Segmental arterial mediolysis after fenestrated endovascular abdominal aortic aneurysm repair—A rare complication

Eric T. A. Lim, MB, ChB,[a] Andrew Gilkison, FRANZCR,[b,c] Hannah Elstub, FRCPA,[c] Frances Colgan, FRCR,[b] Timothy Buckenham, FRANZCR,[b] *and* Adib Khanafer, FRCS, FRACS,[a] *Christchurch, New Zealand*

ABSTRACT

Segmental arterial mediolysis (SAM) is a rare, noninflammatory, nonatherosclerotic condition that occurs commonly in mesenteric vessels. There are no known predisposing risk factors to the development of SAM. We present a case of a 67-year-old woman who presented with abdominal pain 2 days following discharge after an elective endovascular abdominal aortic intervention. Repeat imaging 2 days after readmission showed the presence of multiple new aneurysms involving the mesenteric vasculature. She underwent attempted endovascular embolization of the largest aneurysm. The postmortem and histopathologic examinations confirmed the diagnosis of SAM. (J Vasc Surg Cases Innov Tech 2024;10:101470.)

Keywords: Aneurysm; Endovascular; Hepatic; Segmental arterial mediolysis; Splanchnic

Endovascular repair of aortic aneurysms using various off-the-shelf and custom-made endografts has gained acceptance in recent years, with encouraging outcomes and lower morbidity and mortality compared with open aortic surgery. The Achilles heel of endovascular repair are endoleaks, wire injury, and contrast allergies. Segmental arterial mediolysis (SAM) has never been reported after endovascular intervention. This could be due to the rarity, unawareness, or lack of understanding of SAM.

SAM is described as a noninflammatory and nonatherosclerotic arteriopathy due to its characteristic histologic appearance.[1-4] This rare medical phenomenon mainly affects mesenteric vessels and, less commonly, the carotid and renal arteries.[4,5] Most patients with SAM tend to present with abdominal pain or massive hemorrhage, which carries a significant mortality risk.[4-6]

Current reported studies pertaining to SAM are limited to case series only, and, to date, there is still much to learn about this pathology.[5-7] We present an interesting medical mystery case that occurred after fenestrated endovascular abdominal aortic aneurysm repair and was later confirmed to be SAM. The patient's family provided written informed consent for the report ofcase details and imaging studies.

CASE REPORT

A 67-year-old woman presented for elective three-vessel fenestrated endovascular abdominal aortic aneurysm repair of a 57-mm juxtarenal abdominal aortic aneurysm. She had a history of polymyalgia rheumatica treated with 5 mg of prednisone daily, diverticulosis, and five previous laparotomy procedures and was an ex-smoker. The procedure was successful, with all three branches performed over both renal arteries and the superior mesenteric artery (SMA). The celiac trunk was not cannulated at any point during the procedure. The patient had an uneventful recovery and was discharged home on day 1 after the procedure.

Two days later, she presented to the emergency department with lower abdominal pain and diarrhea. She was hemodynamically stable. On clinical examination, she was tender over the lower abdomen but without signs of peritonism. The blood test results showed a hemoglobin of 95 g/L (normal range, 115-155 g/L), hematocrit of 0.29 (normal range, 0.35-0.46), white blood cell count of 13.4 $\times$ 10^9/L (normal range, 4-11 $\times$ 10^9/L), neutrophil count of 10.8 $\times$ 10^9/L (normal range, 1.9-7.5 $\times$ 10^9/L), C-reactive protein of 166 mg/L (normal, <5 mg/L), and lipase of 5 U/L (normal range, 10-60 U/L). A stool sample was negative for parasites and viruses. She underwent urgent computed tomography angiography of the abdomen and pelvis, which showed signs of sigmoid colitis, likely secondary to ischemia with no evidence of pneumatosis. She was readmitted and intravenous cefuroxime and metronidazole was started.

