). Other routine antibodies are negative.

A control renal angiography (repeated 1 year later) yielded a satisfactory result with only slight irregularities of the main renal artery and minor defects in the parenchymal phase on the left kidney, while the right renal artery was fully patent and the parenchymal phase of the right kidney was homogenous with typical corticomedullary demarcation (Figure 3).
Discussion

Renal disease in the primary antiphospholipid syndrome (APS) is characterized by non-inflammatory occlusion of renal blood vessels ranging in size from glomerular capillaries to the main renal artery and vein. The clinical signs of thrombi in the glomeruli and small arteries are variable and range from asymptomatic mild proteinuria to acute or subacute renal failure with proteinuria, active urine sediment and (often marked) systemic hypertension [4]. Renal infarcts are characterized by prominent unilateral or bilateral lumbar pain and severe hypertension, as described by Mandreoli and Zucchelli [5]. The incidence of renal involvement in the APS is unclear. Some authors have reported a prevalence of renal disease in up to 25% of patients with APS [6].

Thrombosis of the large renal arteries has rarely been described. This case is exceptional in that selective thrombotic occlusion of both renal arteries occurred without any signs of thrombosis in extrarenal location. Residual diuresis was preserved because the lower pole of the left kidney was fed by an accessory renal artery. A specific reason why isolated thrombosis of both renal arteries occurred is not entirely clear. Primary bilateral ostial stenosis (due to fibromuscular dysplasia?) cannot be excluded. Ostial dissection on both sides after PTA points to this possibility (an accessory branch perfusing the lower pole of the left kidney clearly had no stenosis and remained patent). Severe hypertension was evidently provoked by the progressive decrease of renal perfusion and the ensuing activation of the renin-angiotensin system. Restitution of renal perfusion was followed immediately by a normalization of blood pressure and subsequent improvement of renal function in spite of long-term ischaemia. Cardiac impairment and dilatation, as seen in our patient, is frequent in patients with APS and SLE [7]; it disappeared spontaneously after recanalization of the renal artery.

Thrombolysis of the right renal artery was followed by excellent recovery of renal function. Aspiration of thrombotic mass as from the left renal artery, particularly the upper pole branches, was efficient but not followed by full recovery of the function. Probably delayed recanalization and withholding of thrombolytic therapy account for this outcome. Long-term anticoagulant therapy successfully prevented a relapse of thrombosis during 1-year follow-up. Oral contraceptive could have been an aetiological factor and was stopped.

References


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