

Case Report

Segmental arterial mediolysis: vasculopathy presenting as acute abdominal pain

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ABSTRACT

Segmental arterial mediolysis, initially termed segmental mediolytic arteritis, is a rare noninflammatory, non-atherosclerotic vasculopathy that primarily affects visceral arteries. SAM presents with a unique skip pattern of arterial involvement and is characterized by the degeneration of the arterial medial layer, leading to various vascular abnormalities. We present the case of a fit and healthy 50-year-old Male who sought medical attention for the sudden onset of severe abdominal pain. This led to the diagnosis of SAM after repeated imaging and blood investigations. The considered differential diagnoses include fibromuscular dysplasia, vasculitis, and mycotic aneurysms. This case underscores the importance of clinical awareness and multidisciplinary collaboration in diagnosis and managing SAM, offering valuable insights into its clinical presentation and the complexities surrounding its diagnosis and treatment. Conclusively, SAM is treated conservatively by emphasizing blood pressure control, antiplatelet therapy, and anticoagulation. However, endovascular intervention is deemed necessary if there is evidence of end-organ ischemia.

Keywords: Segmental arterial mediolysis, Vasculopathy, Arteriolytic, Segmental mediolytic arteritis, Vasculitis

INTRODUCTION

Segmental Arterial mediolysis (SAM) was first described by Slavin et al back in 1976. This disease was initially known as segmental mediolytic arteritis and was subsequently renamed once the pathological process involved in this disease was identified.¹ SAM is a rare idiopathic, noninflammatory, non-atherosclerotic vasculopathy involving mostly visceral arteries of abdomen and rare involvement of the coronary, renal, iliac, cranial and pulmonary arteries, with an unknown etiology and usually demonstrated with two or more arterial segments from the same or adjoining vascular beds, typically occurring in a skip pattern.²⁻⁶ The pathophysiology of SAM mostly affects tunica media, leading to the degeneration of smooth muscle cell, causing medial disruption, intramural hemorrhage and

adventitial deposition.³ This often results in developing gaps causing aneurysms, dissections, or stenosis and may present with acute end-organ ischemia or life-threatening bleeding.^{3,4} Clinical presentation entirely depends on artery involvement and underlying pathology.² The gold standard of diagnosing SAM is through histological confirmation; however, with the recent emergence of state-of-the-art imaging modalities, such as computed tomography (CT) and magnetic resonance (MR) angiograms, the approach has shifted towards a greater use of imaging instead of histological sampling.^{2,3} SAM and other similar vasculopathies (e.g., fibromuscular dysplasia, localized vasculitis of the gastrointestinal tract, isolated dissection and aneurysm) have significant overlap, requiring the need to standardize diagnostic criteria.² Clinical recognition of SAM has increased; however, it lacks in diagnostic criteria, guidelines on management options and prognostication of the disease.^{2,5}

We present a case of SAM in a fit and healthy 50-year-old Gentleman.

CASE REPORT

A 50-years-old fit and healthy patient presented to emergency department with sudden onset of abdominal pain. Pain started while attempting to have a forceful bowel movement and radiated across upper abdomen. Patient characterized pain as sharp and stabbing in nature, constant, aggravated by movement and progressively worsening. It also radiated to left flank over time. On examination, patient had a blood pressure of 154/100 mmHg, heart rate of 92 and normal oxygen saturation level under room air.



Figure 1: Axial section of CT Abdomen indicating dissection in coeliac trunk and extending into splenic artery.



Figure 2: Axial section of CT Abdomen showing dissection extending into Common hepatic.

Abdominal examination revealed tenderness over epigastric and left upper quadrant, but the abdomen was otherwise soft, not distended, no palpable masses, and no signs of peritonitis. Blood investigations were unremarkable except for mildly raised C-reactive protein level of 38.4. However, moderate hematuria was noted on urinalysis, prompting a CT scan to exclude renal stones. Incidentally the CT scan revealed an abnormal

appearance in the distal aspect of the coeliac artery, with concern for dissection extending into common hepatic and splenic artery (Figure 1-2). There was also significant narrowing of the splenic artery, suggesting thrombus formation leading to acute pancreatitis and focal splenic infarction (Figure 3). Additionally, there was atheroma at the origin of superior mesenteric artery without any significant stenosis (Figure 4).



Figure 3: Coronal section demonstrating significant narrowing of splenic artery.

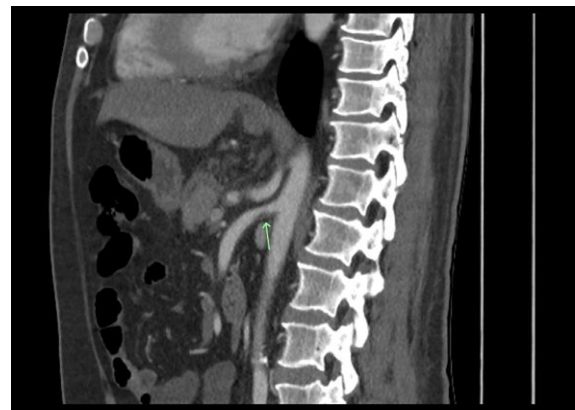


Figure 4: Sagittal section demonstrating atheroma at origin of SMA.

