A case of segmental arterial mediolysis: a vasculitis mimic

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Segmental arterial mediolysis (SAM) is a rare but increasingly recognised non-inflammatory, non-atherosclerotic and non-infectious vasculopathy of unknown aetiology that can potentially lead to life threatening manifestations. It involves disruption of the arterial medial layer causing dissection, aneurysm, occlusion or stenosis of abdominal arteries, with cases reported of coronary and intracranial vessel involvement. Patients commonly present with abdominal pain and/or spontaneous intra-abdominal haemorrhage. SAM may be confirmed histologically, but in practice usually relies on clinical and radiological interpretation and expertise. It typically involves medium-large sized vessels which can mimic vasculitis. Management includes surgical or endovascular procedures and strict blood pressure control. Mortality has been reported from 22%–50%. The natural clinical course of this arteriopathy is unpredictable, and continued surveillance for several years is recommended to detect early disease progression and to maintain optimal blood pressure control.

A 40-year-old dairy farmer presented to the emergency department with nausea, vomiting and dizziness. She was found to be hypertensive to 184/97mmHg. Symptoms were thought to be related to a viral illness, and she was discharged. She re-presented 12 days later with similar symptoms, along with confusion and developed abdominal pain, and hypovolemic syncope with a blood pressure of 78/60mmHg. An extensive septic screen was negative. Her haemoglobin was 122 g/L which precipitously dropped to a nadir of 83 g/L (N=115–155). A computed tomography scan and angiogram of the abdomen revealed large volume hemoperitoneum due to an inferior pancreatoduodenal artery (PDA) pseudoaneurysm, and subsequent superior mesenteric artery dissection (see Figure 1). Renal infarcts and renal artery beading was also identified (see Figure 2). The PDA pseudoaneurysm was confirmed on catheter angiography. Her C-reactive protein on presentation was normal and gradually increased to 109 mg/L (N<5). Her lipase was normal on presentation and when repeated during her admission.

Differentials raised included systemic vasculitis, fibromuscular dysplasia (FMD), segmental arterial mediolysis, or collagen disorder such as Ehlers-Danlos or another connective tissue disorder (CTD). There were no clinical features of a CTD or autoimmune vasculitis on examination. Documented blood pressures in hospital and the community varied with the highest documented reading being 191/128mmHg. Antinuclear antibody and anti-neutrophil cytoplasmic antibodies were not detected. An aortopathy 52-gene panel returned negative. Management involved blood transfusion with four standard units and fluid resuscitation, PDA pseudoaneurysm coiling (see figure 3), multi-agent management for hypertension and antiplatelet therapy. Within two weeks, she was discharged home feeling well with a plan for blood pressure monitoring in the community and surveillance CT scans.

Of note, eight years earlier, the patient had a vertebral artery dissection with subarachnoid haemorrhage which was successfully treated with coiling. One year later, she re-presented with dizziness and headache, was found to have recurrent aneurysm, went on to have a revision, and a flow diverting stent was inserted. She also received anti-platelet therapy and made a full recovery.

Discussion

Segmental arterial mediolysis (SAM) is increasingly recognised as a vasculitis mimic, and may present with a number of similar vascular abnormalities frequently complicated by abdominal haemorrhage. It is interesting to speculate if her prior presentation with subarachnoid haemorrhage was related. Of relevance, her current presentation occurred approximately two months after she stopped her anti-hypertensive medications. It was felt that her presentation with abdominal pain, large volume hemoperitoneum with mesenteric artery involvement made SAM more likely than FMD. An awareness of the condition is relevant for vascular and general surgeons, and rheumatologists/immunologists in particular—the latter often consulted regarding
possibilities of vasculitis or associated connective tissue condition (eg Ehlers-Danlos, Marfan syndrome). As clinicians, we are very dependent on our vascular radiology colleagues for their expertise in evaluating the imaging findings. An accurate diagnosis avoids the unnecessary introduction of high dose corticosteroid therapy, which is tempting to commence (and continue long-term) in an unwell patient with radiology findings suggestive of medium-large vessel vasculitis. SAM can be difficult to distinguish from FMD, with some proposing that SAM may represent a precursor of certain types of FMD.\textsuperscript{9–10} Strict blood pressure control and radiological surveillance are the mainstay of management.

**Figure 1:** SMA dissection
Figure 2: beaded appearance of renal artery (1), renal infarct (2).
Figure 3: Pre (1) and Post (2) PDA pseudoaneurysm coiling
COMPETING INTERESTS
Nil.

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