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SUCCESSFUL AND STENTING OF THORACO ABDOMINAL IN A CASE OF MEDIAN ARCUATE LIGAMENT SYNDROME

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Keywords: *MALS, aortic stenting, endovascular repair of MALS, MALS and staged interventions.*

Abstract

Median Arcuate Ligament Syndrome (MALS) or Celiac Artery Compression Syndrome (CACS) is a rare clinical entity, and presents as symptoms of acute intestinal obstruction. CT angiography is diagnostic in a hemodynamically stable patient. Treatment modalities of choice still remain controversial. In patients with acute presentations it is usually managed by surgical (exploratory laparotomy/ laparoscopic) repair or endovascular repair or combined interventions.

We hereby present a case of 33 years old gentleman who was diagnosed to have acute intestinal obstruction secondary to MALS and underwent emergency exploratory laparotomy, adhesiolysis and release of median arcuate ligament followed by angioplasty and stenting of the Thoracic Aorta.

Introduction

MALS is a rare clinical entity caused by anatomical location of median arcuate ligament and was first described in 1963. Patients may present symptoms suggestive of acute intestinal obstruction; mesenteric ischemia or it can be an incidental finding. CT angiography remains the investigation of choice. The choice of treatment (surgical or endovascular interventions) remains controversial, further study to elucidate the role of endovascular intervention in such cases needs to be done. However, in a patient presenting with symptoms of acute abdominal obstruction, exploratory laparotomy and release of MAL should be done and later on the vascular problem can be tackled.

Case report

A 33 years old gentleman was electively admitted for the management of the distal thoracic aorta stenosis secondary to Median Arcuate Ligament Syndrome (MALS). He gave history of bilateral gluteal claudication (left more than right) and erectile dysfunction for 7 months. He had no complains of calf claudication, parasthesia or weakness of the lower limbs. He was a non-smoker with no history of bronchial asthma, or any thromboembolic events (stroke, MI, amaurosis fugax, limb ischemia, DVT, pulmonary embolism) in any other systems in the past.

General examination of the patient revealed a conscious, oriented and afebrile patient with heart rate 85/min, B.P-220/110 mmHg in the upper limb, and SBP not recordable in bilateral lower limbs and respiratory rate 20/min. Bilateral lower limbs pulses was very feeble with definite radio femoral delay in both sides. Cardiac examination revealed bruit and systolic murmur at the back. Abdominal examination and other systemic examination were unremarkable.

1-month prior to this presentation, he had acute complains of continuous diffuse abdominal pain associated with vomiting and obstipation for almost 40 hours. General examination was normal but systemic examination had revealed diffuse tenderness, guarding and absent bowel sounds.

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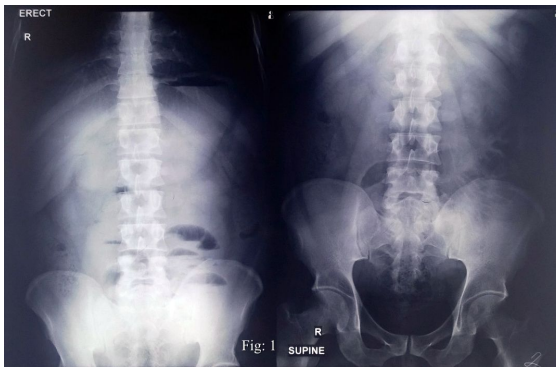

Fig: 1 – X-ray revealed few air fluid level under the diaphragm

X-ray abdomen (erect) (Fig: 1) showed few air fluid levels under the diaphragm and CT abdomen revealed dilatation of proximal small bowel lumen (2.2-3.4cm) and collapsed distal small bowel and entire large bowel lumen. There was focal stenosis of the proximal abdominal aorta for a length of approximately 24mm and focal stenosis of proximal celiac axis with hooked appearance suggestive of MALS. CT angiogram of the abdominal aorta and its branches revealed smooth narrowing of the proximal abdominal aorta with 83% area of stenosis at the narrowest segment involved. Kinking and narrowing of the proximal segment the celiac trunk were noted at the arcuate ligament level with about 87% area of stenosis. Rest of the celiac trunk, Common hepatic artery, right and left hepatic artery, gastro-duodenal artery, left gastric artery, splenic artery and cystic artery were normal in course and calibre.

