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Case Report

Median arcuate ligament syndrome and a laparoscopic coeliac trunk first approach: a literature review and a proposed algorithm

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ABSTRACT

Median arcuate ligament syndrome (MALS) is a rare condition where chronic recurrent abdominal pain is related to the compressive effects of the median arcuate ligament on the coeliac artery. The mechanism behind this phenomenon is incompletely understood but thought to be both ischemic and neuropathic in nature. As a result, the management of this condition remains controversial. Furthermore, while there have been a variety of options both radiological and interventional described for the investigation of MALS, there has yet to be a consensus in how patients exhibiting symptoms should be assessed and worked up. In this article, we described a laparoscopic coeliac trunk first approach in a young female patient and following review of the literature, propose an algorithm that can be used in the assessment and management of suspected MALS.

Keywords: Median, Arcuate, Ligament, Coeliac, Dunbar, Algorithm

INTRODUCTION

Median arcuate ligament syndrome (MALS) is a rare condition first described by Harjola in 1963 and subsequently by Dunbar and Marable in 1965. It is classified by a myriad of non-specific symptoms however the pathophysiology of this phenomenon remains poorly understood. There are a variety of options available for diagnosis however due to the rarity of the condition, there is no consensus on how these patients should be worked up, and similarly, the management of this conditions is controversial. Using a unique case of laparoscopic coeliac trunk first approach in a young female patient, we performed a literature review in order to propose an algorithm that can be used in the assessment and management of suspected MALS.

CASE REPORT

A 21-years-old female presented with a year history of grumbling upper abdominal pain associated with nausea. There were no obvious exacerbating or relieving factors and the pain was unrelated to food. The patient was otherwise fit and well, and there was no significant family history. She underwent an abdominal ultrasound which demonstrated an incidental finding of high flow velocity seen within the coeliac trunk with peak systolic velocity of 400 cm/sec, suggesting a 75% stenosis. The remainder of the ultrasound was unremarkable. A subsequent computed tomography mesenteric angiogram (CTMA) demonstrated focal narrowing of the proximal coeliac artery 5 mm distal to its origin with approximately 56% luminal narrowing and post obstructive fusiform aneurysmal dilatation, thought to be secondary to median arcuate ligament extrinsic compression (Figure 1).

The patient was taken for elective operative management. A 10 mm Hasson port was placed infraumbilically, followed by two left upper quadrant ports (10 mm and 5 mm) and a 5 mm right upper quadrant port. The pars flaccida was divided and the left gastric artery retracted caudally with a vessel loop. The dissection was

commenced on the crus below the hiatus until the aorta was seen on pre-adventitial tissue; as such the hiatus was preserved. The left gastric artery was then traced down to the coeliac/solar plexus divided along with tough tissue on the coeliac trunk exit (Figure 2-4). Post division of the ligament shows a clear path of the coeliac artery origin. A lift test was performed to confirm release and a final check demonstrated adequate exposure of the coeliac trunk. The ports were removed and closed in layers thereover. A post-operative CTMA demonstrated improved calibre of the coeliac trunk with minimal constriction and less narrowing at the origin compared to the pre-operative CTMA (Figure 5). The patient was discharged 3 days later and was well upon her 30 days follow-up with improvement of her symptoms.

Figure 1: Pre-operative CT mesenteric angiogram demonstrating focal narrowing of the proximal coeliac artery and post obstructive fusiform aneurysmal dilatation secondary to median arcuate ligament extrinsic compression.

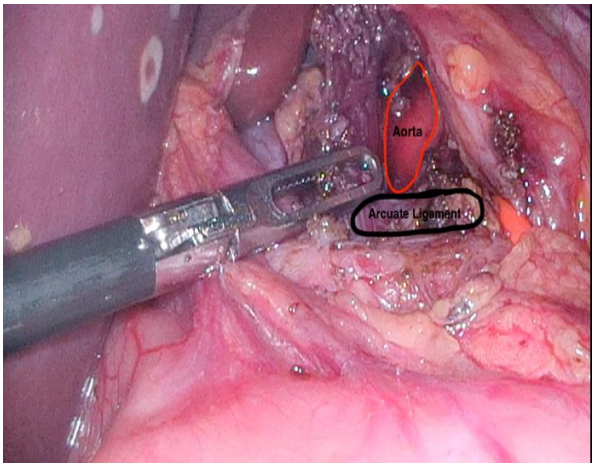

Figure 2: Exposure shows the aorta with a tight band of median arcuate ligament; the left gastric artery has been retracted with a vessel loop.

