



## Case report

## Invasive investigation in cases of visceral AVM may be fatal: A rare case report

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## ABSTRACT

**Introduction and importance:** Visceral arteriovenous malformations (AVMs) are extremely rare with only a few cases described in the literature. We have encountered a mesenteric AVM in a 23-year-old girl. Considering the rarity of this entity and diagnostic dilemma, we herein describe a case of mesenteric AVM along with the review of literature.

**Case presentation:** A 23-year-old female presented with pain and lump in lower abdomen. During her workup to conclude a final diagnosis, Fine needle aspiration cytology (FNAC) was done. Post FNAC patient went into shock and immediately emergency laparotomy was done. The bleeding mass was resected along with involved gut and anastomosis was done. Histopathology suggested AVM. She was doing well at 2 months of follow up.

**Clinical discussion:** AVM is the rare cause of ischaemic colitis. It can create a diagnostic dilemma with its unusual presentation and its rarity even for both radiologists and surgeons. Usually such malformation reported after trauma or any surgical intervention, but in our case there was no such history of trauma which makes this case more interesting. Invasive investigation is recommended in such condition but needs to be very cautious. As in this index case invasive procedure lead to severe bleeding. Although definitive treatment are embolization and surgery.

**Conclusion:** Invasive procedure should be avoided in case of AVM. If angiography is not available its mandatory to keep operating room ready before any invasive procedure.

## 1. Introduction

Visceral arteriovenous malformations (AVMs) are extremely rare with only a few cases described within the literature. AVMs more often involve the hepatic (45%), splenic (30%), superior mesenteric artery (SMA), gastro duodenal arteries and very rare to find in the territory of the inferior mesenteric artery (IMA) [1]. Typical presenting clinical features include abdominal pain, lower and upper gastrointestinal bleeding, abdominal mass, portal hypertension and ischaemic colitis [2].

We have encountered a mesenteric AV malformation in a 23-year-old female. Considering the rarity of this entity and diagnostic dilemma, we herein describe a case of mesenteric arteriovenous malformation along

with the review of literature. The work has been reported in line with the SCARE 2020 criteria [3].

## 2. Case presentation

A 23-year-old female presented to our out patient department with chief complaints of pain and lump in lower abdomen for 1 month, pain was dull aching in nature, sudden in onset, without aggravating or relieving factors with alternate episodes of loose stools. There was no history of blood present in the stool. On examination there was a palpable globular mass of 15x15cm in hypogastric region which was extending from midline to right iliac fossa with smooth surface, firm consistency, indistinct lower margin, and restricted mobility in all