With clinical features and CT abdomen/angiogram suggestive of sub acute small bowel obstruction (mechanical) secondary to MALS he underwent emergency exploratory laparotomy, adhesiolysis and release of median arcuate ligament. His postoperative period was uneventful and planned for revascularization of the aortic stenosis in staged manner.

CT angiogram (1 month after surgery) at our center (Fig 2- 4), revealed luminal narrowing of the distal thoracic aorta, for a length of approximately 6.8 cm. Tightest point noted was just proximal to the diaphragmatic hiatus with luminal calibre of approximately 7 mm. The aortic calibre at proximal segment of the luminal narrowing was approximately 13.8 mm and the distal segment of the luminal narrowing was approximately 16.1 mm. The *Celiac trunk* and its branches, Superior Mesenteric artery and its branches, Inferior Mesenteric artery and its visualized branches, Renal arteries on both side, bilateral Common Iliac arteries, bilateral External & Internal iliac arteries were normal.

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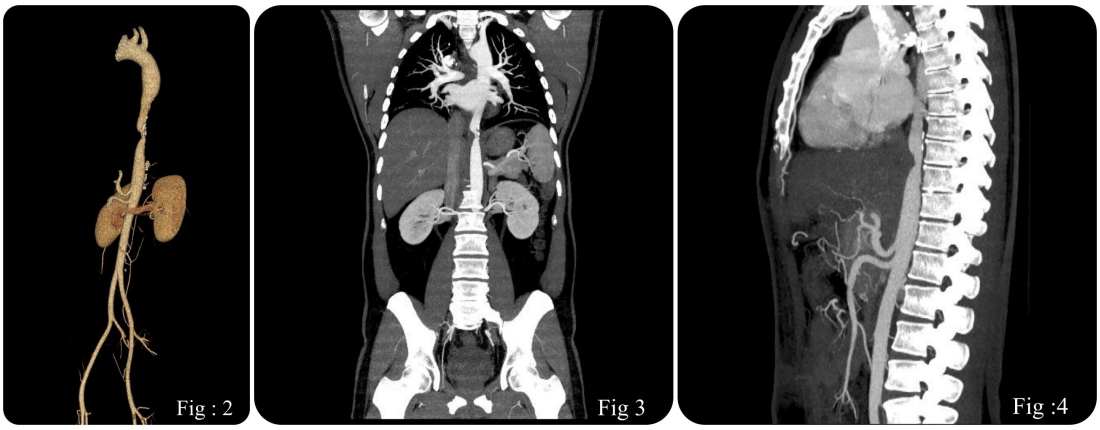

(Fig 2- 4: luminal narrowing of the distal thoracic aortic calibre, with maximum narrowing just proximal to the diaphragmatic hiatus. Normal Celiac trunk and its branches, SMA and its branches, IMA and its visualized branches, Renal arteries on both side, bilateral CIA, bilateral EIA AND IIA).

As per the management plan, the patient was taken for aortic angioplasty and stenting. Both left radial and right femoral artery puncture was done and gradients measured. Pressures proximal to stenosis were 240/110mmHg and distal to stenosis were 110/82mmHg with peak-to-peak pressure gradient of 130mmHg. After this two Proglide XL (ABOTT VASCULAR) were inserted in the right femoral artery and 14F COOK sheath was inserted. The lesion was crossed with TERMO wire and exchanged with Amplatz super stiff wire (Fig: 5), the tightest segment was pre dilated with 9x40mm balloon (RIVA)(Fig: 6) and stented with 18x45mm CP stent (NUMED) (Fig: 7, 8). Post angioplasty and stenting (Fig: 9, 10) the proximal pressure was 187/106mmHg and distal pressure was 183/104 mmHg with peak-to-peak pressure gradient of 4mmHg. All catheters were removed and the femoral artery was closed with PROGLIDE XL (ABBOTT VASCULAR) as recommended.

