Coeliac artery stenosis and antiphospholipid (Hughes) syndrome/antiphospholipid antibodies

Sirs,

Arterial and venous thromboses are cardinal features of the antiphospholipid (Hughes) syndrome (APS) (1). Recent reports suggest that arterial stenotic lesions may also be observed in APS patients (2, 3). For example, there are anecdotal reports of mesenteric artery stenosis in patients with APS (4, 5). We have observed a spectrum of coeliac artery disease ranging from narrowing to complete occlusion in APS patients and describe 17 patients with coeliac artery lesions and antiphospholipid antibodies (aPL). Fourteen patients fulfilled the Sapporo criteria for APS. Three patients only had positive aPL. Thirteen patients had SLE (12 with APS and 1 aPL only) and 2 primary APS. Ten patients complained of abdominal angina and weight loss or failure to thrive. One other patient with lupus nephritis had recurrent chronic gastric erosions, which failed to respond to conventional therapy of proton pump inhibitors; her gastric biopsy was suggestive of ischaemia. All but one were young women of Caucasian origin. Their median age was 42 years (29 to 78 years). Only one who was overweight (BMI 26), had diabetes mellitus, all other had BMI <25. Two patients smoked more than 7 cigarettes/day without any evidence of peripheral vascular disease, one other had high cholesterol for which she was on a statin and five patients had arterial hypertension. They all had normal cardiac rhythm and none were receiving oestrogen containing hormone replacement therapy. Six of 14 patients with APS were anti-coagulated for their previous thrombotic events and remaining received aspirin. We used magnetic resonance angiography as the principal method of investigation. All our patients were screened at the end-inspiratory phase minimising the error of median arcuate ligament compression syndrome (6).

All patients had coeliac artery stenosis with variable severity. One patient had calcification at the origin of the coeliac trunk. Six patients had complete occlusion of the coeliac artery (Table I) including 2 patients with secondary APS where there was retrograde filling of the coelic artery from the superior mesenteric artery. This may explain the lack of abdominal symptoms in these patients. The patient with recurrent gastric erosions and ischaemic gastritis on biopsy had significant narrowing of the coeliac trunk. The lesions observed in these patients were not typical of atherosclerosis as evidenced by the smooth and regular aorta. The lesions were at and distal to the ostium and were smooth and regular (Fig. 1).

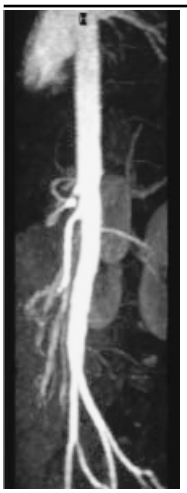

Fig. 1. MRA showing coeliac artery stenosis in a patient with APS.

A previous report by our group described an inferior mesenteric artery occlusion causing large bowel infarction in a patient with SLE and positive aPL (4). There is also a case series in which coeliac artery stenosis was observed in a patient with HIV and aPL (5). In a case report 37-year-old woman with primary APS who developed widespread necrosis of the small intestine and ascending colon, histological examination of the resected superior mesenteric artery showed complete thrombosis 2 cm distal from its origin with smooth muscle proliferation (7). Similarly, Sipek-Dolnicar examined histopathological and immuno-histological findings in his 14 patients with SLE of whom 13 had anticardiolipin antibodies (aCL). The autopsy samples of these patients showed frequent vascular occlusion due to thrombosis, thrombotic microangiopathy related changes and arterial intimal fibrous hyperplasia. Surprisingly immune complexes were sparingly seen in those patients with aCL (8). The accelerated atherosclerosis is also a known complication of APS (9). The intima –medial thickness has also been shown to increase in APS patients who did not have any traditional atherosclerotic risk factors (10). These findings suggest the possibility of thrombosis, endothelial dysfunction and/or accelerated atherosclerosis individually or together may be responsible for the coeliac artery stenosis in patients with APS/aPL.

In summary, our data describing 17 patients with lesions of celiac trunk indicate that, as in the renal arteries, the coeliac arteries may also be involved in patients with aPL. We have previously described renal artery stenosis in aPL positive patients with uncontrolled hypertension (2). This preliminary data extends this observation by reporting a case series of aPL positive patients with coeliac artery lesions. We have now embarked upon a controlled study to assess the prevalence of CAS and it's association with aPL.

Table 1.

Patients (n = 17)	Symptomatic	Asymptomatic	Total Occlusion	Incomplete occlusion
Primary APS (n = 2)	1	1	1	1
Secondary APS /aPL (n = 13)	8	5	4	9
Positive aPL only (n = 2)	1	1	1	1

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