Laparoscopic treatment of median arcuate ligament syndrome

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Summary	Median arcuate ligament syndrome (MALS) refers to a clinical syndrome caused by compression of the median arcuate ligament due to the fibers of this ligament that connect the diaphragmatic crura on the two sides of the aortic foramina, forming the anterior edge of the aortic foramina. If MALS is suspected, invasive digital subtraction angiography and computed tomography angiography or magnetic resonance angiography (MRA) can be used to verify the location of the celiac trunk. A disrupted or increased blood flow in the proximal end of the celiac trunk can be detected with doppler ultrasound, indicating stenosis. Treatment needs to alleviate celiac trunk compression. A common procedure involves separation of the ligament fibers and other surrounding tissues around the beginning of the celiac trunk. This can be achieved by either laparotomy or laparoscopic surgery. Patient prognosis is good, with a cure rate of about 80%.

Keywords: Median arcuate ligament syndrome, laparoscopic treatment

1. Introduction

The median arcuate ligament is the fibrous ligament that connects the diaphragmatic crura on the two sides of the aortic foramina, forming the anterior edge of the aortic foramina. The celiac trunk mostly starts below the median arcuate ligament and then divides into the hepatic artery, splenic artery, and left gastric artery. In 10% to 24% of the population, the celiac trunk is adjacent to the median arcuate ligament and may be compressed by the ligament, resulting in reduced blood supply for the corresponding organ and symptoms (1). Katz-Summercorn *et al*. (2) found that in 92.6% of 99 autopsies, the celiac trunk is adjacent to the median arcuate ligament (the distance could not be measured), and in 33.7% of those cases the celiac trunk was compressed or distorted.

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome or Dunbar syndrome, mainly refers to a clinical syndrome caused by compression of the median arcuate ligament. MALS was first postulated in the 1960s (3,4) and was commonly seen in slender women ages 20 to 40 years (1). The typical triad of MALS syndromes is postprandial abdominal pain, weight loss, and an abdominal vascular murmur, but the appearance of these 3 symptoms at the same time is not common; other manifestations include anorexia, nausea, vomiting, diarrhea, and fatigue. Symptoms may be aggravated after exercise or when the body is in a certain position. The clinical manifestations after surgery are abdominal pain (80%), weight loss (48%), an abdominal vascular murmur (35%), nausea (9.7%), and diarrhea (7.5%) (5). The complications of MALS include gastroparesis and pancreatic duodenal aneurysm, etc., the mechanism of which includes long-term chronic ischemia, development of collaterals, and a long-term extensive decrease in blood flow (6-8). The diagnosis of MALS requires clinical manifestations and imaging studies and ruling out other causes that may lead to similar symptoms. Treatment is primarily performed by releasing the median arcuate ligament, removing the abdominal nerve plexus, and/ or selective vascular reconstruction.

2. Etiology and pathogenesis of MALS

The pathogenesis of MALS has not been definitively determined and may include the mechanisms described below.

The starting point of the celiac trunk is too high or the median arcuate ligament is too long, compressing the celiac trunk, or abdominal ganglia are fused (including the superior mesenteric ganglia), compressing the celiac trunk (9). The compressed celiac trunk may cause limited blood flow and organ ischemia. Evidence to support the hypothesis of celiac trunk compression is that stenosis of the proximal end of the celiac trunk is found in the form of a hook in some patients during imaging studies; other findings are post-stenotic hemangiectasis and development of collaterals. The symptoms may resolve after release of the ligament during surgery. A study was conducted to determine whether the gastric mucosa in patients with MALS was ischemic by ascertaining gastric tension (10). When surgery was performed to release the ligament or intra-abdominal vascular reconstruction was also performed, ischemia was significantly alleviated.

