Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/19904
DOI URL: http://dx.doi.org/10.21474/IJAR01/19904

RESEARCH ARTICLE

SEGMENTAL ARTERIAL MEDIOLYSIS - A RARE DISEASEPRESENTED WITH COMMON SYMPTOMS

Dr. V.R Rajendran, Dr. Minsha Fathima, Dr. Nimisha Rajendran, Dr. Nuzair Ansari

Manuscript Info

Manuscript History
Received: 11 September 2024
Final Accepted: 21 October 2024
Published: November 2024

Abstract

A rare case of segmental arterial mediolysis- (SAM) presented with recurrent attacks of common symptoms like epigastric pain radiating to the back, and right lumbar pain with radiation to the groin in 55 yr old male patient. Routine investigations x-ray abdomen, ultrasound, and blood investigations are normal. NECT was non-contributory, but CECT shows circumferential soft tissue thickening that involved the celiac trunk extending into the trifurcation and the proximal part of the splenic artery, resulting in significant luminal narrowing and irregularities in the celiac artery. Focal narrow-necked saccular outpouching was noted arising from the celiac artery from its origin and directed laterally. There is no evidence of a thrombus or active leak. Focal dilatation at the bifurcation of the common hepatic artery, extending to proximal gastrodudenal artery with adequate contrast opacification and distal runoff. Reporting the features of this rare disease diagnosed by CECT.

Introduction: -

SAMis a non-atherosclerotic, non-inflammatory arteriopathy characterized by the disruption of the arterial wall's outer media layer thus leading to the formation of dissecting aneurysms, arterial stenosis, end-organ ischemia, and/or profound hemorrhage, resulting from the structural compromise of the affected vessels.[(1)]. The common clinical presentation is pain abdomen and hemorrhage, classically observed in middle-aged or elderly individuals. Computed tomography and angiographic imaging findings can overlap with those seen in various forms of vasculitis. It is essential to differentiate and exclude inflammatory arteritis to ensure proper management, as immunosuppressants generally used to treat vasculitis may be ineffective or even complicate the vasculopathy associated with SAM. Timely diagnosis is therefore vital for the effective management of this uncommon entity.

The exact aetiology of the disease is unknown, although researchers have hypothesized vasospasm and vasoconstriction as possible precursors to SAM.

Slavin and Gonzalez-vital first described in 1976. SAM is more commonly seen in men. Hypertension, hyperlipidemia, and tobacco use are the risk factors. Commonly affected vessels are, superior mesenteric 53%, hepatic 45%, celiac 36%, renal 26%, and splenic 25% arteries, with aneurysms seen in 76%, dissections in 61%, and arterial ruptures in 46%. Notably, involvement of the aorta, iliac, carotid, and intracranial arteries has also been documented in history.[(1)]

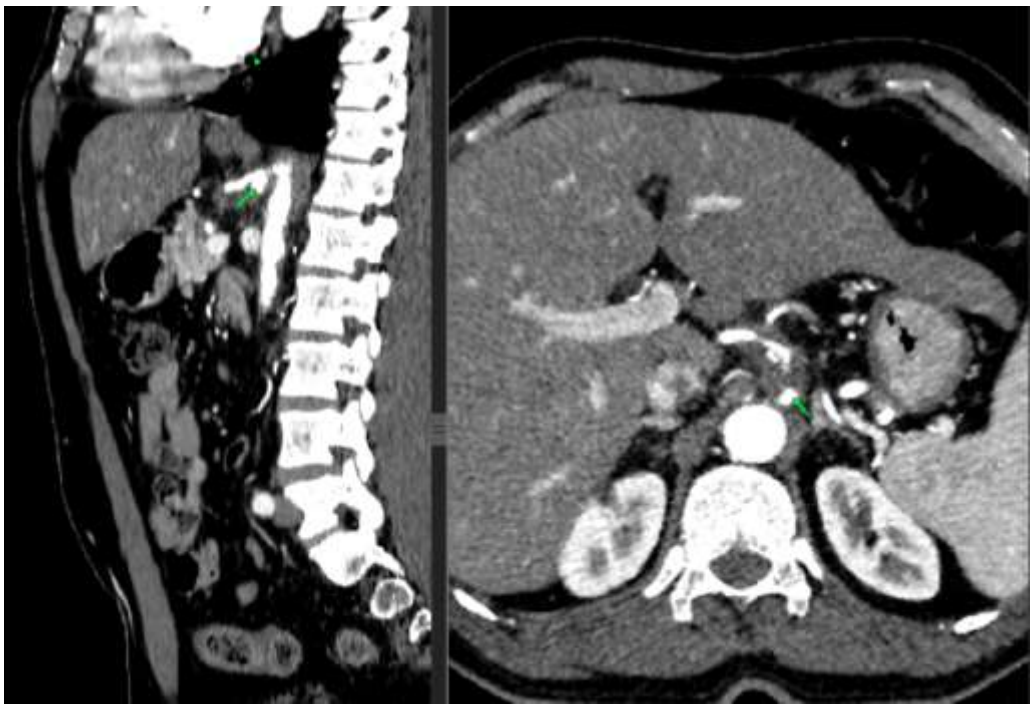
Corresponding Author:-Dr. V.R Rajendran
Address:-Professor and HOD, KMCT Medical College, Manassery, Kozhikode 673602, Kerala, India. vrajendranclt@gmail.com

