

## CASE REPORT

# Have You Seen SAM?

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**Introduction:** Segmental arterial mediolysis (SAM) is a rare, non-atherosclerotic, non-inflammatory vascular disease mostly affecting medium to large sized abdominal arteries which may cause aneurysms, stenosis, and haemorrhage.

**Report:** A case is reported of a patient with SAM affecting the renal arteries bilaterally, where the diagnosis was made by excluding other inflammatory, immunological, and infectious mimickers.

**Discussion:** As SAM carries a significant mortality and morbidity from end organ ischaemia, infarction, or haemorrhage, it should be considered in any patient presenting with abdominal pain.

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## INTRODUCTION

Segmental arterial mediolysis (SAM) is a rare, but important vasculopathy of unknown aetiology characterised by disruption of the arterial medial layer, with resultant susceptibility to vessel dissection, stenoses, occlusions, and aneurysm formation that can result in end organ haemorrhage, infarction, and ischaemia.<sup>1</sup> It mostly affects medium to large sized abdominal arteries, but any vessel can be involved, including intracranial, coronary, and retroperitoneal arteries.<sup>2</sup>

Although histological diagnosis is the gold standard, this is rarely feasible. SAM exhibits a characteristic pattern of arterial involvement and morphological changes on imaging such as catheter directed digital subtraction angiography (DSA) or computed tomography angiography (CTA). However, before the diagnosis of SAM can be made, vasculitides must be excluded by clinical and laboratory testing.<sup>3–5</sup>

## REPORT

A 53 year old Caucasian male presented to a tertiary hospital in 2014 with a 5 week history of bilateral loin pain and malaise, but no weight loss. He was noted to be hypertensive (186/94 mmHg) and had an elevated creatinine of 132 µmol/L. A CTA revealed bilateral renal artery stenoses, aneurysms, and occlusions with multiple renal infarcts (Fig. 1), but the other visceral arteries, including coeliac, superior mesenteric (SMA), and splenic, were disease free (Fig. 2).

A catheter directed DSA confirmed the CTA findings (Fig. 3).

Differential diagnoses were polyarteritis nodosum (PAN), fibromuscular dysplasia (FMD), and segmental arterial mediolysis (SAM). Given the acute clinical presentation with pain and malaise, with simultaneous bilateral renal artery aneurysms and infarcts, FMD was considered unlikely. Blood tests for elevated erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), globulins, anti-neutrophil cytoplasmic antibodies (ANCA), and hepatitis B surface antigens (Hep B Ag) were performed and the patient was commenced empirically on oral high dose prednisolone (80mg daily) for the treatment of PAN, in addition to therapeutic enoxaparin. An angiotensin converting enzyme inhibitor (ramipril) 5 mg twice daily and an  $\alpha$  blocker (prazosin) 0.5 mg three times daily were commenced for the management of hypertension in consultation with a renal physician. ESR and CRP were within normal limits and the patient was discharged with well controlled blood pressure (120–150 mmHg) and pain.

Subsequently, normal results were received for ANCA, globulins and Hep B Ag. Further multidisciplinary review led to a diagnosis of SAM on the basis of the clinical and radiological picture. Prednisolone was therefore ceased and the patient was monitored serially with inflammatory markers and renal function tests. Follow-up CTA imaging over the ensuing 5 months did not demonstrate progression (Figs. 4 and 5) and the patient's renal function normalised. The patient continues to be monitored by duplex ultrasound, inflammatory markers, and renal function tests.