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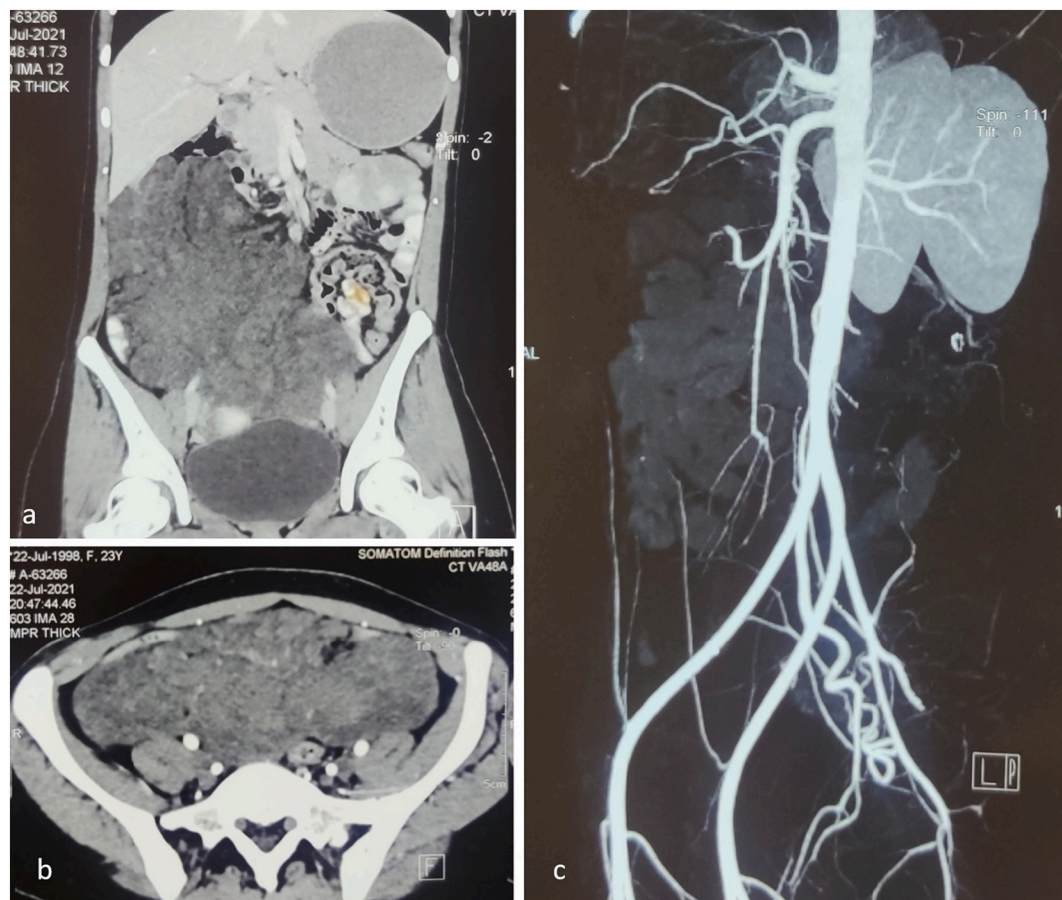
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planes. There was no hepato- splenomegaly. On CT angiography of abdomen there was a large infiltrative mass measuring  $20 \times 18.7 \times 8.2$  cm within the mesentery extending from the sub-diaphragmatic region up to the pelvis. Anteriorly it was abutting the anterior abdominal wall, posteriorly abutting the retroperitoneal vascular structures including aorta and IVC (inferior vena cava). Inferiorly it was seen abutting the dome of the bladder. The mass was encasing the SMA and its distal ileo-colic branches and arterial arcade along the caecal, and hepatic flexure regions. The adjoining SMV and tributaries were also seen encased by the same. The IMA was displaced by the mass. The small bowel loops and colon were displaced left laterally. The mass was abutting the right common iliac artery along its entire length, however with maintained intervening plane. The mass showed no evidence of post contrast enhancement and few tiny foci of calcifications within (Fig. 1). Our preliminary diagnoses were Fibrous tumour of the peritoneum, hemangiopericytoma, Pseudomyxoma peritonei, and Desmoid tumour. Considering the clinical symptoms including pain abdomen, and increase frequency of stool, the preliminary diagnosis of ischemic colitis cannot be excluded. To confirm the diagnosis, we performed a CT guided fine needle aspiration biopsy. Post procedure patient had severe abdominal pain and per abdomen was tender with drop in haemoglobin. Emergency exploratory was performed by an additional professor and his team with 12 years of experience in surgical disciplines. Intra operatively there was a 25x30cm multi-lobulated mass arising from the mesentery at the level of jejunum and proximal ileum, just abutting the ileo-colic vessels (Fig. 2). We dissected the ileo-colic vessels, ligated at the base of tumour and 40 cm of small bowel was resected with tumour with the help of GI stapler followed by side to side jejuno-ileal anastomosis. Two drains were placed in both sides of pelvis and abdomen. On