Case Report

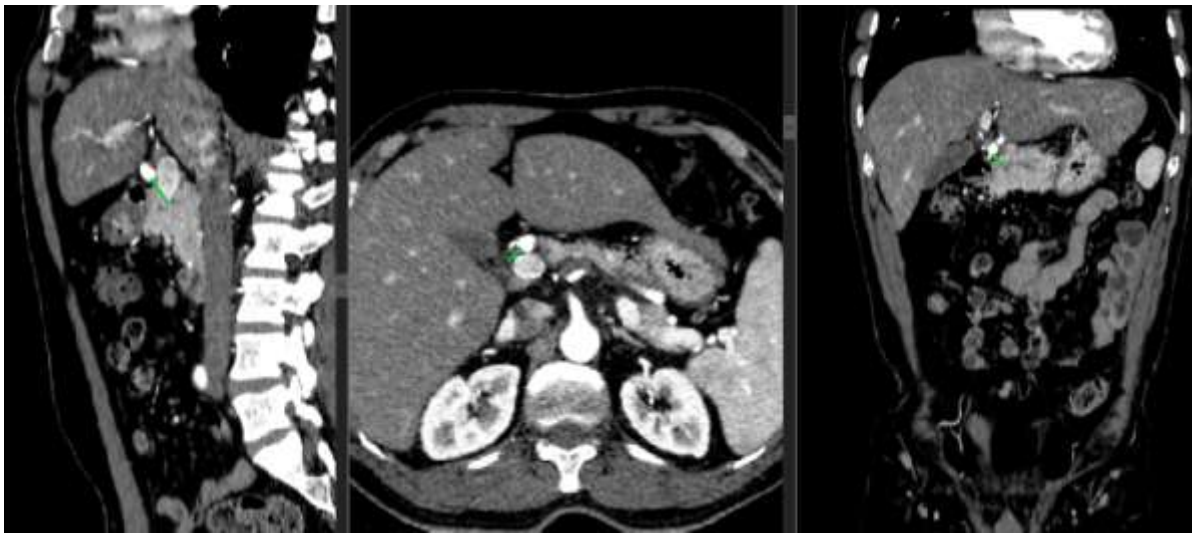
A 55-year-old Indian male, with no notable comorbidities, arrived at the emergency department reporting a six-month history of severe, episodic abdominal pain.Initially localized to the epigastric region with radiation to the back, the pain later shifted to the right lumbar area, radiating towards the groin, and is alleviated with analgesics. There was no history of fever, chills, nausea, vomiting, diarrhoea, or blood in the stool. There was no history of hypertension, diabetes, CAD, heartburn, or relevant surgical history. He had normal appetite, bowel and bladder habits. No significant family history. Based on the clinical presentation, pancreatitis and ureteric colic, as the potential diagnoses were raised.However, Blood investigations including serum amylase, serum lipase, creatinine, and urine investigations were within normal limits.