Two days following admission, her abdominal pain worsened but she remained hemodynamically stable. An urgent repeat computed tomography angiography imaging study showed the presence of multiple new pseudoaneurysms of various sizes over the hepatic artery, the SMA, and the left gastric artery (Fig 1). One of the hepatic artery pseudoaneurysms measured 30 mm.

From the Department of Vascular, Endovascular and Transplant Surgery,[a] Department of Radiology,[b] and Forensic Pathology South Island,[c] Christchurch Hospital.

Correspondence: Eric T. A. Lim, MB, ChB, Department of Vascular, Endovascular and Transplant Surgery, Christchurch Hospital, Private Bag 4710, Christchurch 8140, New Zealand (e-mail: eric_lta@hotmail.com).

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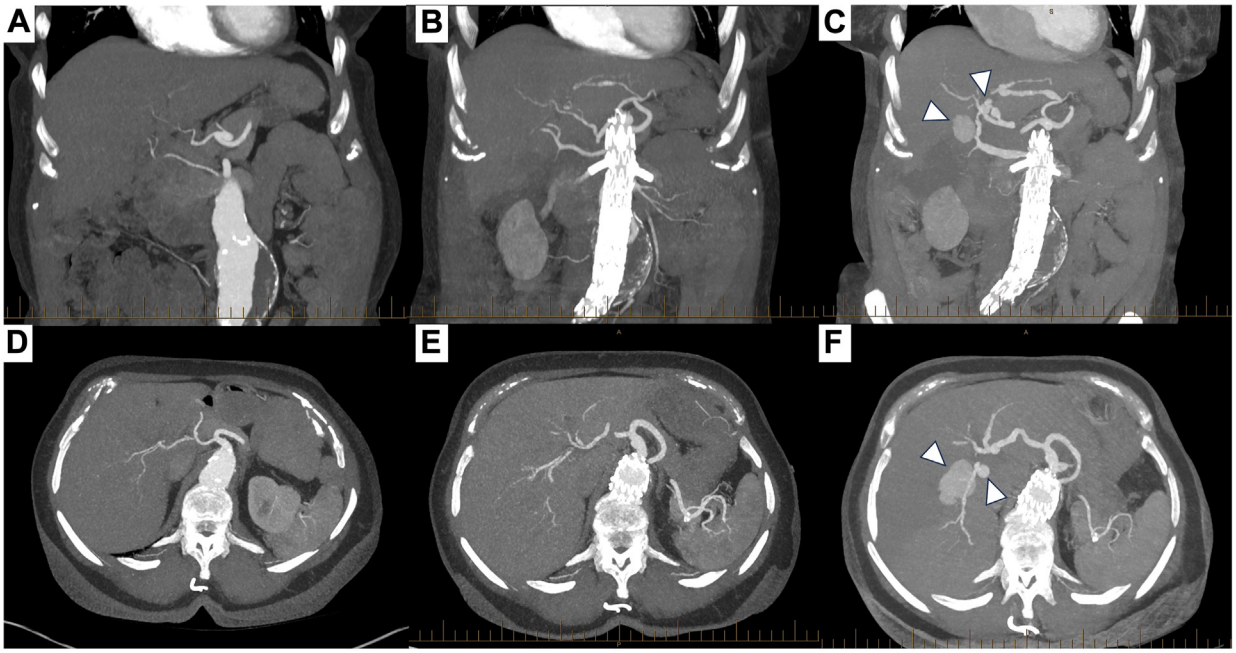

