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Median Arcuate Ligament Syndrome: A Case Report

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ABSTRACT

Median Arcuate Ligament Syndrome (MALS) is a rare vascular compression disorder of the celiac artery with non-specific symptomatology that can mimic other, common abdominal diseases. We present one such case of MALS and review the relevant literature on this rare topic.

Key Words: *Artery; compression; celiac; diaphragm; fundoplication*

INTRODUCTION

The Median Arcuate Ligament (MAL) is an arch-like fibro-fascial ligament that is formed by the fibres of the right and left diaphragmatic crura along the anterior border of the aortic hiatus [1-3]. Anatomical variations leading to superior origin of the celiac trunk from the aorta and low insertion of MAL over the origin of the celiac trunk may cause compression; when compression is significant, it can present with a wide variety of symptoms that may range from chronic epigastric pain, nausea, vomiting, epigastric fullness and delayed gastric emptying [1-4]. This rare condition is known as the Median Arcuate Ligament Syndrome (MALS), Dunbar syndrome, or even, celiac artery compression syndrome [1-4].

CASE REPORT

A 27-year-old male presented in the emergency with symptoms of dull upper abdominal pain, anorexia and weight loss for a period of two years. He had been diagnosed as a case of hiatal hernia earlier and had undergone Nissen fundoplication nine months ago, but his symptoms had not settled after surgery. On physical examination the abdomen was soft and non tender and his laboratory studies were within normal limits. Ultrasound of the abdomen was non-contributory. A contrast enhanced computed tomography (CECT) scan of the abdomen was performed, which showed narrowing of the celiac trunk at its origin, measuring approximately 1 mm in transverse axis with post stenotic dilatation with thickened median arcuate ligament, suggestive of median arcuate ligament syndrome. Computed tomography angiography (CTA) revealed an attenuated celiac artery at its origin along with post stenotic dilatation (Figure 1).

The patient was taken up for surgery. Intra-operatively, the right and left diaphragmatic crura were identified and the MAL was seen covering the origin of the celiac trunk off the aorta; the celiac artery was thread-like, with faint pulsations. The MAL was divided in the midline and all soft tissue including lymphatics were carefully dissected to widely expose the aorta and celiac trunk and a clear view of the anatomy was obtained (Figure 2). Once this was done, the celiac artery showed an immediate increase in caliber and pulsations.

The post-operative course was uneventful and the patient was discharged on the second postoperative day without any dietary restriction.

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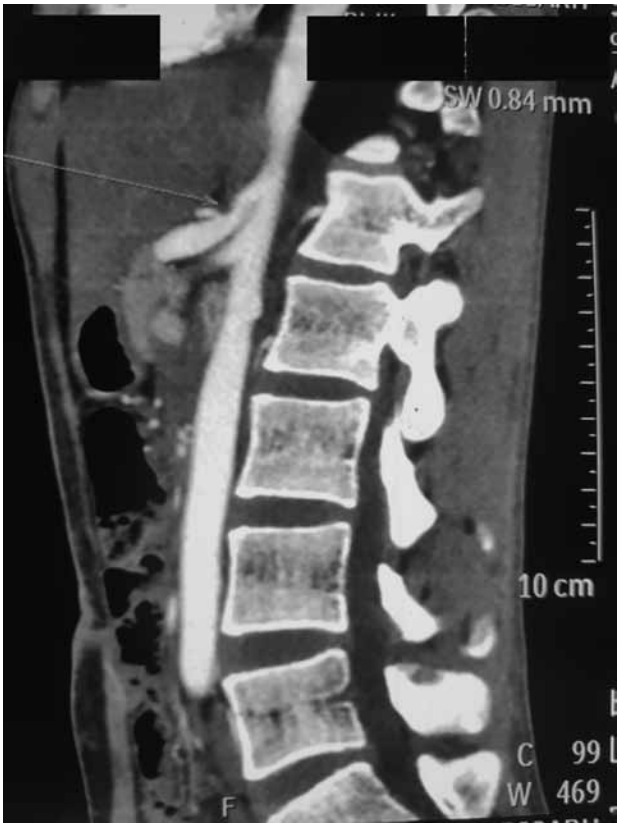

