Visceral ischemia: could it be segmental arterial mediolysis

Sunil Agarwal · Edwin Stephen · Dheepak Selvaraj · Kapil Mathur · Shyamkumar Keshava · Sunil Thomas Chandy

Abstract
We present two cases of segmental arterial mediolysis, which can present with dissecting aneurysms or thrombosis of the visceral branches of the abdominal aorta. Segmental arterial mediolysis (SAM) causes ischemic bowel disease and has characteristic CT and angiographic features.

Keywords Abdominal pain · Segmental arterial mediolysis

Introduction
Segmental arterial mediolysis (SAM) is a rare non-arteriosclerotic non-inflammatory vascular disease that presents with aneurysms, dissection or thrombosis of the visceral branches of the abdominal aorta causing ischemic bowel disease. The diagnosis is most often based on its characteristic CT and angiographic features. It is likely to be increasingly identified with the growing use of CT scans in the evaluation of abdominal pain but nevertheless, to the best of our knowledge, has not been reported in Indian literature.

Case 1
A 52-year-old man presented to the emergency room with abdominal pain in the left upper quadrant radiating to the left side of the chest and vomiting for 5 days and fever for 2 days.

On examination, his pulse rate was 120/min and blood pressure was 170/100 mmHg. His abdomen was distended with mild tenderness and guarding in the left hypochondrium and epigastrium. Active bowel sounds were heard on auscultation. All routine blood investigations were normal. Contrast-enhanced CT, arterial and portal venous phases showed linear luminal narrowing and a thrombosed false lumen extending from the celiac artery origin to the splenic artery upto the splenic hilum, suggestive of celiac artery dissection (Fig. 1). Multiple splenic infarcts were seen. His echocardiogram was normal.

The patient’s blood pressure was stabilized and he was administered heparin. His condition improved and he was discharged on antihypertensives and anticoagulants. He was asymptomatic at 3 months follow up.

Case 2
A 52-year-old hypertensive man presented with spasmodic central abdominal pain, which increased on taking food, since 20 days. There was no history of abdominal distension, vomiting, diarrhea or melena.

On examination, his pulse rate was 100/min and blood pressure was 180/110 mmHg. There was mild ten-
derness over the epigastrium. All peripheral pulses were well felt.

Routine blood tests were normal. CT angiography showed dissecting aneurysms of the celiac artery, common hepatic artery, SMA and right renal artery. There were multiple areas of narrowing and dilatation of both renal arteries and focal areas of thickening of the walls of celiac, SMA and renal arteries. The abdominal aorta appeared normal.

Abdominal aortogram done subsequently showed a proximal aneurysm with distal narrowing before the trifurcation in the left renal artery (Fig. 2). The distal lesion was dilated with 3 mm × 12 mm coronary balloon and then the aneurysm was excluded with a Jostent® (Abbot Laboratories, Illinois) coronary stent graft. The celiac axis had a proximal aneurysm and was occluded distally. The splenic and gastroepiploic arteries were thrombus laden and were wired separately and dilated with 3 mm × 20 mm balloons and later the splenic artery was stented.

Post-procedure the patient improved. At 2 months follow up he was asymptomatic and his antihypertensive requirement had reduced.

SAM is a rare non-arteriosclerotic non-inflammatory vascular disease that mainly affects the visceral arteries of the abdomen presenting either with ischemic bowel disease or shock.\(^1\) It is postulated that SAM is the result of an inappropriate vasospastic response expressed in a splanchnic vascular bed undergoing vasoconstriction as a response to shock or severe hypoxemia. SAM is initiated by the transformation of the arterial smooth muscle cytoplasmic contents into a maze of dilated vacuoles containing edema-like fluid.\(^2\) These gaps may be filled with fibrin, thrombi, or granulation tissue and can lead to saccular aneurysms, dissecting aneurysms, or thrombosis.\(^1\)

Various forms of vasculitis must be considered in the differential diagnosis of segmental arterial mediolysis. Systemic inflammation with inflammatory destruction of the wall of the mesenteric arteries is seen in polyarteritis nodosa, Takayasu’s arteritis, Behcet’s syndrome, and Henoch–Schönlein purpura.\(^3\)

The typical digital subtraction angiography (DSA) features of the disease were first described by Heritz et al.,\(^4\) who found a pattern of focal aneurysms, beading, and narrowing of the splanchnic and renal arteries with an otherwise normal vascular appearance. Histology is the gold standard of segmental arterial mediolysis diagnosis; 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 morphologic changes after excluding vasculitis by clinical and laboratory findings.

The treatment of SAM is limited to surgical or endovascular interventional treatment for symptoms related to ruptured aneurysms or thrombosed arterial segments.\(^1\)

**References**