LESSON OF THE MONTH

A rare case of shock

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CASE HISTORY

A 48 year old non-smoking women presented with a six hour history of colicky right upper quadrant pain radiating to the back preceded by an episode of syncope. Past history included hyperthyroidism treated by partial thyroidectomy.

Initial examination showed mild right upper quadrant tenderness. Investigations demonstrated a normocytic anaemia (haemoglobin 92 g/l), white cell count $15.3 \times 10^9/l$, with normal liver function tests and amylase. Twelve hours later she developed shock associated with a distending abdomen.

Emergency laparotomy showed a haemoperitoneum and huge retroperitoneal haematoma with catastrophic bleeding from the duodenojejunal region requiring 13 units of blood and 8 litres of colloid and crystalloid. Haemostasis was achieved with aortic cross suturing around a branch of the superior mesenteric artery (SMA) and packing the peritoneum. Packs were removed 24 hours later without complication.

A postoperative mesenteric angiogram delineated multiple small aneurysms in the gastroduodenal artery and its pancreatic branches, hepatic artery, and ileal branches of the SMA with microaneurysms of the main renal, lobar, and segmental arteries (fig 1). Differential diagnosis included polyarteritis nodosa (PAN) and fibromuscular dysplasia (FMD).

Further investigations, including erythrocyte sedimentation rate, C reactive protein, autoantibody screen, antineutrophil cytoplasmic antibodies, rheumatoid factor, and cardiolipin antibodies, were unremarkable. Hepatitis B and C and syphilis serology were negative. α_1 Antitrypsin levels, fun-

doscopy, urine microscopy, echocardiography, and muscle biopsies were also normal.

Subsequent magnetic resonance angiography (MRA) with gadolinium enhancement was performed to determine the extent of disease and identify vessels amenable for biopsy. Intracranial and superficial femoral vessels appeared unaffected. Mesenteric images demonstrated mild irregularities of the SMA and the suspicion of a splenic artery aneurysm only. Biopsy was not performed.

Further evaluation of her family history showed that her father and paternal uncle had both died in the fifth decade after a second myocardial infarction and a cerebrovascular accident, respectively.

Fibromuscular dysplasia was considered to be the diagnosis. The patient has remained well during three years' follow up without immunosuppression. Tachycardia and hypertension (recorded during waking hours on 24 hour monitoring) was controlled with propranolol.

DISCUSSION

FMD is a non-inflammatory, non-atherosclerotic vascular disease which may clinically and angiographically mimic PAN.¹ FMD, however, typically presents in young white women, whereas PAN characteristically presents in men in the fifth decade of life. Arterial narrowing and aneurysms in FMD reflect abnormal deposition of collagen and smooth muscle hyperplasia in contrast with cellular infiltration, fibrinoid necrosis, and healing elastic lamina in PAN. Classic PAN is restricted to medium sized vessels (Chapel Hill Criteria), but FMD may affect both medium and small vessels.² End organ damage due to ischaemia, haemorrhage, and infarction is common to both conditions.

FMD most commonly affects the renal and cerebral arteries, occurring in 89% and 25% of cases respectively.¹ FMD is recognised as a common cause of renovascular hypertension and may have a prevalence of 1% in the American population.³ FMD of the mesenteric arteries is, however, rare occurring in only 9% of cases of FMD. Mesenteric FMD is usually asymptomatic owing to collateral circulations but may present with mesenteric ischaemia (postprandial abdominal pain, weight loss, and epigastric bruit), recurrent gastrointestinal bleeds or rarely, as in this case, with acute abdomen and shock.

The diagnosis of FMD is definitively made by histopathology, which will also allow the disease to be classified according to the dominant arterial wall layer affected—for example, intimal, medial, or adventitial. Biopsy, as is in this case, is often not feasible. The diagnosis in this case was suggested by the angiographic “string of beads” appearance representative of

Figure 1 Digital subtraction mesenteric angiogram demonstrating “string of beads” of fibromuscular dysplasia

Abbreviations: FMD, fibromuscular dysplasia; MRA, magnetic resonance angiography; PAN, polyarteritis nodosa; SMA, superior mesenteric artery

alternating thinned media and collagen containing fibromuscular ridges characteristic of medial FMD. Of note, FMD was not clearly visualised on MRA, reflecting the reduced sensitivity of this technique for small vessel disease compared with conventional angiography. FMD may also lack gadolinium enhancement in contrast to vasculitis.⁴ The lack of acute phase reactants in this case were against a diagnosis of PAN. Infarction complicating FMD may confuse this issue and, similarly, the initial leucocytosis in this case might have reflected the catastrophic intra-abdominal bleed.

The family history in this case is suggestive of FMD, which appears to have an autosomal dominant inheritance with incomplete penetrance.⁵ Cerebrovascular accident is a common manifestation of carotid FMD, and cardiac death may rarely occur owing to stenosis or dissection of the coronary arteries, narrowing of the sinus node artery or, possibly, owing to coexisting cardiomyopathy.^{6,7}

The aetiology of FMD is unknown, although hormones, smoking, drugs—for example, methysergide and ergotamine, have been implicated along with a genetic component. α_1 Antitrypsin heterozygosity may be a risk factor for FMD but was not present in this case.⁸ Deficiency of this powerful inhibitor of circulating proteolytic enzymes—for example, collagenase, could disrupt the structural integrity of connective tissue. FMD has also been described in association with Alport's syndrome, a disease characterised by abnormal type IV collagen. Pheochromocytoma, frequently in a homolateral kidney, may also occur with FMD.⁶ The occasional regression of FMD after tumour resection has suggested that localised high concentrations of catecholamines may contribute to the pathogenesis of FMD—for example, by inducing vasospasm. The presence of persisting tachycardia and hypertension in this case led to a search for pheochromocytoma, but this was not present.

This woman has remained well three years after diagnosis without treatment, emphasising the need to consider this diagnosis to avoid inadvertent immunosuppression.

THE LESSONS

- Mimics of vasculitis must be considered in the absence of systemic manifestations of disease to avoid inappropriate treatment.
- Full evaluation of family history may not be possible in an emergency, but subsequent review may help lead to the correct diagnosis.
- MRA is helpful for assessing medium vessel disease but may miss disease of its branches.

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