CT imaging was performed with an iodinated contrast medium. Axial coronal and sagittal sections of images were obtained. CT scan shows circumferential soft tissue thickening involving the celiac trunk extending into the trifurcation and the proximal part of the splenic artery for a length of 4.2 cm resulting in significant luminal narrowing and irregularities in the celiac artery. Focal narrow-necked 2.3 mm saccular outpouching measuring 7.2x6.3 mm noted arising from the celiac artery- 2.5 mm from its origin and directed laterally. There is no evidence of a thrombus or active leak

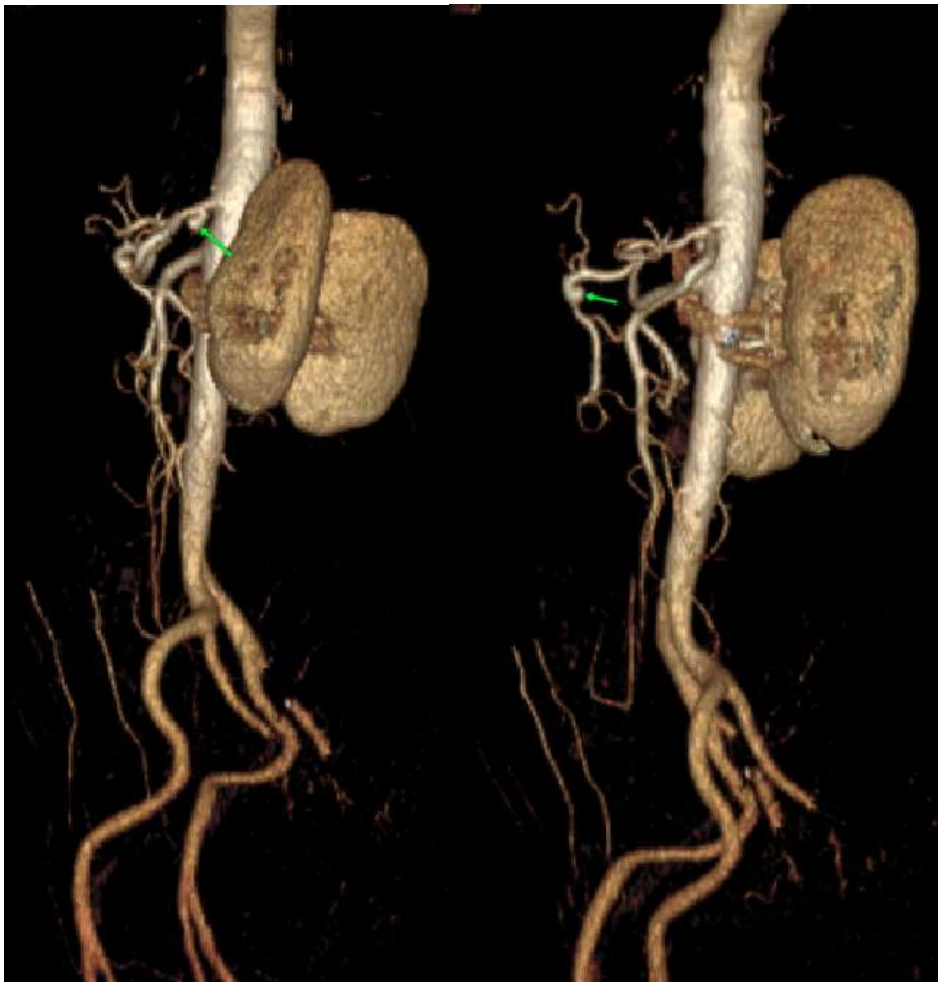
Focal dilatation at the bifurcation of common hepatic artery extending to and involving proximal gastroduodenal artery measuring 98x83 mm with adequate contrast opacification and distal runoff. No evidence of thrombus or extravasation. These findings favors the diagnosis of celiac artery dissection with aneurysmal dilatation, gastroduodenal artery aneurysmal dilatation suggesting vasculitis- SAM. The other arteries show no evidence of aneurysm luminal narrowing or occlusion. End organs like the stomach, spleen, pancreas, and liver do not show any evidence of ischemic changes or bleeding/hematoma. The bowel wall and the mesentery appear normal. No evidence of bowel wall thickening or edema, narrowing, or dilatation. The renal system, renal artery, aorta, and iliac vessels appear normal.