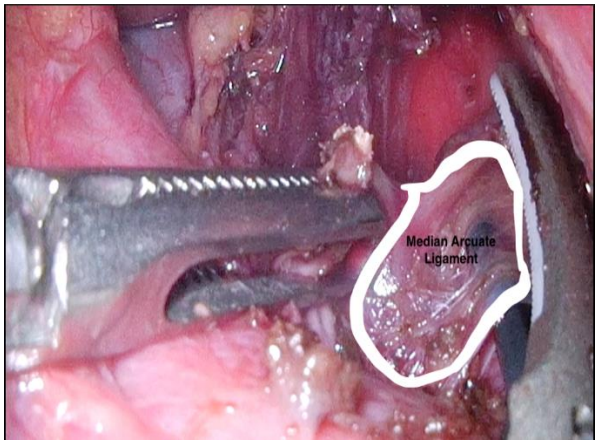

Figure 3: The fibres are divided with a harmonic tissue sealer to ensure the hot plate is away from the aorta.

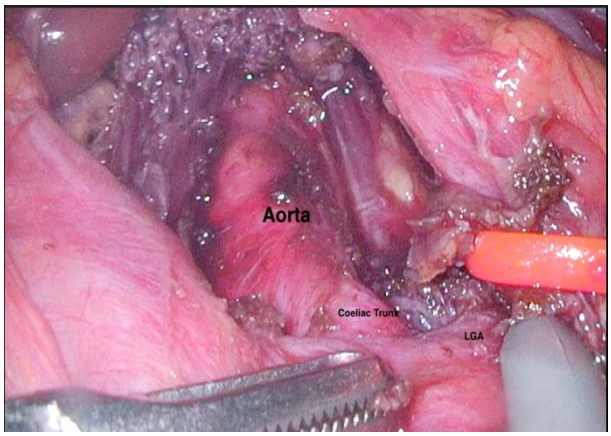

Figure 4: Following division, the origin of the coeliac trunk and the left gastric artery (LGA) is clearly demonstrated, without any residual tissue.

Figure 5: Post-operative CT mesenteric angiogram demonstrating improvement of focal narrowing and decreased angulation of the proximal coeliac artery following laparoscopic release.

DISCUSSION

MALS, also known as coeliac artery compression syndrome, coeliac axis syndrome, and Dunbar syndrome, describes a condition where chronic recurrent abdominal pain is related to the compressive effects of the median arcuate ligament on the coeliac artery. The typical symptoms often exhibited by patients include post-prandial epigastric pain, weight loss and nausea and/or vomiting. The mechanism behind this phenomenon is incompletely understood but thought to be both ischaemic and neuropathic in nature.

The median arcuate ligament is a fibrous arch that connects the crura of the diaphragm above the origination of the coeliac axis off the abdominal aorta, which usually occurs between T11 and L1 however a wide variation has been described.[1] Similarly, there are variations of the location of the median arcuate ligament.[2] It has been surmised that due to abnormal anatomical variations of either or both the coeliac axis and the median arcuate ligament, the symptoms may be ischaemic in nature. However, due to the rich collateral circulation from the superior and inferior mesenteric arteries, perfusion of the abdominal viscera should not otherwise be compromised, which is supported by patients with radiological evidence of coeliac axis compression but remain asymptomatic.[3]

The pain associated with MALS is also thought to be mediated by the coeliac plexus or ganglion, which originates from the splanchnic, phrenic and vagus nerves, as well as the parasympathetic and sympathetic nervous system. This plexus lies adjacent to the median arcuate ligament with subsequent compression theorised to also contribute to symptoms, however this is somewhat controversial.[4]

