Case Report

Median arcuate ligament syndrome

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Abstract

Chronic abdominal pain has several aetiologies, and many a times, the aetiology is one of the exclusions. Median arcuate ligament syndrome (MALS) also called coeliac artery compression syndrome is a form of chronic mesenteric ischaemia, although rare is an important cause. A 20-year-old male presented with recurrent episodes of pain abdomen, with a history of multiple admissions for the same, before the surgery. His physical examination and laboratories were normal, but the computed tomography (CT) done initially gave a doubtful diagnosis of MALS. A CT angiogram was done, which showed a definite kinking of the coeliac axis. After all the other causes of abdominal pain were ruled out, the patient was taken up for surgery with a provisional diagnosis of MALS. Intraoperatively, there was fibrosis around the coeliac axis with kinking. The fibrosis was cleared, and pulsations of the artery were confirmed. Postoperatively, the patient had an uneventful recovery.

Keywords: Coeliac artery compression, Dunbar syndrome, median arcuate ligament syndrome

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INTRODUCTION

The median arcuate ligament (MAL) passes in front of the aorta at the level of the L1 vertebra, just above the origin of the coeliac artery. In 16%, a low MAL covers the coeliac artery and may compress it.[1] This condition is known as coeliac artery compression syndrome or MAL syndrome (MALS) (Figure 1). Angiographically, there is coeliac artery compression that augments with deep expiration and post-stenotic dilatation. Coeliac artery compression syndrome has been implicated in some variants of chronic mesenteric ischaemia. Most patients are young females between 20 and 40 years of age. Abdominal symptoms are non-specific, but the pain is localised in the upper abdomen, which may be precipitated by meals.[2] We report a case of MALS.

CASE REPORT

Our patient is a 20-year-old male who presented with a history of recurrent upper abdominal pain. The patient had severe pain which was sudden in onset and required him to get admission in a surgical ward four times in 4 months preceding the surgery. There was no history of weight loss. At the time of admission, the patient was consistently complaining of upper abdominal pain, but examination of his abdomen revealed no positive findings; bruit was not evident in the epigastric area. Investigations at the time of admission did not show any

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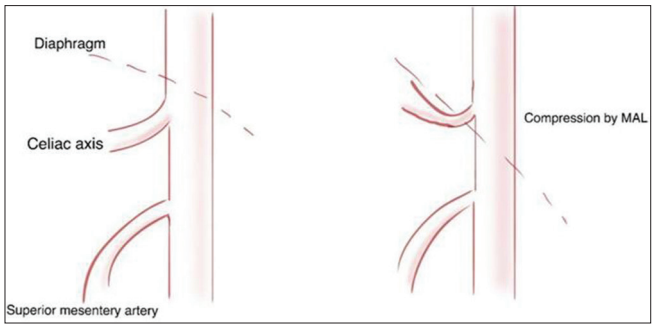

Figure 1: Normal anatomy and pathological anatomy of MAL syndrome
MAL = median arcuate ligament

elevation of amylase or lipase that could point towards pancreatitis which would be the normal pathology in such presentations. Ultrasonography of the abdomen also revealed no abnormality. Contrast-enhanced computed tomography (CT) abdomen was done initially which gave doubtful diagnosis of MALS. The patient was treated conservatively with nil per mouth and analgesics. Upper gastrointestinal (GI) endoscopy and lower GI endoscopy showed no causative lesions. At the time of the fourth admission, a CT angiography of the abdomen was done, which revealed a hook-shaped coeliac axis along with mild post-stenotic dilatation (Figure 2). A provisional diagnosis of MALS was done and decision was taken to subject the patient to operative intervention after proper counselling of the patient. Laparotomy was done and it revealed kinking of the coeliac axis along with perivascular fibrosis. The constricting fibres of the MAL were divided along with the release of the fibrosis. Pulsations of the arterial tree were confirmed finally before closing the abdomen (Figure 3). The patient had an uneventful recovery and is asymptomatic during the follow-up of 4 months post-surgery.