## DISCUSSION

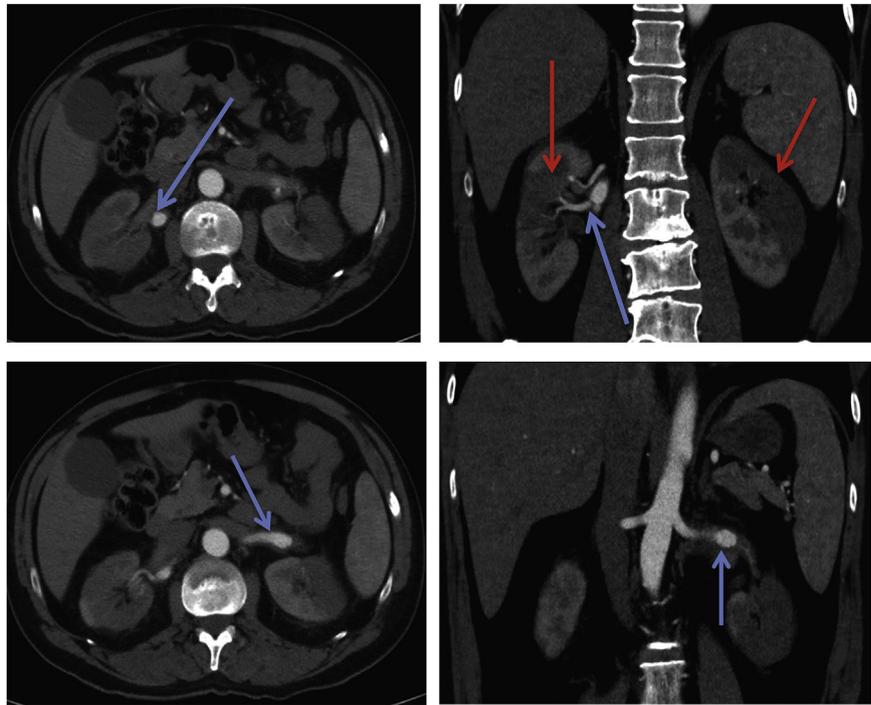
Segmental arterial mediolysis (SAM) was first described by Slavin in 1976, in three autopsy cases of ruptured large abdominal muscular artery aneurysms resulting in massive haemorrhage and death.<sup>3</sup> SAM is not a systemic disease, but rather a rare and acute non-atherosclerotic and non-inflammatory arteriopathy of unknown aetiology with life

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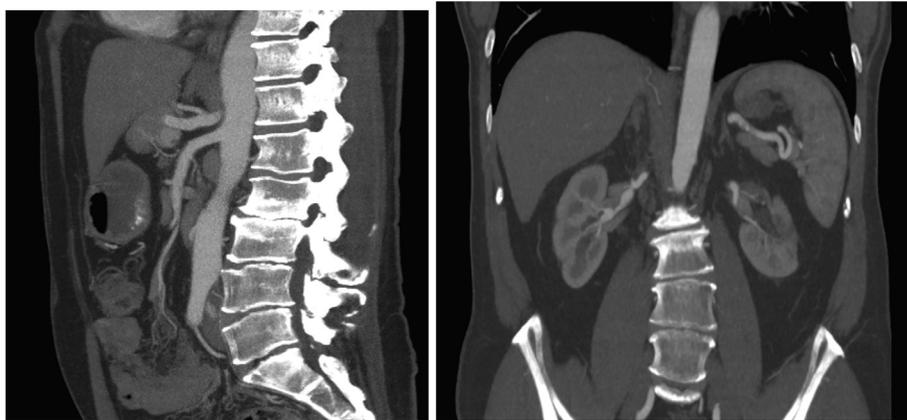
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**Figure 1.** Coronal and axial CT angiogram (arterial phase) demonstrating right and left renal artery aneurysms (blue arrows) and bilateral renal infarctions (red arrows). There is an approximately 14mm left renal and 10mm right renal aneurysm.



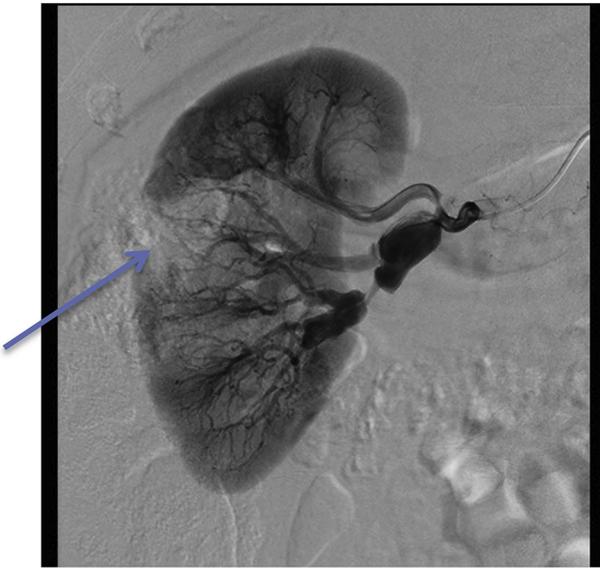
**Figure 2.** Sagittal and axial sections of CT angiogram demonstrated no aneurysms or stenoses within the coeliac, splenic, and superior mesenteric arteries.

threatening manifestations. Radiological and autopsy studies have demonstrated that two or more arterial segments from the same or adjoining vascular beds may be affected, and there is an acute phase mortality of nearly 50% from vessel rupture.