Conservative management commenced with a heparin infusion initiated, and strict blood pressure control. Patient was then referred to general surgery team due to acute splenic infarction and acute pancreatitis. General surgical team also decided to manage the patient conservatively as there were no further exhibited symptoms, and blood investigations remained normal. SAM was the initial diagnosis after considering the patient's history, physical examination, CT findings and ruling out other potential causes. A vasculitis screen was performed to exclude inflammatory cause of vasculopathy, but it yielded normal results. On third day of admission, the case was discussed in multi-disciplinary team meeting in which concluded that there was no indication for urgent surgical intervention. This decision

was based on the patient's improving abdominal pain, no changes in liver function tests and, no evidence of hepatic ischemia. The patient was discharged home on fourth day of admission and seen in the clinic with a repeat CTA for surveillance of aneurysmal degeneration. CT Angiogram showed appearances of coeliac trunk consistent with SAM and no further progression in dilatation. No additional imaging was planned, and patient was reassured about his symptoms and advised to seek medical attention if he experienced any further episodes of abdominal pain. He was also instructed to have regular follow-up with his general practitioner and was advised on the importance of adequate blood pressure control.

DISCUSSION

We present the case of a 50-year-old fit and healthy male presenting to our ED with abdominal pain. This case highlights the challenge in diagnosing SAM, and the importance of clinician awareness. The most common acute clinical presentation is abdominal pain, as SAM frequently involves visceral arteries of the abdomen. SAM was identified by Slavin et al. in 1976 commonly among elderly patients and was dubbed 'Segmental Mediolytic Arteritis'.^{1,7} Despite being a rare arteriopathy characterized by noninflammatory degeneration of the arterial medial layer, SAM has been increasingly diagnosed due to the refinement of cross-sectional imaging and widespread availability of focused arterial assessment methods, such as CT Angiography and MR angiography.⁴ Numerous studies have been conducted to establish epidemiology, diagnostic criteria and best treatment option. However, since most patients with this disease present asymptomatic and rarely, few reports exist with no mass studies assessing treatment outcomes.⁷ Pathophysiology of SAM is not entirely understood and has been the topic of extensive study; it is mainly guided by imaging and histologic findings.^{7,8} It is often thought that SAM is a variant of Fibromuscular dysplasia.⁴ This disease often affects medium-to-large arteries, distinguished by lysis of the tunica media and development of vacuoles in smooth muscles cell of the outer medial wall of arterial wall.^{7,9} Subsequently, this leads to formation of granulation tissue and fibrosis. Inflammatory markers such as erythrocyte sedimentation rate (ESR) and C-reactive protein are not often elevated in SAM. Patients may present with a variety of symptoms, such as abdominal pain or discomfort, hematochezia, back pain or even more severe symptoms like intraperitoneal hemorrhage and dissection.³ In this case study, it was noted that presentation was mild but persistent, leading to clinical suspicion of SAM. Gold standard of diagnosing Segmental Arterial Mediolytic is via histology.^{3,7} The presence of fibrin and collagen deposits with the destruction of smooth muscle cytoplasm of the media is suggestive of SAM. In addition, absence of inflammatory cells is a key finding that is exclusive to SAM, unlike vasculitis. However, with the recent state-of-the-art MRI and CT Angiography (CTA) imaging, SAM could be identified early as it can accurately reveal

patterns consistent with SAM. An important angiographic diagnosis is the lysis of medial layer of the arterial wall, which may result in stenosis, aneurysm formation, occlusion or even dissection.^{3,7} As in this study, medium-sized vessels were affected, which is consistent with SAM, such as dissection of the coeliac trunk and hepatic arteries, as well as narrowing of the splenic artery. Numerous studies suggested that SAM occurs in medium-sized vessels with skip patterns, affecting different segments and involving the circumference or only a part of the arterial wall, mainly tunica media.⁴⁻¹¹ Numerous differential diagnoses must be considered prior to diagnosis SAM, as there are multiple mimics. Fibromuscular Dysplasia (FMD) should be considered as it presents very similarly to SAM, often exhibiting beading or webs on imaging, posing a challenge in differentiating FMD and SAM. FMD usually affects middle-aged females with premature hypertension, which could be the main determining factor in distinguishing it from SAM, as neither favors a certain age group nor gender.^{3,7,8,12-14} Vasculitic diseases such as Takayasu's, Behcet and Henoch-Schonlein purpura often exhibit destruction of smooth muscles but are often accompanied by systemic inflammation and raised inflammatory markers, which distinguishes them from SAM.^{3,6,9} Atherosclerosis must also be considered; however, it often affects diffusely and at bifurcations, which is unlike SAM, which affects the vessel at different segments and often spares bifurcations.^{3,7,12} Other differentials to consider are Marfan and Cystic medial necrosis, which can be ruled out by characteristic clinical findings. Mycotic aneurysms should also be considered, but it always manifests with systemic infection, unlike SAM.³ SAM is often misdiagnosed due to its multiple mimics, leading to incorrect treatments. Treatment of SAM is conservative, with a focus on blood pressure control, antiplatelet therapy, or anticoagulation. Active surveillance with CT Angiography may also be suggested.^{2,3,7,9} In the event of clinical deterioration or end-organ ischemia, an endovascular approach should be the treatment of choice unless there is evidence of intraperitoneal hemorrhage, which would then warrant exploratory laparotomy.³ The patient we reported above was managed conservatively with adequate blood pressure control, antiplatelet therapy and repeat imaging, which showed no further deterioration. It is important to note that no standardized guidelines are present for the diagnosis and management of SAM, which underscores the need for randomized controlled trials.