Sagittal and axial sections of contrast enhanced CT of the abdomen reveals circumferential soft tissue thickening seen encasing the celiac trunk, extending up to its trifurcation and up to the proximal part of the splenic artery with resultant luminal narrowing and wall irregularity of the celiac artery. There is also a focal narrow necked saccular aneurismal dilatation of the celiac trunk at its origin and directing laterally.



Similar focal aneurismal dilatation has also been noted involving the common hepatic artery at the level of its bifurcation and extends in to the proximal gastroduodenal artery.



3D reconstructed images illustrate a focal aneurysm at the origin of the celiac trunk (left), with involvement extending in to the bifurcation of the common hepatic artery extending to gastroduodenalartery (right)

Discussion:-

The first cases of this type of intra-abdominal arterial pathology was described by Slavin and Gonzales-Vitale in 1976and termed it segmental mediolytic arteritis described the (1)An identical histologic pattern of arterial "medial necrosis," first documented by Gruenwald in 1949 in the coronary arteries of newborns, is also observed in this pathological condition.[2] SAM has also been recognized as a non-atherosclerotic, non-hereditary vasculopathy, distinctly lacking an inflammatory component, as highlighted in numerous studies.[3,4,5,6]

The exact pathogenesis of SAM remains unclear; however, repeated vasoconstrictive stimuli and vasospasm have been linked to the medial lysis observed in affected vessel walls, a defining histologic hallmark of this condition.[4,8]

While histologic confirmation remains the gold standard, it is often challenging to obtain; meaning, the diagnosis of SAM is primarily based on characteristic imaging findings from CECT, CTA, and MRI, alongside clinical and laboratory exclusion of other potential diagnoses.

The pathophysiology of SAM is hypothesized to involve repeated vasoconstrictive responses within the splanchnic vascular bed, which may lead to arterial mediolysis.[4,8].

Slavin et al. identified four key diagnostic lesions of SAM: mediolysis, separation, arterial gaps, and reparative fibrosis—each playing a critical role in outlining the presentation of the disease.[1,3,4,11]. Mediolysis refers to the partial or complete vacuolization and lysis of the outer arterial media, weakening this layer of the vessel wall; thus leading to the formation of arterial gaps, patchy transmural loss of the external elastic lamina, and separation of the media from the adventitia. Consequently, dissecting hematomas and aneurysms can develop at these arterial gaps, potentially causing sudden, massive hemorrhage in the intra-abdominal or retroperitoneal space. Dissection or the presence of associated thrombi can lead to arterial luminal occlusion, which in turn causes ischemia or infarction of vital end-organs, including the bowel, liver, pancreas, kidneys, and mesenteries.[3,5,6]. The acute phase is succeeded by a late reparative phase, characterized by the formation of abundant granulation tissue which is eventually replaced by fibrosis. This process ultimately culminates in vessel remodeling, restoring a smooth and uninterrupted wall.[3,15].

SAM predominantly affects the splanchnic arteries in middle-aged and elderly individuals. While it can be clinically silent, when symptoms do manifest, they can be in the form of abdominal pain, distension, intra-abdominal hemorrhage, and hypovolemic shock. Associations can include hypotension, ischemic bowel, and vomiting.[11,12,16,17]. Less commonly, patients may present with haematuria or acute flank pain if the renal artery is involved[12,18], or Haemobilia secondary to an aneurysm eroding into the biliary system. [12].

SAM has been infrequently identified in the coronary arteries of newborns, typically in those experiencing hypoxia or asphyxia related to in-utero or immediate postpartum fetal distress.[2,3,4] SAM has also been documented in the cerebral arteries of young adults, often manifesting with severe, acute headaches lasting one to two days, followed by a sudden cerebrovascular event. Alternatively, affected individuals may present with a constellation of symptoms, including persistent dizziness, episodic hemicrania, and visual disturbances, signaling underlying vascular compromise.[7,17].