SAM is characterised by disruption of the arterial media leading to loss of the supporting muscular wall.<sup>3,4</sup> Four histological characteristics are described: mediolysis, separation, arterial gaps, and reparative fibrosis. Mediolysis refers to partial or total vacuolisation and lysis of the outer arterial media, resulting in weakening, formation of arterial gaps, and separation of the media from the adventitia.<sup>6</sup> This is followed by fibrin deposition at the media-adventitial junction, which predisposes to dissecting aneurysms. It can occur in a section of the arterial circumference or in its entirety, and

characteristically exhibits a segmental distribution. These lesions may rupture and announce the disease with a high mortality from haemorrhage. Stenoses and occlusions are caused by dissections or reparative granulation tissue with overlying organising thrombi.<sup>4</sup> The definitive pathogenesis of SAM is unknown, but it has been suggested that the arterial lesions develop as a consequence of an inappropriate response by endothelium to vasospasm such as hypoxia, hypotension, or sepsis.<sup>7</sup> The main clinical symptom is abdominal pain; a manifestation of abdominal haemorrhage or end organ ischaemia and infarction. Less commonly, it may present with haematuria or flank pain from renal artery involvement.

Demographically, SAM typically affects those of late middle age and the elderly, with a median age of 57 years.<sup>8</sup> No gender differences have been observed. No risk factors



**Figure 3.** Digital subtraction angiography (DSA) showing proximal right renal artery branches with stenoses and aneurysm. There is also an established infarct in the medial posterior pole of the kidney (arrow).

have been identified. It typically affects multiple splanchnic arteries as evidenced by a case report in 2016 which describes a patient with SAM of the coeliac, right renal, jejunal branch of the superior mesenteric, left gastric, and splenic arteries.<sup>9</sup> However, arterial involvement in the kidneys, liver, bowel, and pancreas is typically restricted to a small area in each involved organ.<sup>4</sup> In a recent systematic review of 85 cases of SAM between 1976 and 2012, the splenic artery was the most commonly involved in 29% of the cases followed by the coeliac trunk and renal arteries.<sup>8</sup> The overall SAM related mortality rate was 26%, of which 62% died before any intervention.<sup>5</sup>

Endovascular embolisation of the sequelae of SAM is a minimally invasive treatment option, and can be used to provide a temporary bailout in the acute phase before definitive treatment is carried out at a later stage. Open reconstructive surgical intervention using autologous vein grafts can be reserved for patients with recurrent bleeding or

in whom embolisation has failed.<sup>9</sup> In one study, 88% of patients had a successful outcome using endovascular techniques (79% coil embolisation alone). The overall open mortality rate was approximately 9%.<sup>8</sup> Stable cases can be treated successfully with non-surgical measures, as illustrated in the present study.

The differential diagnoses of SAM include FMD, PAN, and isolated mesenteric artery dissection (IMAD). FMD tends to affect children and young to middle aged women, with alternating stenoses and aneurysms. Dissections are less common and the renal and carotid arterial systems are preferentially affected.<sup>9</sup> FMD is rarely painful and is usually asymptomatic or associated with symptoms of occlusive disease.<sup>6</sup> Histologically, FMD is classified into three main types based on the dominant arterial wall layer involved: the intima, media, or adventitia (peri-medial). Intimal FMD, which constitutes less than 10% of all FMD cases, is characterised by circumferential deposition of collagen in the intima with long smooth stenoses.<sup>10</sup> Approximately 80% of FMD are of the medial type; the lesion is a homogenous collar of elastic tissue that presents as multiple stenoses interspersed with aneurysmal outpouchings, with a preserved internal elastic lamina. Peri-medial FMD accounts for 10% of dysplastic arteries and involves excessive tissue deposition at the media-adventitial junction.

