CASE REPORT

POLYARTERITIS NODOSA AND THE ANTIPHOSPHOLIPID SYNDROME

B. DASGUPTA, M. K. ALMOND* and A. TANQUERAY†

Departments of Rheumatology, *Nephrology and †Radiology, Southend General Hospital, Prittlewell Chase, Westcliff-on-Sea, Essex SS0 0RY

SUMMARY

We describe a case of classical polyarteritis nodosa (PAN) with visceral aneurysms presenting with renal infarction and hypertension. The female patient also had all the laboratory features of the antiphospholipid syndrome (APS) and 2 months into her illness developed a large iliofemoral thrombosis. She responded well to immunosuppressive therapy and anticoagulation. Repeat arteriogram showed regression of the visceral aneurysms. The link between PAN and APS, and the therapeutic dilemma posed by this association, are discussed.

KEY WORDS: Classical PAN, Visceral aneurysms, Anticoagulation, Antiphospholipid syndrome.

CASE REPORT

A 64-yr-old woman was admitted acutely under the surgical team at Southend Hospital with severe abdominal pain for several hours in November 1994. She had suffered with classical migraine in the past, both her pregnancies were complicated by hypertension and she had two episodes of pulmonary emboli (PE) while on hormone replacement therapy. She had been well until 10 days pre-admission, when she developed headaches.

On admission, she looked very ill with tenderness in the upper abdomen. She was hypertensive with a blood pressure of 220/130 mmHg. Urinalysis showed 4+ proteinuria, 3+ haematuria. Full blood count revealed haemoglobin 10.5 gm/dl, white cell count 5.2 x 10^9/l, platelets 244 x 10^9/l and the erythrocyte sedimentation rate (ESR) was 109 mm/1st h. Her biochemical tests showed elevated ALT 56 IU/l, gamma GT 627 IU/l, alkaline phosphatase 457 IU/l, urea 8.5 and creatinine 144 μmol/l. Hepatitis B surface antigen was negative, and abdominal ultrasound and ERCP failed to reveal any abnormality. The coagulation profile revealed a prothrombin time of 12.4 s, INR 0.9, fibrinogen >999 (200–400) mg/dl, and activated partial thromboplastin time (APTT) was prolonged at 56.6 s (21–34). Lupus anticoagulant was strongly positive and she had high titres of anticardiolipin IgG 97 GPLU/ml and IgM 43 MPLU/ml (normal range <20) antibodies. Antinuclear antibodies were positive, DNA binding borderline elevated at 114 IU/ml (normal range <100) and perinuclear ANCA was positive.

A diethylene triamine pentacetate scan showed a small right kidney with reduced uptake and excretion. A second ultrasound confirmed a small-sized right kidney. She then underwent abdominal arteriography. The flush aortogram showed occlusion of the right renal artery and multiple aneurysms in the branches of the hepatic arteries (Fig. 1). Selective left renal arteriogram showed multiple small aneurysms in the intrarenal vessels (Fig. 2).

A diagnosis of classical polyarteritis nodosa (PAN) with renal involvement was made, and the patient started on a regimen of pulsed cyclophosphamide and methylprednisolone. This consisted of 2-weekly i.v. pulses of cyclophosphamide (500 mg/m² body surface area) and methylprednisolone (500 mg) for 12 weeks. After 12 weeks, the i.v. cyclophosphamide pulses were administered at monthly intervals for a further 6 months along with...
gradually tapering doses of oral prednisolone. Cyclophosphamide was replaced by azathioprine after 9 months. Her hypertension was treated with amlodipine and labetolol. Despite the laboratory features of the antiphospholipid syndrome (APS), a decision was made not to anticoagulate in the presence of widespread aneurysms and risk of bleeding. However, 2 months into her treatment, she presented with an acutely swollen left thigh and leg. Venography showed a large iliofemoral thrombus and she was warfarinized.

She responded very dramatically to therapy, with full blood count and ESR returning to normal within 3 weeks. Her blood pressure came under control and microscopic haematuria resolved. Her creatinine remains mildly elevated at 140–150 $\mu$mol/l. A repeat angiogram a year later shows virtually complete resolution of the microaneurysms (Fig. 3). Her current medications are azathioprine 100 mg daily, prednisolone 5 mg daily, amlodipine and warfarin. Twenty months post-diagnosis, she remains stable and asymptomatic.

**DISCUSSION**

This patient presented with abdominal pain and hypertension caused by renal infarction due to classical PAN and the APS. Classical PAN is a systematic necrotizing vasculitis involving small and medium-sized arteries, and visceral arterial aneurysms were noted in the original description by Kussmall and Maier in 1866. The presence of aneurysms is associated with more severe disease, such as loss of weight, severe hypertension, abdominal pain, cardiac involvement, central nervous system involvement and poorer prognosis [1, 2]. In contrast, this woman has done very well so far and repeat angiogram has shown a significant decrease in the number and size of the aneurysms, indicating arterial healing. Regression of aneurysms with treatment has been reported so far only in PAN associated with hepatitis B virus infection [3].

The clinical features of APS include arterial and venous thromboses, recurrent miscarriages, strokes, migraine, livedo reticularis and thrombocytopenia in the presence of prolonged APTT, positive lupus anticoagulant and elevated levels of anticycliclin antibody. It may be secondary to diseases such as systemic lupus erythematosus or occur in isolation [4]. The association of APS with PAN is a rarity, although it has been described with PAN as well as a few other vasculitides [5]. Was this woman’s condition due to vascular inflammation (PAN), vascular thrombosis (APS), or both? The past medical history of recurrent PE, migraine and pregnancy-induced hypertension would fit with a pre-existing APS, and Norden et al. [5] speculated whether antiphospholipid antibodies favour the development of vasculitis. It is
impossible to evaluate the impact of these two factors separately. However, the associations of the two conditions posed a therapeutic dilemma since anticoagulation in the presence of multiple aneurysms may be hazardous in view of the real risk of abdominal bleeding from rupture of visceral aneurysms [6].

REFERENCES