Imaging Findings

The imaging characteristics of SAM closely resemble those of various vasculitides. In fact, the radiologic presentation of SAM can be virtually indistinguishable from that of other small-to-medium vessel vasculitides, including polyarteritis nodosa, fibromuscular dysplasia, and Wegener's granulomatosis.[16]. The clinical or laboratory data help to differentiate SAM from other conditions.

SAM most commonly involves the large abdominal aortic branches[8,19].Slavin et al. has reported to document the involvement of the celiac artery and its branches, the superior mesenteric artery, renal arteries, as well as the inferior mesenteric artery and its branches.[4] Involvement of the iliac arteries, common carotids, and intracranial arteries are also not uncommon[15]

On CTA, magnetic resonance angiography (MRA), or catheter angiography, the hallmark features of SAM include alternating aneurysms and stenoses, often described as "beading," along with irregular vascular wall contours and occlusions.[13,16,20,21]. The hallmark of SAM is dissecting aneurysms and pseudoaneurysms[6,12]. Aneurysms may be fusiform or saccular[10] Focal areas of vascular dilation, accompanied by proximal stenosis, may represent a dissection or a dissecting hematoma.[17,18] Affected arteries may become elongated and kinked[1,3,14].

CT offers a few added benefits such as enabling the detection of omental, mesenteric, or retroperitoneal hematomas, along with secondary indicators of end-organ damage, such as bowel wall thickening, pneumatosis coli, pancreatitis, and renal or splenic infarction. Furthermore, it demonstrates superior sensitivity in identifying arterial wall thickening, a key feature associated with dissection.[1,4,20,22,23,24]

Management

Antiplatelet and anticoagulant agents are used. Other medications used are beta blockers, angiotensin-converting enzyme inhibitors. Antihypertensive agents are also used[18]Oral corticosteroid therapy is a safe and effective treatment option. Interventional procedures like stenting are more effective than surgery.

Conclusion:-

Segmental arterial mediolysis is a rare non atherosclerotic, noninflammatory arteriopathy, involving the medium-sized arteries, especially of the abdominal aorta and its branches of young adults or elderly patients. The presentation of the symptoms varies from asymptomatic to massive intra-abdominal hemorrhage to infarction of the end organs. The underlying pathophysiology is hypothesized to involve recurrent vasoconstriction and vasospasm of the affected vessels. As histopathological conclusion is challenging to achieve, diagnosis primarily relies upon the imaging findings of CECT, CTA, and MRA supplemented by clinical findings and lab parameters.

Acknowledgment:-

We express our sincere thanks to Dr Shabeeb Asst Professor, Dr Jinu CK Professor and others at department of Radiodiagnosis KMCT medical College Manassery Calicut for their help and support

Conflict of interest:

None

References:-

1. Slavin R E, Gonzalez-Vitale J C. Segmental mediolytic arteritis: a clinical pathologic study. Lab Invest. 1976;35:23–29. [PubMed] [Google Scholar]
2. Gruenwald P. Necrosis in the coronary arteries of newborn infants. Am Heart J. 1949;38(6):889–897. [PubMed] [Google Scholar]
3. Slavin R E, Cafferty L, Cartwright J., Jr Segmental mediolytic arteritis. A clinicopathologic and ultrastructural study of two cases. Am J Surg Pathol. 1989;13(7):558–568. [PubMed] [Google Scholar]
4. Slavin R E, Saeki K, Bhagavan B, Maas A E. Segmental arterial mediolysis: a precursor to fibromuscular dysplasia? Mod Pathol. 1995;8(3):287–294. [PubMed] [Google Scholar]
5. Armas O A, Donovan D C. Segmental mediolytic arteritis involving hepatic arteries. Arch Pathol Lab Med. 1992;116(5):531–534. [PubMed] [Google Scholar]
6. Inayama Y, Kitamura H, Kitamura H, Tobe M, Kanisawa M. Segmental mediolytic arteritis. Clinicopathologic study and three-dimensional analysis. Acta PatholJpn. 1992;42(3):201–209. [PubMed] [Google Scholar]
7. Leu H J. Cerebrovascular accidents resulting from segmental mediolytic arteriopathy of the cerebral arteries in young adults. Cardiovasc Surg. 1994;2(3):350–353. [PubMed] [Google Scholar]
8. Hashimoto T, Deguchi J, Endo H, Miyata T. Successful treatment tailored to each splanchnic arterial lesion due to segmental arterial mediolysis (SAM): report of a case. J Vasc Surg. 2008;48(5):1338–1341. [PubMed] [Google Scholar]
9. Lie J T. Segmental mediolytic arteritis. Not an arteritis but a variant of arterial fibromuscular dysplasia. Arch Pathol Lab Med. 1992;116(3):238–241. [PubMed] [Google Scholar]
10. Shimohira M, Ogino H, Sasaki S, et al. Transcatheter arterial embolization for segmental arterial mediolysis. J Endovasc Ther. 2008;15(4):493–497. [PubMed] [Google Scholar]

