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Median Arcuate Ligament Syndrome



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Median arcuate ligament syndrome is an uncommon disorder first described in the 1960s. It is characterized by epigastric abdominal pain accentuated by meals and weight loss associated with nausea, vomiting and gastroparesis. Abnormal gastric electrical rhythm has also been reported. Abdominal bruit is a striking feature that is present in some cases. It is a diagnosis of exclusion that should be considered when there is a subjective presentation of severe epigastric and right upper quadrant abdominal pain which is out of proportion to objective findings. Whether using Doppler study, CT angiography, MRA or angiography, the main and most important goal is assessing both inspiratory and expiratory phases of the celiac artery to demonstrate reduction in the compression during inspiration. The treatment is surgical release of the median arcuate ligament to achieve decompression of the celiac artery and the celiac plexus. An evolving role for endoscopic ultrasound both in diagnosis and management is also discussed.

INTRODUCTION

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome (CACS), Dunbar syndrome and celiac axis syndrome is a cause of chronic epigastric and right upper quadrant abdominal pain that is explained by the median arcuate ligament, a fibrous tissue connecting the two crura of the diaphragm, compressing the celiac artery and its companion structure, the celiac ganglion.¹ Theories explaining the pathophysiology of the disease suggest either neurogenic or vascular origin.² The

clinical presentation is with abdominal pain, weight loss and gastroparesis. It is a diagnosis of exclusion. Most patients will undergo extensive laboratory and imaging workup looking for the diagnosis and some might undergo unnecessary surgical procedures like cholecystectomies and appendectomies.³ The treatment traditionally has been surgical with the goal of releasing the median arcuate ligament (MAL), resulting in restoration of blood flow to the celiac artery and relieving the entrapment of the celiac ganglion fibers.^{4,5}

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Etiology and Pathophysiology

MALS or CACS, also named Dunbar syndrome is a chronic epigastric and right upper quadrant abdominal pain exacerbated by eating and explained by the compression of the celiac artery by the median arcuate ligament at the level of the diaphragm.⁴

Understanding the anatomy of the MAL and the extensive collateral blood supply formed by the celiac artery, superior mesenteric artery (SMA) and the inferior mesenteric artery (IMA) is a key to understanding the pathophysiology of MALS.

The MAL is a fibrous structure that connects the diaphragmatic crura and crosses the aorta just proximal to the origin of the celiac artery. The celiac plexus originates from somatic branches of the phrenic and vagus nerves, parasympathetic preganglionic and sympathetic postganglionic fibers, and preganglionic splanchnic nerves. Its anatomic relationship to the celiac artery origin means that the MAL would compress both the celiac artery as well as the celiac ganglion.⁶ (Figure 1).

It was first suggested that the compromise of blood supply by compression of the celiac artery would lead to post prandial ischemia and hence worse abdominal pain. However the SMA and IMA are widely patent in this entity and the stomach has extensive collateral blood flow from the gastric and epiploic arcades leading to the conclusion that ischemia is not the explanation for the abdominal pain and other associated symptoms. It was suggested later that pressure on the celiac plexus might be the main etiological factor in MALS² and thus it could also explain the accompanying findings often present in addition to the abdominal pain, namely nausea, vomiting, gastroparesis, and gastric electrical dysrhythmias.

Many theories have been proposed to explain this altered anatomy including congenital origin as it has been found in children and twins,^{7,8} as well as traumatic origin.⁹ The presentation in adults could be explained by a combination of a congenitally early take off origin of the celiac artery and/or an abnormally thickened MAL with or without accompanying arteriosclerosis in the vessel that may predispose to the “narrowing” finding on expiration.

Clinical Manifestation

Most patients report chronic epigastric and right upper quadrant abdominal pain which is accentuated after meals. Weight loss is explained by reluctance to eat for the fear of provoking pain. There are also symptoms that are manifestations of delayed gastric emptying, namely nausea and vomiting. In addition the normal gastric electrical rhythm has been reported as irregular (e.g. tachygastria) contributing to nausea and vomiting as well as impaired gastric motility. This is

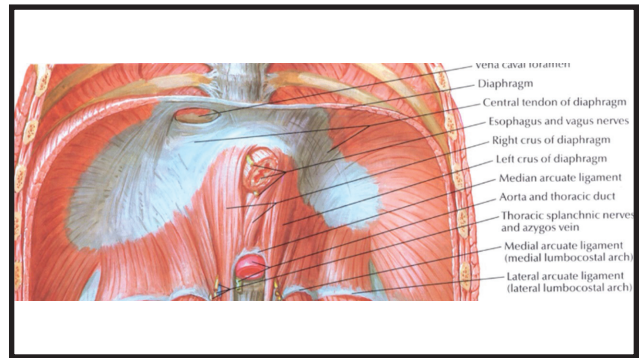


Figure 1. Illustration of the anatomical relation of the median arcuate ligament (MAL) of the diaphragm to the aorta. The proximity of the origin of the celiac artery to the MAL is the reason why both the celiac artery and the celiac ganglion can be compressed by the MAL.