MALS is more prevalent in women compared with men (4:1) and more common in patients of 40-60 years of age. Epigastric pain is the most common presenting symptom (often post-prandial or exacerbated by exercise), followed by unintentional weight loss, nausea and vomiting. Due to its low incidence and non-specific symptoms, the diagnosis of MALS is often delayed and one of exclusion. Patients will often have had a multitude of investigations, including a CT abdomen, which may demonstrate coeliac artery stenosis with or without post-stenotic dilation or aneurysm. A dedicated CT or MR angiography with 3D reconstruction study can also be useful to provide multiple angles of the compression and anatomical relations to surrounding structures.[2,5] The diagnosis of MALS can be confirmed with ultrasound or catheter-based angiogram and is aided by concurrent respiratory manoeuvres.

Duplex ultrasound has the benefits of being non-invasive and more pronounced dynamic respiratory manoeuvres when compared to other modalities, however findings are operator dependent. Diagnostic findings indicative of coeliac artery stenosis included increased systolic flow velocities, visible external compression of the coeliac artery and post stenotic dilation.[5] The Gruber group found that duplex ultrasonography had a sensitivity of 83% and specificity 100% diagnosing MALS using a peak systolic velocity of >350 cm/sec, 210% change in pulse volume amplitude with inspiration and expiration, and a coeliac artery deflection angle of 50^{o}.[6]

Catheter-based angiogram can also be considered in the diagnosis of MALS. The advantages of this modality are the ability to evaluate flow dynamics and collateral circulation, as well as directly visualising coeliac artery compression with dynamic respiratory manoeuvres. The cranial movement of the diaphragm during expiration can be visualised to worsen the compression of the coeliac artery, and conversely, the caudal movement during inspiration relieves it. A patent coeliac artery without any evidence of compression on expiration excludes a diagnosis of MALS. Other positive findings include post stenotic dilation, pressure gradient measurement over the coeliac artery and retrograde filling via collateral supply. The main disadvantages include the invasive nature of catheter angiography as well as the indirect visualisation of the source of compression (which can be better appreciated on CT or MRI).[7]

Median arcuate ligament release with concurrent ganglionectomy is the mainstay of surgical treatment for MALS which can be offered via an open, laparoscopic or robotic approach. The median arcuate ligament and coeliac plexus fibres are resected to skeletonise the coeliac artery thus relieving the compressive symptoms. The usual surgical management often involves dissection of the hiatus, however in our case, we were able to preserve it. There is conflicting evidence regarding the superiority of either laparoscopic or open release, however given laparoscopic methods are still relatively new, the risk of converting to open remains significant given the high morbidity associated with acute bleeding of the supra-coeliac aorta. Some groups also consider coeliac artery revascularisation or reconstruction during the index operation, however historically this was more common during open procedures.[7] Whilst there is a paucity of evidence on which method is more effective long term, there have been multiple groups who have reported good outcomes from laparoscopic release alone.[8,9] Robotic approaches to median arcuate ligament release offer enhanced 3D visualisation, improved stability and ergonomics, and increased ability to operate in more confined spaces, and the results have been promising with comparable short and intermediate term outcomes to laparoscopic approaches.[10,11]

The short-term results of laparoscopic versus open, as well as ligament release alone versus combined with coeliac artery manipulation, are comparable, but there is again insufficient evidence to determine assess recurrence or restenosis.[7] This is further compounded by the fact that the pathophysiology behind MALS remains poorly understood, and so the potential placebo effect remains uncertain.

Post-op patients following ligament release are subsequently followed up with a routine visit at 1 month. The Columbo group suggests that those who are asymptomatic do not require routine long-term surveillance or progress imaging unless symptoms recur, after which duplex ultrasonography and subsequent catheter-based angiography is re-pursued.[12] Persistent stenosis may warrant revascularisation, either percutaneous via coeliac artery angioplasty/stenting or surgically via bypass grafting.[7,12]

We proposed the following algorithm for the management of suspected MALS (Figure 6).

