CASE REPORT

Endovascular Treatment of a Splenic Aneurysm Associated With Segmental Arterial Mediolysis

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Introduction: Segmental arterial mediolysis is a rare disorder characterised by disintegration of the medial layer of an arterial wall usually affecting the intra-abdominal splanchnic vessels.

Report: A case of 50 year old man who presented with sudden-onset left sided flank pain is reported. A computed tomography mesenteric angiogram showed haemorrhage and a stable left upper quadrant haematoma arising from 8 × 8 mm splenic artery aneurysm.

Discussion: The patient underwent a successful endovascular coiling procedure to exclude the aneurysm and for complete resolution of his symptoms.

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INTRODUCTION

Segmental arterial mediolysis (SAM) is a rare non-atherosclerotic and non-inflammatory arteriopathy characterised by lytic degeneration of the media, resulting in aneurysm formation and vessel rupture. Typical presentation of SAM includes acute onset of abdominal pain from haemorrhage.

The case of 50 year old man who presented with bleeding from a splenic artery aneurysm and associated mesenteric panniculitis is described.

Consent was obtained from the patient to publish this case report along with the images.

CASE REPORT

A 50 year old man presented with left sided flank pain for 2 days.

His past medical history was significant for treated hypertension and dyslipidaemia. On examination he appeared generally well and his vital signs were within normal limits. His abdomen was soft with mild tenderness in the left upper quadrant and no bruit. All his peripheral pulses were symmetrical and intact. There were no clinical features of cutaneous vasculitis, mononeuritis multiplex or pulmonary, visual, auditory, or sinus disease.

Blood investigations showed stable haemoglobin 161 g/L, white cell count 8.8 × 10^9/L and platelet count 183 × 10^9/L.

The kidney and liver function tests were normal. Urine analysis was unremarkable. Subsequent immunological tests showed C-reactive protein 28.5 mg/dL. The erythrocyte sedimentation rate was 14 mm/hour. The vasculitis screen, including ANCA, MPO and PR3 antibodies, myeloperoxidase, and proteinase antibodies, were negative. The hepatitis and HIV serology were negative.

Chest X-ray was unremarkable. A triple phase computed tomography (CT) scan of the abdomen showed a small, hyperdense collection in the retrogastric, perisplenic, and retropancreatic space. There was moderate fat stranding surrounding the root of the mesentery with a few intermediate sized lymph nodes, the largest node measuring 14 × 9 mm, suggestive of mesenteric panniculitis (Fig. 1).

A CT mesenteric angiogram showed a small aneurysm arising from the splenic artery measuring 8 × 8 mm and a stable left upper quadrant haematoma (Fig. 2). There was significant oedema and thickening of the vessel walls of the coeliac artery and its branches namely common hepatic, proper hepatic, gastroduodenal, and splenic arteries (Fig. 3). There was no abnormality of the superior or inferior mesenteric arteries.

A positron emission tomography CT scan showed mild to moderate fludeoxyglucose (FDG) uptake (standardised uptake value max. 4.0) along the vessel wall of coeliac trunk, common hepatic artery and proximal splenic artery corresponding to diffuse vessel wall thickening noted on CT mesenteric angiogram. The aortic arch was mildly dilated to 34 mm with mildly increased FDG uptake along the wall of the aortic arch suspicious of underlying medium or large vessel vasculitis.

A temporal artery biopsy was negative for giant cell arteritis. A specialist rheumatology and immunology
opinion was sought. The patient was diagnosed with SAM based on the clinical, radiological, and laboratory findings.

An endovascular coiling procedure was performed to occlude the aneurysm. Multiple Nester® Embolisation Coils were used to occlude the aneurysm. Completion angiogram showed no extravasation of contrast from the aneurysm (Fig. 4).

The patient was followed-up with no further episodes of abdominal pain or discomfort.

**DISCUSSION**

The first case of a distinct arterial lesion in the large abdominal muscular arteries was described by Slavin et al. in 1976 and was termed “segmental arterial mediolysis” (SAM) in 1995. Alterations of SAM stem from two separate lesions: mediolysis and a tear that separates the outer medial muscle from the adventitia. As a result, sudden haemorrhage due to aneurysm rupture, or dissection occurs in the abdomen, retroperitoneum, or brain. Cases of SAM that are complicated by intra-abdominal haemorrhage have mortality rates approaching 25—50%.