In patients with conventional chronic mesenteric ischemia, however, the obstruction or severe stenosis of at least two of the three mesenteric vessels may result in symptoms of abdominal pain due to the presence of extensive collaterals carrying intestinal blood; however, the superior mesenteric artery and inferior mesenteric artery may not be affected in patients with MALS since they can theoretically provide a sufficient blood supply to the intestines. Evidence contradicting the theory of ischemia also includes angiography of asymptomatic patients which indicated compression of the median arcuate ligament (11,12). When surgery is performed to relieve compression, not all symptoms are alleviated (5). Therefore, abdominal pain may be associated with compression and intermittent ischemia of the visceral plexus (13). The abdominal nerve plexus is adjacent to the median arcuate ligament, and its source consists of preganglionic visceral nerves, phrenic nerves as well as somatic nerve of the vagus nerve, the preganglionic nerves of parasympathetic nerves, and postganglionic nerves of the vagus nerve. Abdominal pain can be caused by vasoconstriction or direct stimulation of the sympathetic nerves when the celiac plexus is involved.

Some studies contend that delayed gastric emptying is involved in the occurrence of MALS (13,14). A study of gastric myoelectric activity in a patient may facilitate readjustment of the gastric electrical rhythm and relief of symptoms, improving emptying of the contrast agent after compression of the celiac trunk is relieved.

In addition, one report described MALS in a couple of identical twin sisters (15) while another described compression of the celiac trunk in a father and a daughter as well as three brothers (16), suggesting that genetic factors may be involved in the development of this syndrome.

## 3. Diagnosis and treatment of MALS

The diagnosis of MALS requires the combination of clinical manifestations and imaging studies and ruling out other etiologies such as gallbladder disease, peptic ulcer, appendicitis, and inflammatory bowel disease (17), but there are no uniform diagnostic criteria.

A commonly used form of screening for MALS is Doppler ultrasound (18). If blood flow is disrupted or increased in the proximal end of the celiac trunk, this indicates stenosis. Gruber et al. (19) used a peak systolic expiratory velocity greater than 350 cm/ s and bending of the celiac trunk greater than 50º as criteria to identify MALS with a sensitivity of 83% and a specificity of 100%y. Selecting a certain systolic expiratory velocity could improve the sensitivity of diagnosis because celiac trunk compression is more severe due to lateral migration of the aortic and celiac trunk (20,21) and because the diameter of the celiac trunk increased significantly at end-systole compared to that at end-diastole (21). The advantages of Doppler ultrasound include its lack of invasiveness and the ease with which it is performed. Although it cannot determine the status of vessels during inspiratory and expiratory phases, it can be performed in a sitting or standing position.

If MALS is suspected, invasive digital subtraction angiography (DSA) or non-invasive computed tomography angiography (CTA) and magnetic resonance angiography (MRA) can be used to verify the location of the celiac trunk.

DSA can detect the presence of proximal stenosis and post-stenosis hemangiectasis and dilated vessel morphology and dynamic blood flow in the celiac trunk. In patients with MALS, the pancreaticoduodenal arterial arch in the superior mesenteric artery and retrograde filling of gastroduodenal artery into the celiac trunk can be observed. DSA can also detect the arterial pressure gradient across the origin of the celiac trunk, which helps to determine whether the celiac trunk is compressed (1). Kalapatapu et al. (22) proposed a new diagnostic method involving the administration of vasodilators via a catheter into the superior mesenteric artery during angiography. The method yields a positive result if the patient's symptoms reappear and the collateral blood reperfusion is absent. The study population consisted of 8 patients, 4 of whom had positive results. Three of those patients were cured after surgical alleviation of compression. Symptoms had other causes in 2 of the 4 patients who had negative results. Given the small sample size, the reliability of this method must be confirmed with additional observations.

CTA and MRA are forms of non-invasive imaging to identify celiac trunk compression. The advantages of CTA are that the examination is quick and inexpensive and that it better depicts intra-abdominal structures.
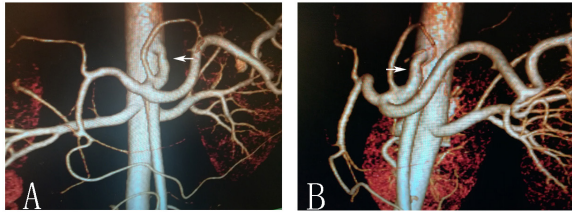
Figure 1. Three-dimensional rendering of the celiac artery according to CTA. (A) Anterior view and **(B)** Lateral view of the celiac artery showing stenosis of the celiac artery. The celiac artery was twisted and overlapping over its entire length.