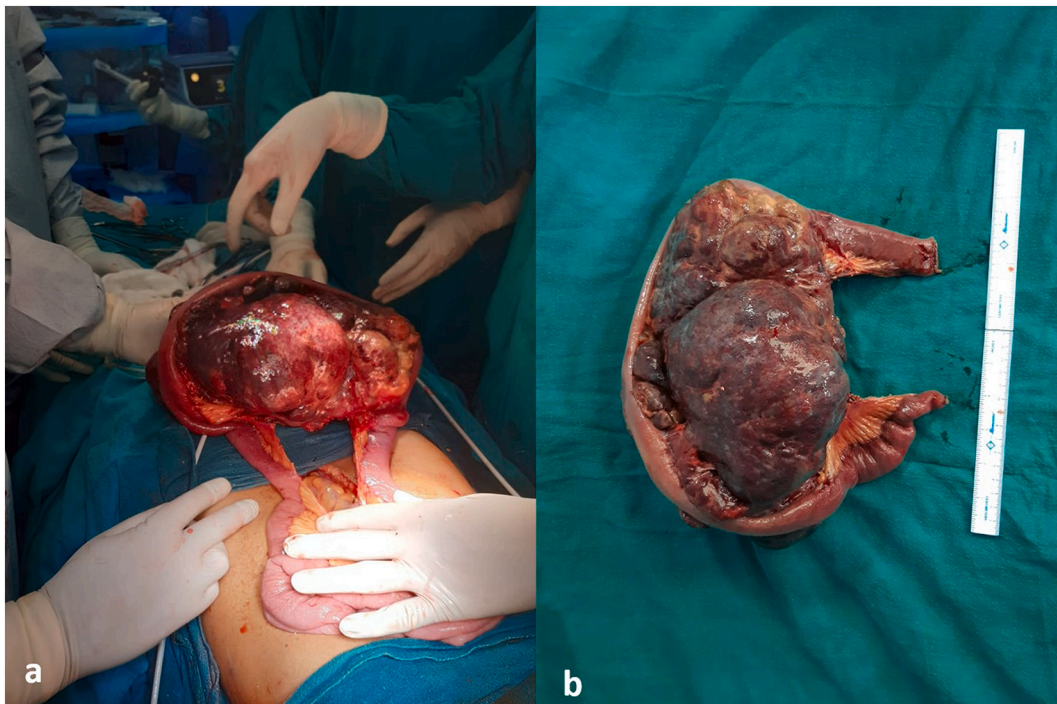
histopathological examination it was a well circumscribed tumour present on mesenteric side with proliferation of variably sized intercommunicating dilated and congested thin and thick walled vessels with intervascular stroma predominantly composed of fibro fatty tissue with small lymphoid collection along with vascular channels comprised of arterial, venous and lymphatic system. IHC (Immunohistochemistry) was positive for CD31 and CD34 for endothelial cells and D240 for lymphatics, which confirm the diagnosis of arteriovenous and lymphatic malformation (Fig. 3). After 2 months of follow up with ultra sound abdomen, patient is doing well.

### 3. Discussion

Ischaemic colitis (IC) is the most prevalent type of gastrointestinal ischemia (50–60% of all episodes) [4,5]. Its real incidence is probably underestimated because many patients suffer only mild or transient damage that remains undiagnosed. Females have more predilection for IC and more in elderly patients, majority of them being over 60 years of age. Patients with IC frequently present with co-morbid conditions, but in this index case patient belonged to young age group and she didn't have any co-morbidities. IC is frequently classified as occlusive and non-occlusive with the latter being the predominant mechanism [6] and AVMs are an exceptionally rare cause of IC. There are various factors which make colon prone to ischaemic changes like reduced blood supply per 100 g tissue, watershed areas such as splenic flexure and recto sigmoid junction, poor auto regulation during hypotension. The suggested mechanism through which an AVM causes IC is a combination of reduced arterial blood flow to the mucosa secondary to a steal phenomenon and submucosal oedema due to venous hypertension [7].



**Fig. 1.** (a, b, c): a) large infiltrative mass measuring  $20 \times 18.7 \times 8.2$  cm is seen within the mesentery extending from the sub-diaphragmatic region up to the pelvis b) Anteriorly it is abutting the anterior abdominal wall, posteriorly abutting the retroperitoneal vascular structures aorta and IVC c) Tortuous mesenteric vessels.



**Fig. 2.** (a, b): Multi-lobulated mass of size 25 × 30 cm arising from mesentery at the level of jejunum and proximal ileum, just abutting the ileo-colic vessels.