The most common sites of visceral arterial aneurysms are the splenic, hepatic, superior mesenteric, coeliac, gastric, gastroepiploic, jejunal, ileal, and colonic arteries and their tributaries. Inada et al. reported 27 cases of SAM in Japan with a third of the cases having multiple intra-abdominal aneurysms. Matsuda et al. reported intracranial and intra-abdominal aneurysms, associated with SAM in a patient who suffered subarachnoid haemorrhage due to rupture of an intracranial aneurysm.

Distinction between isolated visceral artery dissection that could be of several types and segmental arterial mediolysis is important. In superior mesenteric artery (SMA) dissection the characteristic finding of a “double lumen sign” of the SMA on axial views of CT images requires...
that both true and false lumen are patent. However, about 25% of visceral dissections have an occluded false lumen (often called type Ib). Additionally, they can have lumen variations and aneurysm formation with extravasation both close to aortic take-off or more distally. That is the same as aortic type A or B dissection and penetrating aortic ulcers and intramural haematomas—they are probably related. Currently our knowledge is limited.

There were no other convincing clinical manifestations of Von Recklinghausen’s neurofibromatosis polyarteritis nodosa, Wegener’s granulomatosis or Churg Strauss vasculitis, and certainly none to suggest Ehlers Danlos syndrome (EDS). The patient was not hypermobile and did not have the typical phenotype of patients with EDS. There were no clinical manifestations of Marfan’s syndrome, which would be self evident and would probably have presented with other somatic manifestations several decades prior to this patient’s presentation in his sixth decade. Kawasaki disease is not seen in adults and is a paediatric form of vasculitis. The patient did not have the fevers, typical strawberry tongue, or any evidence of coronary arterial disease that are cardinal manifestations of this condition.

The rapid subsidence of the inflammatory markers without any immunosuppressive intervention is against a systemic vasculitic process, although it must be conceded that there are case reports of isolated intra-abdominal manifestations of polyarteritis nodosa or giant cell arteritis that can present with omental haemorrhage/haematoma, or involve isolated organ systems in the abdomen, including the gall bladder, as well as the uterus and the testicle. Surgical excision in such cases has been found to be curative as there is no systemic disease.

It is most uncommon to have significant abdominal involvement with a vasculitic process over an extended period to produce microaneurysms/aneurysms without symptoms of end organ involvement and especially mesenteric ischaemia.

Fibromuscular hyperplasia could be more difficult to exclude immediately on clinical grounds, but the absence of a history of dissection of upper limb or cervical/cranial vasculature and the absence of upper limit claudication complicating subclavian disease or background of renal arterial dissection/stenosis is the most common presentation and would be useful findings militating against this diagnosis.

The pathogenesis of SAM is uncertain, but is thought to be a response to repeated episodes of vasospasm. Treatment of SAM involves embolisation, surgical bypass, or resection of the injured arteries. Ryan et al. described the first case of coil embolisation. The typical digital subtraction angiography (DSA) features of the disease were first described by Heritz et al., who found a pattern of focal aneurysms, beading, and narrowing of the splanchic and renal arteries with an otherwise normal vascular appearance.

Angiographically, patients may present with one or more of six distinct features: simple arterial dilatation, a single aneurysm, multiple aneurysms, dissecting haematomas, stenosis, and arterial occlusions. The most common sites of involvement include the coeliac axis, the superior mesenteric artery, the renal arteries, and the inferior mesenteric artery.

Histology is the diagnostic gold standard for segmental arterial mediolysis; however, the diagnosis of segmental arterial mediolysis is most often made using DSA or CTA and based on the characteristic pattern of arterial involvement and morphological changes after excluding vasculitis by clinical and laboratory findings.

In the present case, the patient’s symptoms were probably related to bleeding from the splenic artery aneurysm associated with segmental arterial mediolysis.

Although the patient’s symptoms completely resolved after coiling and embolisation of the splenic artery aneurysm, a high degree of vigilance is required with surveillance CT scans as little is known about aneurysmal degeneration due to SAM. In the literature, there are fewer than nine reported cases of splenic aneurysms due to SAM. The authors suggest regular follow-up and monitoring to prevent a catastrophic bleed from rupture of splenic aneurysms.

CONCLUSION
Segmental arterial mediolysis is a rare cause of bleeding from a splenic artery aneurysm. Endovascular treatment with coiling and embolisation is a safe and effective technique to prevent rupture and complications arising from splenic artery aneurysms associated with SAM. Close follow-up is required in patients diagnosed with SAM.

CONFLICT OF INTEREST
None.

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