FIGURE 1

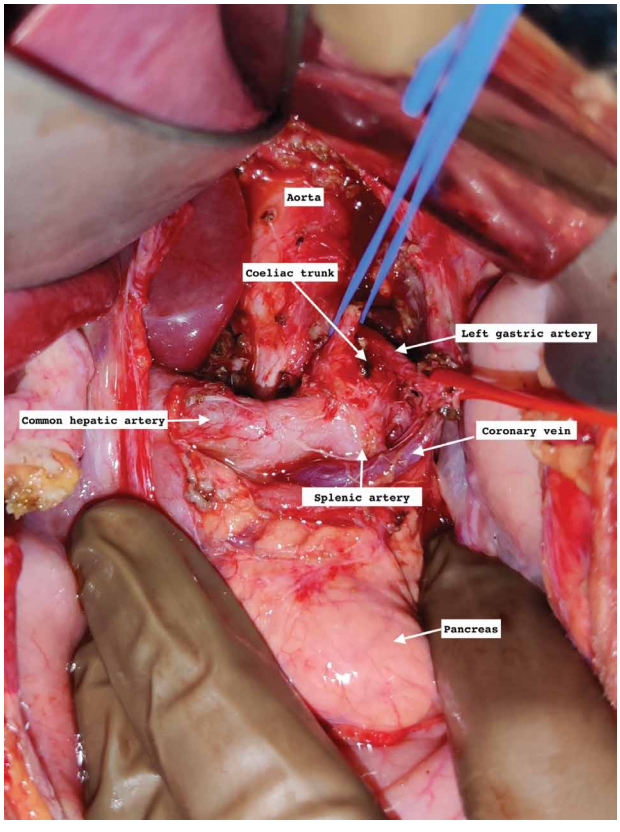

FIGURE 2

DISCUSSION

Ever since the initial descriptions by Harjola (1963) and later by Dunbar (1965), MALS has remained a rare diagnosis, usually by exclusion of other causes [1,3]. It is seen more commonly in thin females in the 3rd to 5th decades of life and presents with chronic epigastric pain (post-prandial or triggered by exercise), nausea, vomiting, weight loss and bloating [1-5]. The exact pathophysiology of MALS is still unclear - most current theories revolve around gastrointestinal ischaemia arising from mechanical compression of the celiac artery by MAL; however, compression of the celiac nerve plexus by MAL may also contribute [1,4]. Usually, the MAL transverses the aorta cranial to the origin of the celiac artery, but low insertion of MAL has been reported in up to 24% of the population [1]. In addition, there may be anomalous higher origin of the celiac trunk from the aorta, and any of these, alone or in combination, may cause compression of the proximal celiac artery, especially during expiration [1,3,6]. However, simply, compression does not always translate into MALS; compression of the celiac artery on angiography is seen in almost 50% of individuals, but not all are symptomatic [6,7].

Clinically, MALS can range from being completely asymptomatic to having very general presenting symptoms. The typical symptoms reported in MALS are post-prandial abdominal pain, weight loss, nausea and vomiting; other presentations include anorexia, nausea, vomiting, fatigue and diarrhea, similar to several other abdominal disorders like gastritis, peptic ulcer disease, hepatitis, cholecystitis, chronic pancreatitis, colorectal malignancy, appendicitis or chronic mesenteric ischaemia [1-7]. On occasion, abdominal bruit (increasing during expiration) may be heard. Given the rarity of the disease and its non-specific symptomatology, the diagnosis is difficult, and depends upon specific findings on abdominal Doppler, CT or MR angiography [1,4-7]. Abdominal doppler is a good initial investigation as it is easy to perform, avoids radiation and contrast, and can reveal post stenotic dilatation and increased flow velocities in the celiac artery that normalise on inspiration [1,2,5]. A combination of a deflection angle of greater than 50% and a high expiratory peak systolic flow velocity (more than 350 cms/sec) is highly sensitive and specific for the diagnosis of MALS [6]. However, this investigation is highly operator dependent, as well as limited by overlying bowel gas. Although conventional angiography is considered the 'gold standard' for diagnosing MALS [1], CTA has the advantage of high resolution three-dimensional reconstruction of the celiac axis as well as better visualisation of any other abdominal pathology [1,5-7]. Focal narrowing of the proximal celiac

artery and the presence of 'J-hook' pattern that normalises on expiration is considered diagnostic; in addition, CTA has better pick up of post-stenotic dilatation, distal aneurysms and collaterals [5-7]. MR angiography can be used instead of CTA in patients with contrast hypersensitivity or renal failure [1].

Surgery remains the mainstay of treatment [2]; decompression of the celiac artery by division of MAL and celiac ganglionectomy is recommended in symptomatic patients having documented compression on imaging in inspiration [1,2,6]. This can be done by open, laparoscopic or robotic means, although there is a recent trend towards the laparoscopic approach [4,7]. It has been reported that up to 85% of patients have symptom relief in the immediate post-operative period; typical pain (post-prandial or exercise induced), age between 40 to 60 years and weight loss greater than 10 kg are predictive of success after surgery [4,6]. Patients with persistence or recurrence of symptoms in the long term need evaluation for persistent stenosis/restenosis of the celiac artery either due to pathological changes (intimal fibrosis, smooth muscle proliferation, abnormal elastic fibres, disruption of medial and adventitial layers) that is often seen in long standing MALS, or abnormal anatomical configuration [4]. The treatment of such patients is by percutaneous transluminal angioplasty (with or without stenting) or arterial reconstruction [1,2,4].

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