Primary mesenteric AVMs are congenital and/or idiopathic and differ from secondary or acquired AVFs that are commonly caused by blunt or penetrating trauma (bullet or knife) or have iatrogenic aetiologies [8–11]. Congenital AVMs result from undifferentiated embryonic vessels failing to regress and interconnect the arterial and venous system. They are distinguished by a dilated feeding artery, a large tangle of vessels representing the AVM nidus with multiple arteriovenous connections and a densely opacified early draining vein or veins [8]. The incidence of such AVM in early age is not common. Table 1 showed that in last 8 years except one case all gastrointestinal AVMs were reported in older age group. In our case it was of 23 year old female. In this index case, the congenital origin of the AVM was the most probable cause, because our patient was absolutely free of any previous surgical procedure or trauma. The CECT abdomen usually demonstrates the enhancement effect but in present case because of major lymphatic component, there was slow flow within the mass which in fact is the main reason of no contrast enhancement [12]. The treatment of inferior mesenteric AVMs is complex and needs patient-specific multidisciplinary approach. It involves either endovascular embolization or surgical intervention. Embolization is successful in most cases described in the literature either as a bridge prior to operative management or as a definitive treatment. It is thought to be safer and to reduce the risk of intraoperative blood loss. Percutaneous endovascular arterial embolization of the feeding artery at the artery–venous junction is the technique of choice [13]. However, it carries the risk of ischemia and passage of embolization material into the portal circulation [14]. So, embolization is not recommended in fistulas with large vessels because of the increased risk of extensive arterial thrombosis and ischemia. Furthermore, collateral formation often follows embolization if surgical intervention is not performed shortly after [15].

AVM is one of the rare causes of IC that should be considered before any invasive investigation. Whenever there is a diagnostic challenge and an invasive investigation needs to be done we should consider AVM as one of the differentials and we should be adequately prepared to deal with possible haemorrhage and other complications.

#### 4. Conclusion

In the clinical setting of ischemic colitis without any demonstrable etiology, we should consider for mesenteric angiography with keeping AVM as one of the differentials. If angiography is not available we should be ready with operating room before any invasive investigations.

#### 5. Patient's perspective

It was a life time experience. After piercing the needle over my abdomen, my condition was so bad and I thought that I will not survive now. Fortunately I had a successful surgery and got a second life. Definitely I would like to give many many thanks to treating doctor and their team.

#### Provenance and peer review

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#### Ethical approval

There is no ethical approval was obtained as it's a case report.

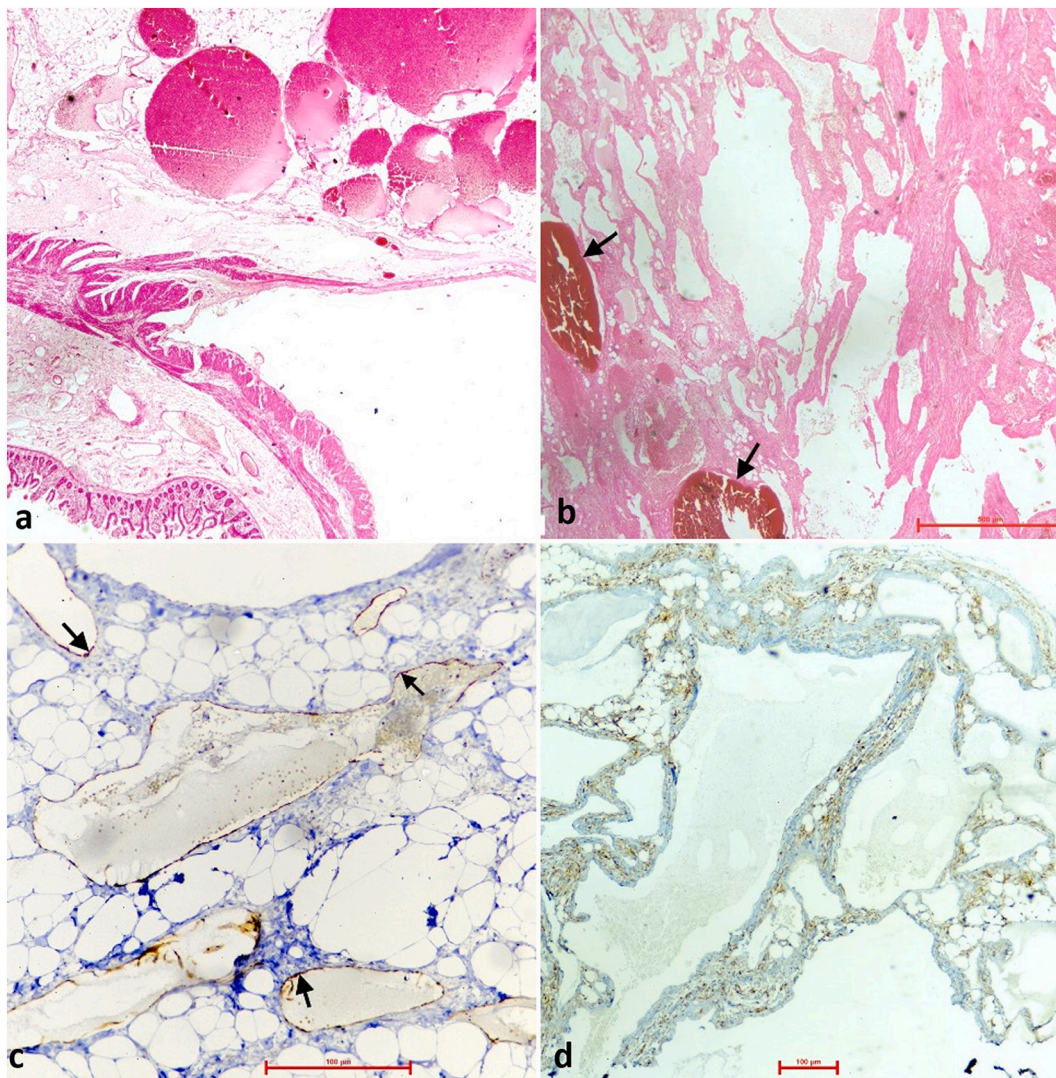
#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-chief of this journal on request.

