

Case Report

Ruptured inferior mesenteric artery aneurysm in a patient with neurofibromatosis type I and its management

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Received: 02 December 2017

Accepted: 28 December 2017

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ABSTRACT

Inferior mesenteric artery aneurysm is very rare and are usually asymptomatic. A high mortality is associated with aneurysmal rupture. The purpose of this paper is to report occurrence of a rare disease and its treatment options. A 64 years old woman presented with complaints of lower abdominal pain radiating to back for 1month, with non-passage of stools for 5days, and had decreased appetite. She had features of Neurofibromatosis type I. On examination, she had tachycardia and had pallor, and abdominal examination showed lumps in right and left iliac fossa which progressively increased in size. Computed tomography scan showed features of leaking inferior mesenteric artery aneurysm with perianeurysmal hematoma. The patient was taken up for surgical exploration wherein after checking the bowel viability, the inferior mesenteric artery was clipped at its origin, ligated and divided followed by hematoma evacuation. Post-operative stay was uneventful, and the patient was discharged on post-operative day 10.

Keywords: Inferior mesenteric artery aneurysm, Neurofibromatosis type 1, Ruptured aneurysm

INTRODUCTION

Neurofibromatosis (NF; von Recklinghausen disease) is an autosomal dominant trait that has a rare but well-documented association with visceral arteriopathy.¹

Aneurysms of splenic, hepatic as well as superior mesenteric artery (SMA) have been reported, however aneurysms of the inferior mesenteric artery (IMA) are very rare, representing less than 1% of all visceral arterial aneurysms.² Most of them are asymptomatic, but when they present as a rupture, a high mortality is associated. Only 2 cases of IMA aneurysm associated with NF type I have been reported in the literature till date.³

His purpose is to present a rare case of a ruptured inferior mesenteric artery (IMA) aneurysm in a woman with neurofibromatosis type 1 and its successful management.

METHODS

A 64 years old woman presented with complaints of dull aching lower abdominal pain radiating to back for 1month, with non-passage of stools for 5days and decreased appetite. There was no history of trauma or previous abdominal surgery. She had history of pulmonary tuberculosis for which she completed category II anti-tubercular treatment in 2009. She had suffered a cerebro-vascular event in 2012 which was managed medically and had recovered by time of presentation. On presentation, she had a pulse rate of 110 per minute, blood pressure of 112/88mmHg and pallor was present. Multiple cutaneous neurofibromas were present over limbs and trunk with multiple hyperpigmented macule over the trunk and back suggestive of NF type I. On abdominal examination, an ill-defined lump of size 2X2 cm was present in the right iliac fossa which was mildly tender, non-mobile with smooth surface and non-

pulsatile. On digital rectal examination (DRE), an external haemorrhoid at 11'o clock was present with no active bleeding and anal tone was increased. On initial investigations, her haemoglobin (Hb) was 4.7gm/dL, total leucocyte count (TLC) of 26000/cu.mm. Ultrasound of abdomen showed localized collection in pericaecal region with internal echoes and bowel loops seen adherent to the collection. Abdominal x-ray was normal.

With a provisional diagnosis of appendicular lump, patient was managed conservatively on nil per oral, intravenous (IV) antibiotics, analgesics and transfusion of 2 units of packed cells. On re-evaluation after 24hours, her pulse was 108/minute, BP was 120/84mmHg, and the lump size doubled to 4X5cm and another lump was palpable in the left iliac fossa of size 10X10cm which was non-tender and non-pulsatile. On DRE, an extraluminal mass was felt 4-5cm from the anal verge causing luminal compromise, the mucosa was normal. Her Hb increased to 7.2gm/dl which later on decreased to 6.0gm/dl. An urgent contrast enhanced computed tomography (CECT) scan of abdomen showed a large saccular aneurysm arising from IMA with large perianeurysmal hematoma extending into bilateral posterior pararenal retroperitoneum and pelvis with hematoma in right psoas major muscle with mild free fluid in abdomen suggesting a leaking inferior mesenteric artery aneurysm (Figure 1, 2).