11. 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):771–778. [PubMed] [Google Scholar]
12. Slavin R E, Inada K. Segmental arterial mediolysis with accompanying venous angiopathy: a clinical pathologic review, report of 3 new cases, and comments on the role of endothelin-1 in its pathogenesis. Int J Surg Pathol. 2007;15(2):121–134. [PubMed] [Google Scholar]
13. Heritz D M, Butany J, Johnston K W, Sniderman K W. Intraabdominal hemorrhage as a result of segmental mediolytic arteritis of an omental artery: case report. J Vasc Surg. 1990;12:561–565. [PubMed] [Google Scholar]
14. 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(6):1463–1469. [PubMed] [Google Scholar]
15. Soulen M C, Cohen D L, Itkin M, Townsend R R, Roberts D A. Segmental arterial mediolysis: angioplasty of bilateral renal artery stenoses with 2-year imaging follow-up. J VascIntervRadiol. 2004;15(7):763–767. [PubMed] [Google Scholar]
16. Ryan J M, Suhocki P V, Smith T P. Coil embolization of segmental arterial mediolysis of the hepatic artery. J VascIntervRadiol. 2000;11(7):865–868. [PubMed] [Google Scholar]
17. Obara H, Matsumoto K, Narimatsu Y, Sugiura H, Kitajima M, Kakefuda T. Reconstructive surgery for segmental arterial mediolysis involving both the internal carotid artery and visceral arteries. J Vasc Surg. 2006;43(3):623–626. [PubMed] [Google Scholar]
18. LaBerge J, Kerlan R L. SCVIR Annual Meeting film panel sessions: case 3. Segmental arterial mediolysis (SAM) resulting in spontaneous dissections of the middle colic and left renal arteries and occlusion of SMA. Society of the Cardiovascular & Interventional Radiology. J VascIntervRadiol. 1999;10:509–513. [PubMed] [Google Scholar]
19. Molloy E S, Langford C A. Vasculitis mimics. Curr OpinRheumatol. 2008;20(1):29–34. [PubMed] [Google Scholar]
20. Rengstorff D S, Baker E L, Wack J, Yee L F. Intra-abdominal hemorrhage caused by segmental arterial mediolysis of the inferior mesenteric artery: report of a case. Dis Colon Rectum. 2004;47:769–772. [PubMed] [Google Scholar]
21. Sakano T, Morita K, Imaki M, Ueno H. Segmental arterial mediolysis studied by repeated angiography. Br J Radiol. 1997;70(834):656–658. [PubMed] [Google Scholar]
22. Wang J JL, Huang T W. Ischemic colitis caused by an isolated dissecting aneurysm of the left colic artery: a presumed case of segmental mediolytic arteriopathy. J Formos Med Assoc. 1994;93(8):715–720. [PubMed] [Google Scholar]
23. Phillips C K, Lepor H. Spontaneous retroperitoneal hemorrhage caused by segmental arterial mediolysis. Rev Urol. 2006;8(1):36–40. [PMC free article] [PubMed] [Google Scholar]
24. Frauenfelder T, Wildermuth S, Marincek B, Boehm T. Nontraumatic emergent abdominal vascular conditions: advantages of multi-detector row CT and three-dimensional imaging. Radiographics. 2004;24(2):481–496. [PubMed] [Google Scholar].