Fig 1. A, Initial preoperative coronal computed tomography angiography (CTA) demonstrating no evidence of aneurysms over the branches of the celiac trunk and hepatic artery. The preoperative juxtarenal abdominal aortic aneurysm can be partially seen at the bottom of the image. **B,** Readmission CTA over a similar slice again demonstrating no evidence of aneurysms over the branches of the celiac trunk and hepatic artery. The previously partially imaged juxtarenal abdominal aortic aneurysm has been excluded with the presence of a fenestrated endograft in the abdominal aorta. **C,** A repeat coronal CTA 2 days later over a similar slice demonstrating the presence of multiple new aneurysms over the right hepatic artery (*arrowhead*). **D,** Initial preoperative axial CTA demonstrating no evidence of aneurysms over the branches of the celiac trunk or hepatic artery. **E,** Readmission axial CTA over a similar slice again demonstrating no evidence of aneurysms over the branches of the celiac trunk or hepatic artery. A fenestrated endograft is now present in the abdominal aorta. **F,** Repeat axial CTA 2 days later over a similar slice demonstrating the presence of multiple new aneurysms over the right hepatic artery (*arrowhead*).

A wedge infarct was present in the liver. The previously noted sigmoid colitis remained unchanged in appearance.

A multidisciplinary discussion with hepatopancreaticobiliary, general surgery, vascular surgery, and interventional radiology and the patient was held. It was suspected that the pain was likely due to the finding of the multiple splanchnic aneurysms, with the largest right hepatic artery pseudoaneurysm of 30 mm the main culprit and at risk of imminent rupture. The decision was made to proceed with urgent endovascular embolization of the 30-mm right hepatic artery pseudoaneurysm. Access was gained from the left common femoral artery with a 6F sheath. There was difficulty cannulating the celiac axis through the scallop fenestration of the endograft, necessitating the use of an 8F Destino steerable sheath (Oscor Inc). The right hepatic artery pseudoaneurysm was then identified and successfully embolized under ultrasound guidance with injection of glue and lipiodol and thrombin (Fig 2). Following the procedure, the patient deteriorated intraoperatively and died. The suspected cause of death at the time was the potential rupture of the multiple visceral pseudoaneurysms.

A hospital postmortem examination was requested, which showed the cause of death was intra-abdominal hemorrhage secondary to rupture of one or more of the known splanchnic pseudoaneurysms. Histopathologic examination found evidence of medial degeneration with focal sites of mediolysis, which were found in the celiac trunk, SMA, splenic artery, hepatic arteries, and both renal arteries, consistent with the diagnosis of SAM (Fig 3).

DISCUSSION

SAM was previously known as segmental mediolytic arteritis after it was first described by Slavin in the mid-1970s.[1,2] As we delve deeper histologically, our understanding of this disease has allowed us to differentiate it from other common vasculitides or radiologic conditions and, thus, the term we use today.[1,3] It is a middle-age medical condition with a male predominance but with no proven predisposing risk factors known to date.[4-6] The only available guideline in diagnosing SAM incorporates both clinical history and imaging findings; however, the guideline has not yet been validated.[2,7]

Radiologically, SAM typically presents with six cardinal features within the affected arterial bed. These include arterial occlusion, arterial stenosis, arterial dilatation, dissecting hematoma, and the presence of single or