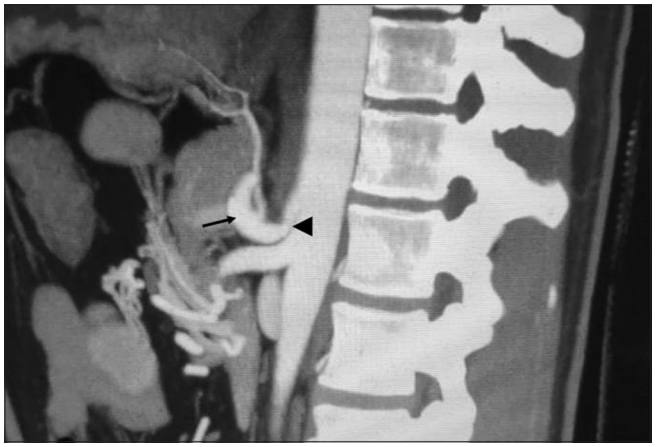

Figure 2: Sagittal computed tomography angiography showing compression of the coeliac artery (arrow head) with post-stenotic dilatation (arrow)

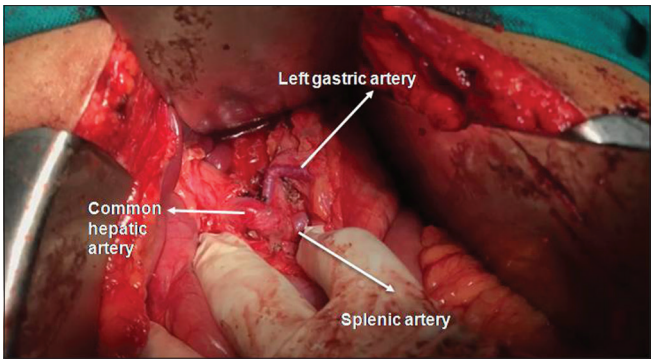

Figure 3: Intraoperative photograph showing the coeliac axis that has been cleared from the median arcuate ligament and fibrosis

DISCUSSION

The coeliac artery compression syndrome was first described by Harjola.[3] The incidence of MALS has been found to be about 10%–24%.[4] This syndrome is frequent amongst young females with a thin body habitus and associated with the classical triad of postprandial abdominal pain, epigastric bruit and the presence of extrinsic coeliac compression revealed by vascular imaging.[5] The clinical presentation in our patient was atypical in that there was no history of weight loss and epigastric bruit was not evident. Various investigations for diagnosing this disease range from the classical percutaneous angiography which is invasive to non-invasive modalities such as Doppler assessment of the coeliac axis, CT angiography and magnetic resonance angiography. Peak systolic velocity in the superior mesenteric artery >275 cm/s demonstrated a sensitivity of 92%, specificity of 96% and overall accuracy of 96% for detecting >70% stenosis. The same authors found a sensitivity and specificity of 87% and 82%, respectively, with an accuracy of 82% in predicting >70% coeliac trunk stenosis. Duplex has been successfully used for follow-up after open surgical reconstruction or endovascular treatment of the mesenteric vessels to assess the recurrence of the disease.[2]

CT angiography, specifically in the sagittal plane, aids in optimal visualisation of the proximal portion of the coeliac axis. Focal narrowing in the proximal coeliac axis with a hooked appearance is seen characteristically. CT angiography should be performed during inspiration as focal narrowing seen during the inspiratory phase may be clinically significant. Associated post-stenotic dilatation and collaterals are also visualised well.[6] The thickness of the MAL over 4 mm is considered significant.[7] Hence, end-inspiratory stenosis, post-stenotic dilation and collaterals seem to be markers of significant compression and clinical symptoms.

The treatment of MALS can be open, minimally invasive and interventional radiological procedures. A case series[8] of four patients treated with endovascular procedures successfully has been reported. However, there is no long-term follow-up supporting the efficacy of endovascular procedures over other modalities. Hence, endovascular procedures can be limited to patients who have contraindications for undergoing surgical procedures.[8]

Open surgery with lysis of the MAL is a straightforward procedure. Minimally invasive ligament lysis can also be done, and studies have shown that there is no difference in recurrence or outcomes with both the procedures although a high rate of conversion to open procedure was seen in the patients treated with laparoscopic approach.[9]

MALS or coeliac artery syndrome also called eponymously Dunbar's syndrome is a rare cause of chronic mesenteric ischaemia. This condition needs the combined efforts of the surgeon, clinical gastroenterologist and the radiologist to come to a diagnosis. Although conventional arteriography is considered standard, high-quality multi-slice CT with volume rendering and three-dimensional reconstruction have made the process observer independent and easier and less invasive. Duplex ultrasonography can be used both for diagnosis and follow-up after surgery. Surgery remains the mainstay for treatment of this elusive disease, and results remain similar with both open and minimally invasive procedures. As of now, interventional radiology procedures are indicated only in those with contraindications for surgery since there are no long-term studies regarding the same. Surgeons and gastroenterologists must equally be aware of this rare entity while treating chronic abdominal pain.

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Conflicts of interest

There are no conflicts of interest.

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