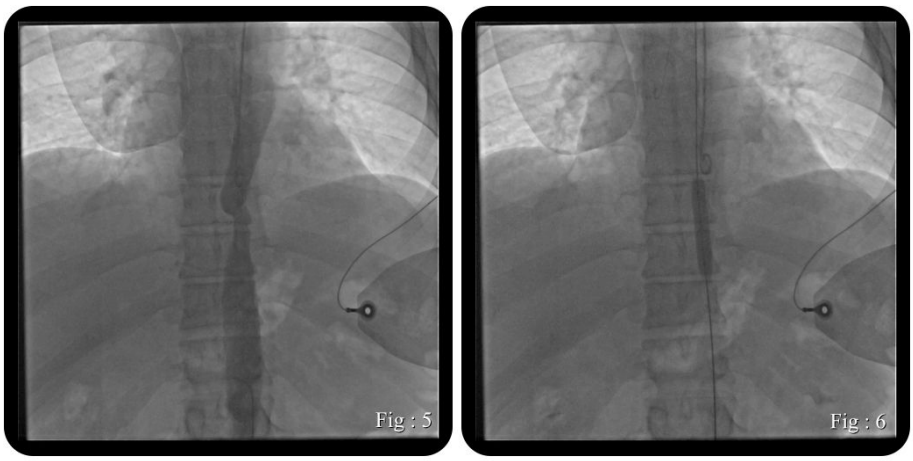

Fig 5, 6 - lesion was crossed and Amplatz super stiff wire was inserted (Fig: 5), the lesion was pre dilated with 9x40mm balloo

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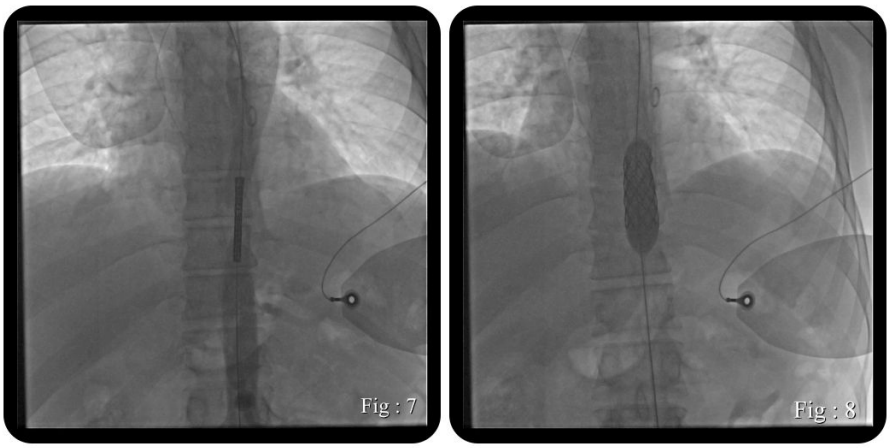

Fig: 7, 8 – lesion stented with 18x45mm CP stent (NUMED and dilated

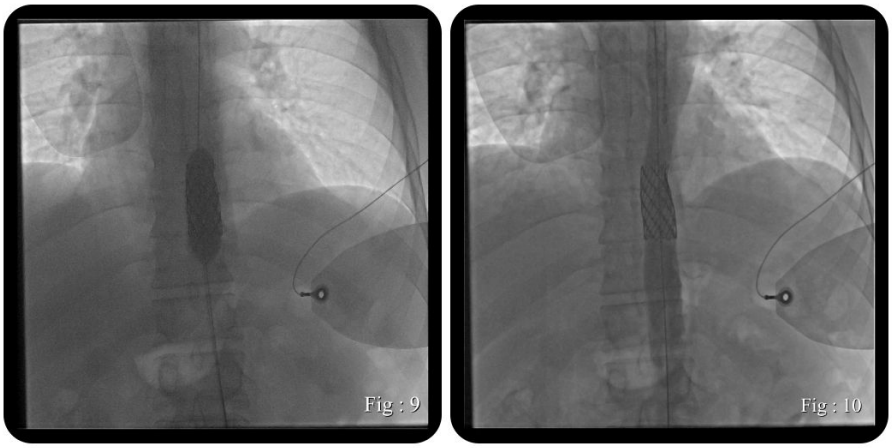

Fig: 9, 10 - Post angioplasty and stenting

There was no dissection (Fig: 10) and good bilateral Femoral, Popliteal, Anterior Tibialis and Posterior Tibialis pulses were felt. Post procedure he was started on dual antiplatelet therapy and antihypertensive and was discharged on the following day in a stable condition.