all consistent with the neurogenic pathophysiology of the syndrome by inhibiting the gastric neuromuscular function.² As previously stated, these patients usually undergo extensive workups including computed tomography (CT) scans, magnetic resonance imaging (MRI), angiographies, and some even undergo surgical procedures including cholecystectomies, appendectomies, laparoscopic or even gynecological procedures to make the diagnosis or at least try to relieve the symptoms^[10].

On physical examination one striking feature is the finding of an abdominal bruit present in up to thirty five percent of patients based on a review of cases from 1963 to 2012.⁶ In addition there is epigastric and/or right upper quadrant tenderness. Other non-specific findings might be weight loss and cachexia.

Making the Diagnosis

As previously explained, MALS is a diagnosis of exclusion to be considered when the subjective presentation of upper abdominal pain is dominant in the absence of any objective findings. Other causes of upper abdominal pain including but not limited to gastroesophageal reflux disease, pancreatitis, cholecystitis, gastric outlet obstruction and chronic intestinal ischemia from vasculitis or arteriosclerosis have to be considered. Extensive testing and therapy directed at these suspected causes have been unsuccessful.

This syndrome is seen more in females ranging in age between 40-60 years. The fact that the abdominal pain is exacerbated by eating and the possible presence of a bruit along with significant weight loss raises



Figure 2. Illustration of the expiratory phase of the angiogram, lateral view, demonstrating the compression of the celiac artery at its origin by the median arcuate ligament consistent with celiac artery compression syndrome.

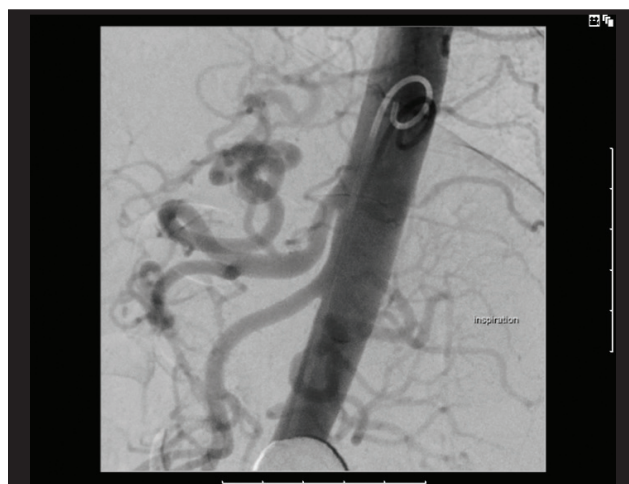


Figure 3. Illustration of the inspiratory phase of the angiogram, lateral view, demonstrating reduced compression on the celiac artery.

the index of suspicion for the diagnosis.⁴ Imaging is required to confirm the celiac artery compression by the MAL and other imaging is necessary to exclude any other causes of the patients' symptoms.

The diagnosis can be made through different imaging modalities including duplex ultrasound, CT angiography, magnetic resonance angiography, and arteriography. The findings that are specific for the diagnosis include compression of the celiac artery with changes in the degree of obstruction related to inspiration and expiration, post compression dilation, and elevated velocities of blood flow.^{6,11} This is caused by two physiological phenomena; the first is that the aorta is displaced both anteriorly and inferiorly during inspiration. Second, as the diaphragm moves down during inspiration, the crura will relax and the compression on the celiac artery will be relieved.

Duplex Ultrasound

Performed during deep expiration, duplex ultrasound shows increase in the blood flow velocity across the compressed area of the celiac artery and supports the presence of constriction caused by the MAL.¹² An example of the change in velocities is illustrated in two cases reported by Alper Ozel et al.¹³ In the first patient, the peak systolic blood flow velocity during deep inspiration and expiration were 135 cm/second and 308 cm/second, respectively. In the second patient, the peak systolic blood flow velocity during inspiration and expiration were 276 and 430 cm/second,

respectively. The advantages of duplex ultrasound over CT angiography or arteriography are that it is noninvasive, less expensive and does not expose the patient to radiation or contrast.

CT Angiography (CTA) and Magnetic Resonance Angiography (MRA)

CTA has several advantages in making the diagnosis over arteriography.¹ It shows the diaphragm and its relation to the celiac and SMA. One can rotate the three dimensional images in real time into different viewing angles to find the optimal one. All these options can be achieved with a single dose of contrast and a single dose of radiation compared to arteriography where each view requires additional contrast and radiation exposure, besides the fact that it is less invasive and less expensive than arteriography.