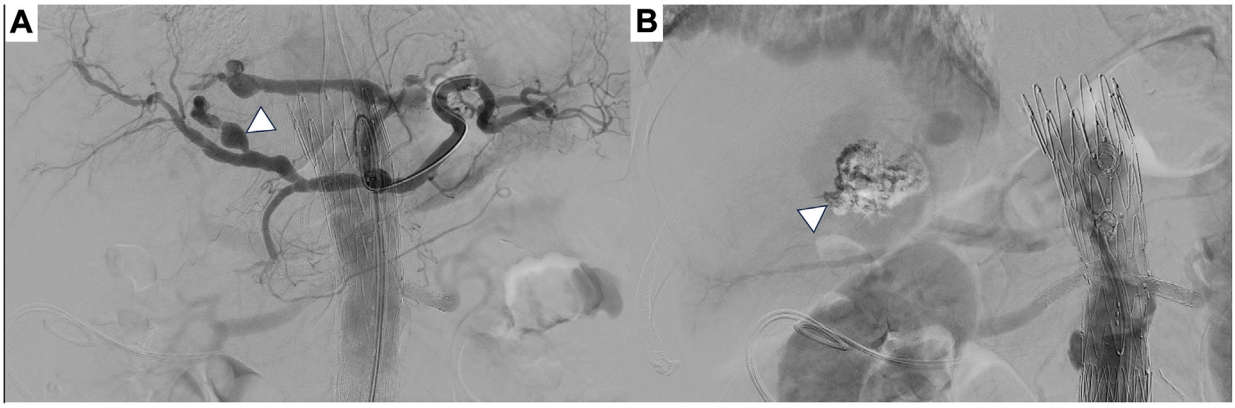

Fig 2. A, Angiography demonstrating evidence of multiple aneurysms over the branches of the celiac trunk, with the largest aneurysm over the right hepatic artery (*arrowhead*). **B,** Angiography demonstrating successful embolization of the right hepatic artery aneurysm using glue/lipiodol and thrombin (*arrowhead*).