CONCLUSION

In conclusion, SAM is a rare and complex vasculopathy that challenges clinicians due to its elusive nature and overlapping clinical features with other vascular conditions. SAM primarily affects medium-to-large arteries, leading to noninflammatory degeneration of the arterial medial layer. This case highlights the importance of clinical awareness and advanced imaging techniques in achieving accurate diagnosis, although histological

confirmation remains the gold standard. Differential diagnosis should include FMD, vasculitis, atherosclerosis, mycotic aneurysms, and Cystic medial degeneration. Treatment of SAM leans towards conservative management emphasizing on blood pressure control, antiplatelet therapy and anticoagulation. In the event of end organ ischemia, an endovascular intervention is warranted.

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REFERENCES

1. Slavin RE, Gonzalez-Vitale JC. Segmental mediolytic arteritis: a clinical pathologic study. *Lab Invest.* 1976; 35:23-9.
2. Skeik N. Segmental arterial mediolysis (SAM): Systematic review and analysis of 143 cases. *Vasc Med.* 2019;24(6):549-63.
3. Tan R. Segmental arterial mediolysis: A case study and review of the literature in accurate diagnosis and management. *Vasc Special Int.* 2019;35(3):174-9.
4. Naidu S. Segmental Arterial Mediolytic: Abdominal Imaging of and Disease Course in 111 Patients. *Am J Roentgenol.* 2018;210(4):899-905.
5. Slavin RE. Segmental arterial mediolysis: course, sequelae, prognosis, and pathologic-radiologic correlation. *Cardiovasc Pathol.* 2009;18(6):352-60.
6. Michael M, Widmer U, Wildermuth S, Barghorn A, Duewell S, Pfammatter T. Segmental arterial mediolysis: CTA findings at presentation and follow-up. *AJR Am J Roentgenol.* 2006;187:1463-9.
7. Pokharel A, Karageorgiou I, Shah S, Bhattarai M, Acharya I, Bateman J. Hepatic segmental arterial mediolysis: A case report and brief literature review. *Clin Case Rep.* 2023;11(7):e7668.
8. Bhesania N, Patil DT, McCleary B, Selvakumar PKC, Radhakrishnan K. Segmental Arterial Mediolytic Presenting as a Pancreatic Mass in a Pediatric Patient: A Case Report. *JPGN Rep.* 2023;4(2):e298.
9. Karna R, Kaye D, Jagdish B, Johnston A, Venkat D. An Unusual Presentation of Segmental Arterial Mediolytic. *ACG Case Rep J.* 2023;9(12):e00942.
10. Inada K, Maeda M, Ikeda T. Segmental arterial mediolysis: unrecognized cases culled from cases of ruptured aneurysm of abdominal visceral arteries reported in the Japanese literature. *Pathol Res Pract.* 2007;203(11):771-8.
11. Matsuura C, Fuchinoue Y, Terazono S, Kondo K, Harada N, Sugo N. Intraperitoneal hemorrhage due to segmental arterial mediolysis associated with cerebral vasospasm after subarachnoid hemorrhage. *Clin Case Rep.* 2022;10(9):e6371.
12. Gheysens G, Van Ballaer J, Maleux G. Segmental Arterial Mediolytic. *J Belg Soc Radiol.* 2021;105(1): 27.
13. Ko M, Kamimura K, Sakamaki A, Niwa Y, Tominaga K, Mizuno K, Terai S. Rare Mesenteric Arterial Diseases: Fibromuscular Dysplasia and Segmental Arterial Mediolytic and Literature Review. *Intern Med.* 2019;58(23):3393-400.
14. Alhalabi K, Menias C, Hines R, Mamoun I, Naidu S. Imaging and clinical findings in segmental arterial mediolysis (SAM). *Abdom Radiol.* 2017;42(2):602-11.

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