MRA has further advantage of no radiation exposure.¹¹ Both CTA and MRA can be used to document post-operative relief of compression.¹⁴

Arteriography

The gold standard for diagnosis of MALS is the lateral view of aortic angiography.⁵ It shows asymmetric focal narrowing in the origin of the celiac artery of more than 50% during expiration with or without distal dilation. Again, the narrowing changes with the respiratory cycle, being improved during inspiration (Figures 2 and 3). Collaterals may be seen in the anteroposterior

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view as well as retrograde filling of the celiac via the dilated gastroduodenal artery.

Venkat Kalapatapu et al proposed a new technique to confirm the diagnosis of MALS.¹⁵ The idea was to steal the blood supply from collaterals by injecting a vasodilator after selective cannulation of the SMA; a positive test leads to reproduction of symptoms. Four out of eight patients who had celiac artery compression and patent SMA had positive test and underwent successful surgical treatment. Three out of four were asymptomatic at follow up and one patient had mild abdominal discomfort.

Treatment

When the diagnosis is confirmed with imaging in patients with persistent and unexplained upper abdominal pain by evidence of celiac artery compression, there is no medical therapy. Surgery is the traditional option for those patients. There are different modalities of surgery. The traditional approach is decompression of the celiac artery and celiac plexus by the division of the fibers of the MAL.^{2,16} Another approach does achieve decompression with angioplasty and vascular reconstruction but does not address the role of the celiac plexus.^{17,18} Laparoscopy and robotic-assisted laparoscopic approach have been utilized successfully.^{6,14,19,20} Intraoperative pre- and post-decompression flow velocity studies are performed routinely to increase the success of the procedure. Post-operative angiography usually still shows some mild compression of the celiac artery (<30%) during expiration.

Evolving and New Concepts

Endoscopic ultrasound (EUS) can be a good predictor of response to surgical decompression and a good way to strengthen the diagnosis of MALS. The technique is EUS guided injection of the celiac ganglion with xylocaine and monitoring over the next weeks whether there is any symptom improvement (Figure 4). If so, then there would be more confidence in the diagnosis and hence the decision for surgery.

Another role for EUS is EUS-guided injection of xylocaine and alcohol into the celiac ganglion to further enhance the results of post-surgical decompression when there is some remaining abdominal pain component. EUS guided therapy could also replace the need for repeated surgery should there be recurrence of the symptoms during long term follow up.¹⁸



Figure 4. EUS-guided celiac plexus neurolysis. The picture to the left shows the anatomic relations of the diaphragm, aorta, the celiac trunk and the superior mesenteric artery as defined by EUS. The picture to the right illustrates the endoscopic injection of the celiac ganglion by EUS.

Outcome and Prognosis

Following surgical decompression, the outcomes have varied widely in the literature mainly because the patients differed in presentations and comorbidities. While symptoms have resolved in most patients after intervention, some others did not have appreciable clinical benefit.^{3,21}

Post-operative pain relief was achieved in most patients treated with open or laparoscopic median arcuate ligament decompression.⁶ Reilly et al. reported in their study the outcome of 51 patients followed after surgical treatment for a mean of 9 years.²¹ They concluded that the factors that were associated with the best success rate were post prandial pain (81 percent of patients were cured), age between forty and sixty (77 percent of patients were cured) and pre-surgical weight loss of 20 pounds or more (67 percent of patients were cured). On the other hand patients with an atypical pain pattern, age more than 60, psychiatric disorders or alcohol abuse and weight loss of less than 20 pounds showed less success rate after intervention. In addition if patients had required chronic narcotic use to address pain prior to surgery their response was also less optimal. ■

SUMMARY AND HIGHLIGHTS

- MALS is an uncommon cause of chronic epigastric and right upper quadrant abdominal pain that is out of proportion to any objective data gathered by diagnostic testing.

- Pain is not controlled medically and patients may be requiring narcotics.
- Additional accompanying findings are nausea, vomiting, unexplained gastroparesis and weight loss.
- Whether using Doppler ultrasound, CT angiography, MRA, or angiography, obtaining both inspiratory and expiratory phases is crucial to make the diagnosis since relief of compression on the celiac artery during inspiration must be demonstrated.
- Surgical decompression relieves abdominal pain, nausea, vomiting, gastric dysrhythmia, and gastroparesis in most patients but a subset have some continuing pain component.
- There are new roles for EUS in the management of MALS.

A) Pre-operatively, it could be a good predictor for surgical response by monitoring patients' symptom relief after EUS-guided celiac ganglion injection.

B) It has a therapeutic role of celiac plexus neurolysis to supplement the surgical outcome. Furthermore, it could be an alternative to future repeat surgery.

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