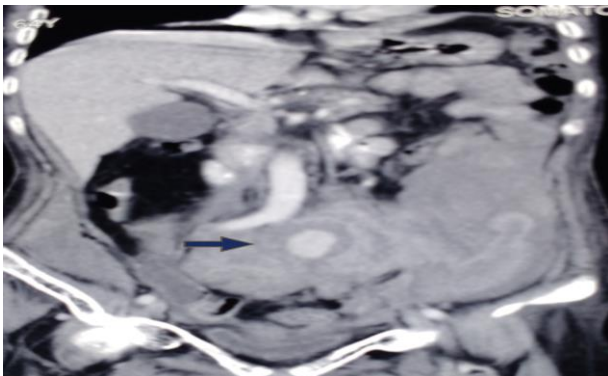


Figure 1: CECT abdomen showing leaking IMA aneurysm (arrow).

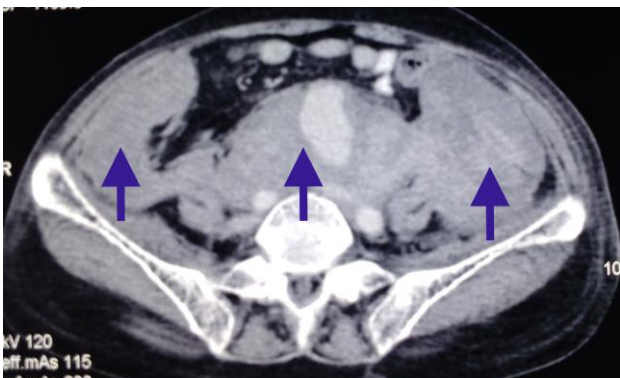


Figure 2: CECT abdomen- showing leaking aneurysm (middle arrow) and hematoma (side arrows).

On angiography, superior mesenteric (SMA) and renal arteries were visualized and found to be normal, but IMA could not be visualized, and the procedure was abandoned. In view of falling Hb, patient was taken up for exploratory laparotomy via midline trans-peritoneal approach. On exploration, there was a large hematoma in left paracolic gutter and surrounding the aorta and overlying bilateral psoas muscle. Origin of inferior mesenteric artery was identified; no active bleeding seen and was occluded using vascular clamp (Figure 3, 4).

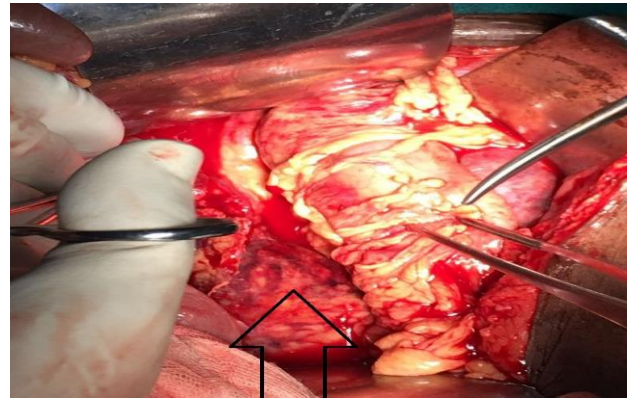


Figure 3: Intra-operative - hematoma surrounding IMA (arrow).

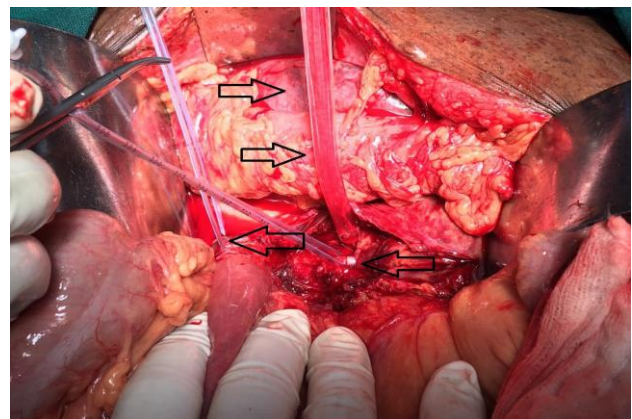


Figure 4: Intra-operative- showing hematoma, colon, SMA (slinged) and IMA (clipped).