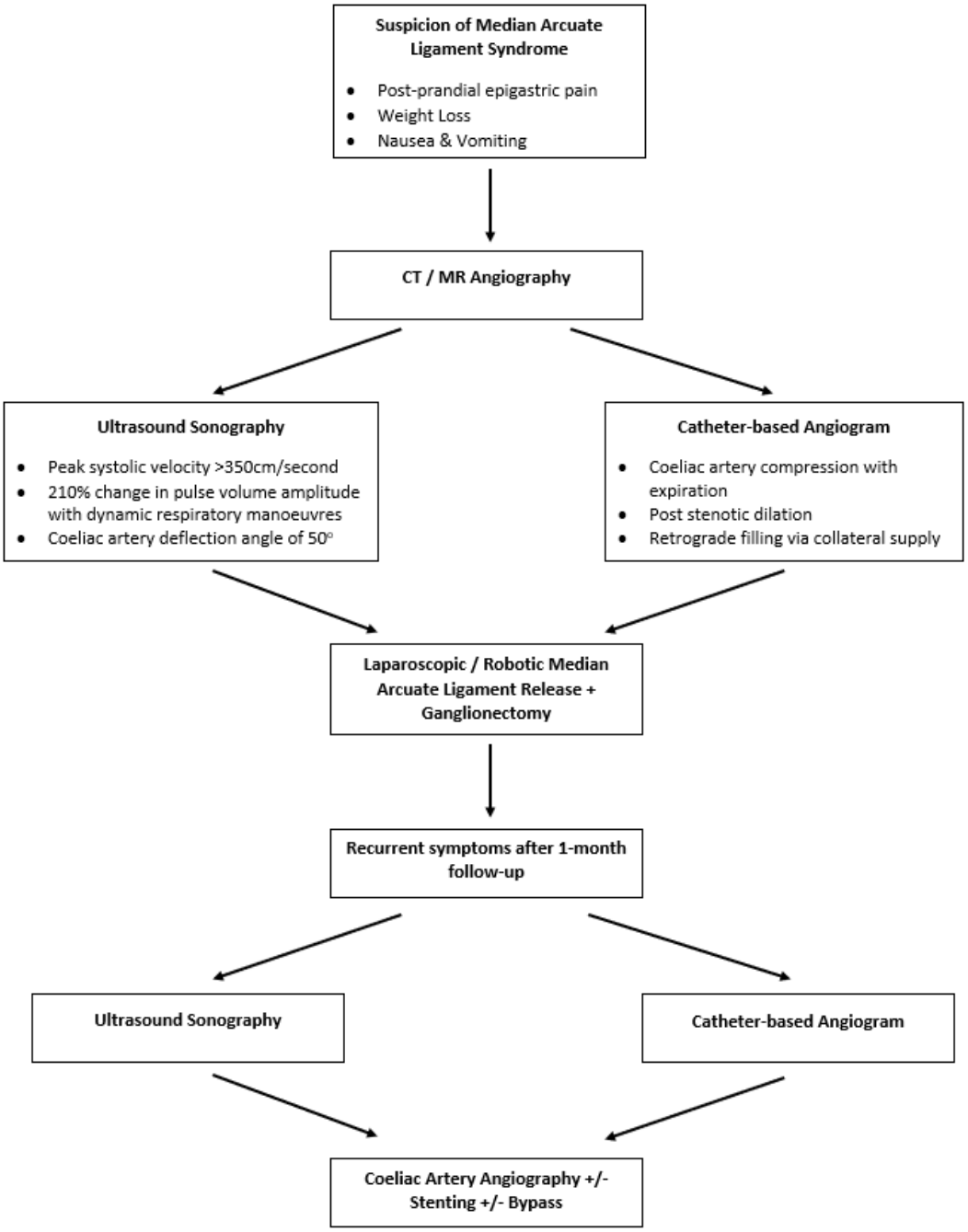

Figure 6: Proposed algorithm for the assessment and management of median arcuate ligament syndrome.

CONCLUSION

MALS is a rare condition that to date remains poorly understood due to the uncertainty of the pathophysiology behind this phenomenon. There are a variety of options available for diagnosis however there is no consensus in the literature on how these patients should be worked up, and similarly, the management of this conditions remains

controversial. Our manuscript proposed a simple algorithm that can be used in the clinical assessment and management of suspected median arcuate ligament syndrome.

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