#### Registration of research studies

Not applicable.





**Fig. 3.** a–d (H & E stain, 100× magnification): A lesion arising from the serosal aspect of the intestine, composed of varying-sized dilated and congested vessels. The lesion involves the muscularis propria. The attached intestinal segment appeared normal (a). Multiple, varying-sized blood and lymph vessels with a few thrombosed capillaries (black arrows) are present in an interconnecting manner (b). D2-40 Immunostain highlights the endothelial lining of the interspersed dilated and irregular lymphatic channels (Black arrows) (c). CD34 immunostain highlighting the vessels with interspersed fat cells nuclei also showing positivity (d).

**Table 1**

Recent case report on AVM of gastrointestinal tract.

S. No	Age (Yrs)	Nature of presentation	Diagnostic modality	Treatment offered	Reported by
1	69	GI Bleeding	Double ballon endoscopy	Laparoscopy small bowel resection	Fujiii etal (2014)
2	34	Hematemesis, Melena, and fatigue	EUS (Left gastric vessels)	Embolization of left gastric vessel	M. Parikh (2017)
3	62	GI Bleeding and bloody stool (Intermittently)	Multiple phase CT and Angiography	Segmental small bowel resection	M.Hirakawa etal (2019)
4	39	Abdominal pain and nausea	CT and Angiography	Endovascular embolization	EC Bandel (2019)
5	48	Anemia (Low haemoglobin)	Endoscopy, Capsule endoscopy, and angiography	Laparoscopic surgery using IV injection of ICG	T Hyo etal (2020)
6	12	Bright red coloured bleeding per rectum	Low haemoglobin, and CT Scan	Resection and anastomosis of the ileal segment	Govindarajan K.K etal (2021)

Comparing the recent few case reports in terms of nature of presentation, diagnostic modality, and treatment offered.

**Guarantor**

Dr. Anil Kumar.

**CRedit authorship contribution statement**

Dr. Sreepriya P P - study concept, design, writing the paper.  
 Dr. Anil Kumar – study concept, design, operated and writing the paper.  
 Dr. Shiv Shankar Paswan- operated the case.

Dr. Utpal Anand- operated the case.  
 Dr. Shreekant Bharti- data collection.  
 Dr. Rahul Ranjan- Review literature.

#### Declaration of competing interest

All authors have nothing to disclose.

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