PAN affects people of variable ages with p-ANCA positive serology and raised ESR and CRP, which were not a feature of this case. Pathologically, PAN demonstrates trans-arterial inflammation and fibrinoid necrosis.<sup>1</sup>

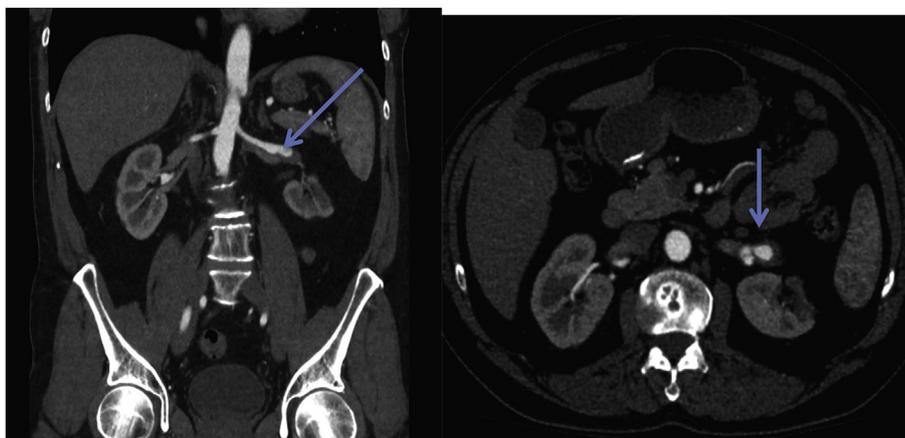
Although the present case was atypical of SAM as only renal arteries were involved, the differential diagnoses of FMD and PAN were excluded based on clinical, laboratory, and radiological findings. Furthermore, the diagnosis of IMAD was excluded as this entity is solely attributed to the superior mesenteric or coeliac artery.

**Table 1** illustrates the key demographics, clinical features, and vasculitic markers in SAM and its mimickers.

It is concluded that SAM is a rare but important cause of unexplained arterial lesions in patients presenting with normal inflammatory and vasculitic markers. It should be



**Figure 4.** CTA demonstrating no significant interval change in abnormalities consistent with SAM of the right renal artery (arrow).



**Figure 5.** CTA illustrating no significant interval change in aneurysms or stenoses within the left renal artery (arrows).

**Table 1.** Demographic, clinical features and laboratory findings distinguishing SAM from other mimickers.<sup>1</sup>

|  | SAM          | PAN      | FMD          | IMAD         |
|--|--------------|----------|--------------|--------------|
| Demographics                           |              |          |              |              |
| Age at onset, years                    | 40–60        | 40–60    | 20–40        | 50           |
| Gender predisposition (male: female)   | 1:1          | 1:1      | 1:3          | 5:1          |
| Clinical features                      |              |          |              |              |
| Hypertension                           | Occasionally | Yes      | Yes          | Yes          |
| Abdominal pain                         | Yes          | Yes      | Occasionally | Yes          |
| Gastrointestinal bleeding              | Yes          | Yes      | Occasionally | Occasionally |
| Inflammatory and immunological markers |              |          |              |              |
| Leukocytosis                           | No           | Yes      | No           | No           |
| Elevated CRP/ESR                       | No           | Yes      | No           | No           |
| ANCA +ve                               | Negative     | Positive | Negative     | Negative     |
| Hepatitis B surface antigen            | Negative     | Positive | Negative     | Negative     |

ANCA = anti-neutrophil cytoplasmic antibodies; CRP = C-reactive protein; ESR = elevated erythrocyte sedimentation rate; FMD = fibromuscular dysplasia; IMAD = isolated mesenteric artery dissection; PAN = polyarteritis nodosum; SAM = segmental arterial mediolysis.

considered in the differential diagnosis of visceral arterial disease characterised by dissection, aneurysm, stenoses, and occlusion in medium to large vessels, particularly when confined to one anatomical location. SAM is a rare but important cause of abdominal pain requiring diligence to enable early treatment before manifestations of life threatening haemorrhage or end organ infarction. Urgent referral to a vascular surgeon for appropriate imaging workup and management is recommended. The present authors recommend a multidisciplinary approach, with engagement from interventional radiologists, immunologists, and physicians as essential to ensure correct diagnosis and treatment where SAM is considered.

#### CONFLICTS OF INTEREST

None.

#### FUNDING

None.

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