No discoloration of the bowel was seen and then IMA was ligated and divided. Early post-operative management was in ICU where her recovery was steady and uneventful. The patient was discharged after 10 days and she follow up has been uneventful.

DISCUSSION

Inferior mesenteric artery aneurysm is the rarest among the visceral artery aneurysms (<1%), the common ones being splenic (60%), hepatic (20%), SMA (6%), coeliac (4%), gastric and gastroepiploic arteries (4%), jejunal, ileal, and colic arteries (3%), duodenal and pancreatic arteries (2%), gastroduodenal artery (1.5%).²

Atherosclerotic disease is the most common cause, other rare causes include polyarteritis nodosa, Takayasu's disease, tuberculosis and Neurofibromatosis.⁴

Most cases are asymptomatic, and symptomatic patients usually have abdominal pain, low back pain, pulsatile abdominal mass, hemorrhagic shock or collapse. Total of 54 cases of IMA aneurysm have been reported out of which 11 were associated with spontaneous rupture/leak, all of whom presented in shock, out of which 4 resulted in death.⁵

Current consensus is intervention for all aneurysms >2cm at the proximal end and 1cm at the distal end of IMA due to increased risk of rupture.⁵ Percutaneous transcatheter coil embolization techniques are being used in increasing frequency. Nevertheless, a high incidence of aneurysmatic sac reperfusion and a relatively high morbidity are associated with this procedure.⁶ In cases where percutaneous techniques fail, surgical repair is indicated and where repair is not possible simple ligation of IMA can be done if patency of SMA and celiac trunk is maintained. Neurofibromatosis has been reported as a rare cause of visceral artery aneurysm due to the fibromuscular dysplasia leading to narrowing and dilatation. Though vascular involvement is common (most common being renal artery stenosis), visceral artery aneurysms are relatively rare.^{7,8}

CONCLUSION

Author experienced a rare case of ruptured IMA aneurysm in a patient with NF-1. In a stable patient, angioembolization may be considered as a definitive management. However, in emergency cases or in cases of failed angioembolization, urgent surgical repair should be considered. Timely diagnosis and management is crucial for survival.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Huffman JL, Gaetan V, Bowers VD, and Mills JL. Neurofibromatosis and arterial aneurysms. Am Surg. 1996;62:311-4.
2. Messina LM, and Shanley CJ. Visceral artery aneurysms. Surg Clin North Am. 1997;77:425-42.
3. Hartmann E, Johnstone JK. Endovascular treatment of a ruptured inferior mesenteric artery aneurysm in a patient With Neurofibromatosis type 1. Jour Vasc Surg. 2015;6:84S-5S.
4. Grego FG, Lepidi S, Ragazzi T, Iurilli V, Stramana R, et al. Visceral artery aneurysms: a single center experience. Cardiovasc Surg. 2003;11:19-25.
5. Edogawa S, Shibuya T, Kurose K, Sasaki K, Tomita H. Inferior Mesenteric Artery Aneurysm: Case Report and Literature Review. Ann Vasc Dis. 2013;6:98-101.
6. Chiesa R, Astore D, Guzzo G, Frigerio S, Tshomba Y, Castellano R, et al. Visceral artery aneurysms. Ann Vasc Surg. 2005;19:42-8.
7. Sacar M, Tulukoglu E, Ucak A, Guler A, Yilmaz AT. Inferior mesenteric artery aneurysm combined with renal artery stenosis in a patient with neurofibromatosis. Perspect Vasc Surg Endovasc Ther. 2006;18:217-20.
8. Oderich GS, Sullivan TM, Bower TC. Vascular abnormalities in patients with neurofibromatosis syndrome type I: clinical spectrum, management, and results. J Vasc Surg. 2007;46:475-84.

Cite this article as: Sheth S, Kansakar N, Agarwal P, Singh R, Mishra A. Ruptured inferior mesenteric artery aneurysm in a patient with neurofibromatosis type I and its management. Int Surg J 2018;5:732-4.