Discussion

Median arcuate ligament syndrome (MALS) or celiac axis compression syndrome (CACS) is a rare clinical entity caused by the compression of the celiac trunk by the Median Arcuate Ligament (MAL). The diaphragmatic crura arises from the anterior surface of the L1-L4 vertebral bodies on the right and

the first two or three lumbar vertebral bodies on the left, and also from the intervertebral disc and anterior longitudinal ligament. It then passes superior and anterior to surround the aortic opening and to join the central tendon of the diaphragm. The diaphragmatic crura on either side of aortic hiatus are united by MAL that usually

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passes superior to the origin of the celiac trunk (1-3). However, in about 10-24%, MAL passes anterior to the celiac artery and causes compression thereby decreasing the blood flow and producing symptoms typically of mesenteric ischemia, such as postprandial pain, abdominal distension, obstipation, nausea etc. (6-9). Also the aortic opening is anterior to T12 vertebrae, between the crura and behind the MAL and hence can cause aortic stenosis due to anatomic variations.

Harjola first described MALS in 1963 as a combination of both clinical and radiological features in a patient with mesenteric ischemia due to extrinsic compression of the celiac artery (4). He also demonstrated that it most commonly occurs in young (20-40 years) females, wherein they present with symptoms of abdominal pain and weight loss that were mostly due to compromised blood flow due to compression of the celiac axis. In 1965, Dunbar et. all reported surgical repair (division of MAL) in 15 patients and successful decompression of the celiac trunk as the treatment. However, subsequent theories for the pathophysiology of MALS suggested that it is due to anatomic variation of MAL compressing the celiac trunk and thereby compromising the blood flow (5-7).

The diagnosis of clinically significant MALS can be made with CT angiogram, which usually demonstrates a characteristic focal narrowing of the proximal celiac axis giving it a hooked appearance, which distinguishes this condition from other causes of celiac artery stenosis. The CT findings may not only be appreciated in axial images alone, as the sagittal plane is optimal for visualizing the proximal portion of the celiac axis and in some cases CT imaging can also identify the actual MAL. CT angiogram is typically performed during inspiration and hence focal narrowing is observed in the inspiratory phase, which may be clinically significant as the transient compression is seen only during expiration in some patients which will not manifest at an inspiratory phase CT (2,10-11).

Treatment modalities of MALS still remain controversial. Surgical correction sometime is difficult as many patients have anatomic abnormality of low insertion of MAL, than actually have symptoms caused by the abnormality. Surgical ligations of the constricting ligamentous bands have been performed but results were variable. Surgical corrections were more helpful in older (50-60 years) patients with post stenosis dilatation with collateral vessel formation and usually have symptoms of postprandial abdominal pain. Initially the open surgical option was describes as the only definitive treatment but due to high rate of postoperative complications and the trend toward minimally invasive laparoscopic surgery. There are very few literatures supporting the relief of symptoms after solo minimally invasive laparoscopic division of MAL (13). Solo endovascular treatment was successfully reported in some literatures, although failures were also there with stent kinking and crushed stents. A recent literature suggests the release of the MAL by surgery (Laparotomy/Laparoscopic) followed by angioplasty and stenting of the effected vessel that they termed as “hybrid procedure” as the gold standard treatment (10-11). Percutaneous angioplasty with stenting is an alternative technique used for the treatment of MALS. The advantages of percutaneous angioplasty are that it is a minimally invasive technique, characterized by short hospitalization and have low morbidity rate. Studies have reported successful endovascular repair of MAL. One of them is the study by Silva et al. (12), in which stenting was employed in four patients with extrinsic compression of the celiac artery with immediate excellent results, but only one of this four patients had a 3-year symptom-free follow up period. However with recent advances in endovascular catheters angioplasty and improved stent quality and techniques solo endovascular model will be of more practice.

Our patient had undergone emergency exploratory laparotomy, adhesiolysis and release of median arcuate ligament, which released his symptoms as the celiac trunk initially compressed was released. However he had developed symptoms of gluteal claudication (BOYD –II) and erectile dysfunction. This was due to thoracic aorta stenosis secondary to MAL, which we repaired by endovascular techniques.

Conclusion

MALS should be one of the differential diagnoses in a patient presenting within chronic abdominal pain, weight loss and vomiting in a younger patient after excluding the common etiologies. CT Angiogram (radiological) findings

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along with clinical features confirms the diagnosis of MALS. Treatment modalities are controversial however; Close cooperation between vascular Interventionist and general surgeon is required. "Hybrid procedures" are gold standard as per the recent literatures. Role of only Endovascular intervention is still under evaluation and probably will standout as the only procedure in the recent future.

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Conflicts of interest

There are no conflicts of interest.

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