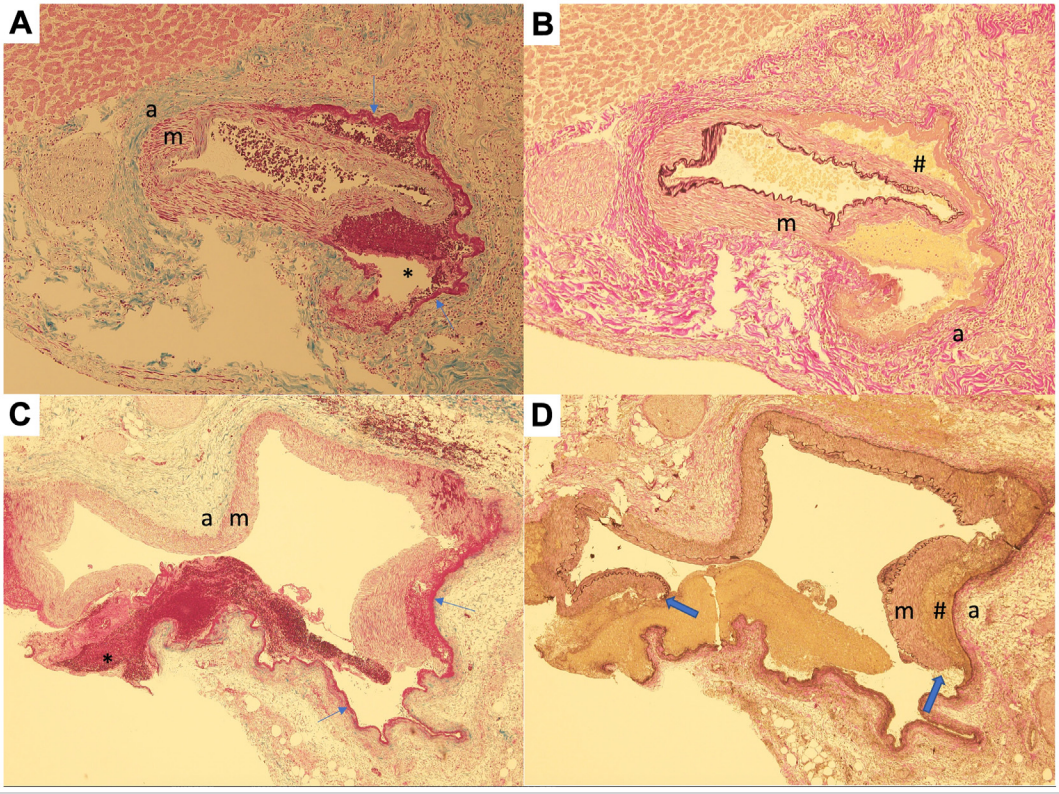

Fig 3. A, Histologic slide demonstrating an intrahepatic artery with the fibrin cap (*thin blue arrows*) and pseudoaneurysm formation (*asterisk*) at the media–adventitial interface (Masson's trichrome stain, original magnification ×100). **B,** Histologic slide demonstrating the same intrahepatic artery shown in **A,** with evidence of an intramural dissecting hematoma (*number sign*; elastin stain, original magnification ×100). **C,** Histologic slide demonstrating the right hepatic artery with evidence of fibrin capped dehiscence (*thin blue arrows*) and apparent point of rupture of the pseudoaneurysm (*asterisk*; Masson's trichrome stain, original magnification ×40). **D,** Histologic slide demonstrating the same right hepatic artery shown in **C,** with evidence of an arterial wall gap formation involving the full thickness of the media (*thick blue arrows*) with a dissecting hematoma (*number sign*) at the media–adventitial interface (elastin Van Gieson stain, original magnification ×40). *a*, Adventitia; *m*, media.

multiple pseudoaneurysms.[1,4,5] The latter three are the most common findings in the arterial bed.[1-3] Some of these findings can also be found in vasculitis, connective tissue disorders, mycotic aneurysms, and fibromuscular dysplasia (FMD). The "beaded string" appearance is commonly seen in FMD, as well as in SAM.[1,5,6] Aneurysms

in SAM do not involve the arterial bifurcation, unlike mycotic aneurysms.[7]

Histologic examination is the gold standard for the diagnosis of SAM. However, this itself is an invasive procedure, and specimens could be challenging to obtain.[1,3,5] It is understood that SAM has two distinct phases—the injurious phase and the reparative phase.[4,5,8] The injurious phase is considered the acute phase of the disease with the finding of mediolysis, which is part of its characteristic name. The resultant process leads to separation of the tunica media layer from the tunica adventitia layer.[4-6] Depending on the degree of mediolysis, this can also result in gaps in the arterial wall between portions of intact arterial wall and affected areas circumferentially[4-6] (Fig 3). The latter phase is considered the healing phase of the disease with its characteristic evidence of proliferation of granulation tissue and presence of fibrosis.[4,5,7,8]

The histologic findings in the reparative phase of SAM have resulted in studies postulating whether SAM itself could potentially be a precursor to the development of FMD.[2-5] This remains an ongoing medical investigation, with most refuting this claim due to the distinct features that distinguish SAM from FMD:[2-4,7]

- Demographic: SAM tends to present in middle-age patients with a male predominance, and FMD presents in younger patients with a female predominance.
- Location: SAM tends to affect the mesenteric vessels, and FMD tends to occur in the renal and carotid arteries.
- Radiologically: SAM tends to present in the form of dissection or pseudoaneurysm, and FMD tends to cause vessel stenosis.

As previously eluded, there are no known predisposing risk factors for the development of SAM. However, reported case series have documented a high prevalence of hypertension, followed by hyperlipidemia, in the patients with SAM.[4,6] This has potentially led to the currently applied management of optimizing cardiovascular risk factors such as good blood pressure control and the use of lipid-lowering medication in the case of an incidental finding of SAM and ongoing management after intervention.[1,4,5] Previous studies have raised the possibility that SAM could potentially be triggered in the context of sepsis, hypotension, or hypoxia.[1,4] To the best of our knowledge, our case is the first documented to occur after an elective endovascular intervention.

Despite its catastrophic complications, conservative management remains the standard of treatment of SAM, and only a limited number of patients will require intervention.[4,5] Thus, SAM could potentially be a self-limiting condition.[5] Due to the rarity of this condition, coupled with only a small proportion of patients requiring intervention, management is not straightforward. Hence, clinical assessment and determination of the hemodynamic status of each patient are warranted. Management of SAM focuses on cardiovascular risk management. Also, because SAM is not a type of vasculitis, steroids are unlikely to be of any benefit and could, in fact, aggravate the lesions in SAM.[3-5] For symptomatic patients, surgery or endovascular intervention remains the mainstay of treatment.[1,2,4]

CONCLUSIONS

SAM remains a challenging clinical condition to diagnose and treat. A histologic diagnosis is key in differentiating this condition from other vascular pathologies. This requires a greater awareness of SAM, which is paramount in the current endovascular era.

DISCLOSURES

None.

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