With the development of CT equipment and CT image processing technology, thin-layer multi-slice spiral CT combined with a 3D reconstruction technique can clearly depict the aorta and its branches, and it has basically replaced traditional aortography (*23*). The sagittal view is the ideal orientation in which to observe the proximal end of the celiac trunk, and celiac trunk compression can be observed in the anterior side at the proximal end in patients with MALS. Hook-like stenosis (which can be distinguished from stenosis due to other causes, such as atherosclerotic stenosis) is evident in the celiac trunk. The morphology of the median arcuate ligament and collateral vessels can be observed in addition to three-dimensional rendering (*1*). CTA is often performed in the inspiratory phase because celiac trunk compression is more severe in the expiratory phase; evidence of compression in the inspiratory phase suggests MALS (Figure 1). Manghat *et al*. used ECG-gated CTA to ascertain changes in celiac trunk flow dynamics and abdominal organ supply in the cardiac cycle, but its diagnostic significance in MALS should be studied further (*21*). Compared to CTA, MRA takes longer for a scan and is more expensive, but its spatial resolution and depiction of calcified plaques are inferior to CTA. However, the absence of radioactivity during MRA and the fact that the contrast agent does not contain iodine make it suitable for use in children, pregnant women, patients allergic to iodine-containing contrast media, and patients with impaired renal function (*24*).

4. Treatment of and prognosis for MALS

Due to the different definitions of MALS and different criteria for subject selection, the appropriate treatment for MALS is a subject of debate.

In light of the pathogenicity of MALS, treatment needs to alleviate celiac trunk compression. Since compression of the celiac trunk is physical rather than due to intravascular lesions, such as atherosclerosis, use of percutaneous balloon dilation or placement of a stent is not feasible (*25*). A common procedure is separation of the ligament fibers and other surrounding tissues around the beginning of the celiac trunk in
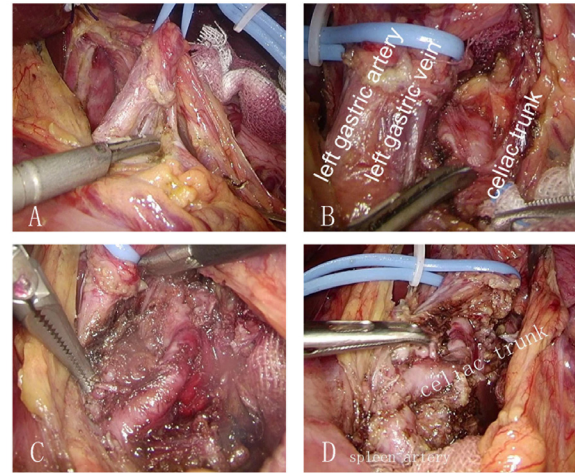
Figure 2. Laparoscopic treatment of median arcuate ligament syndrome. (A), Isolation of the left gastric vessels, which were elevated and held in place with a band; **(B)**, Dissection of the left gastric vessels to the root, and exposure of the celiac trunk. **(C)**, The celiac trunk was compressed by the median arcuate ligament around its circumference. Incision of the ligament with an ultrasonic scalpel to the root of the celiac trunk, and exposure of the aorta; **(D)**. Continued incision of the ligament to the root of the splenic artery at the distal end.

order to relieve compression. In some hospitals, removal of the celiac plexus is usually performed given ischemia leading to celiac trunk compression and involvement of the nerve plexus. In addition, celiac trunk dilation might be performed in a small number of procedures, or through vascular anastomosis, bypass, and reconstruction of the celiac trunk, which can be achieved by either laparotomy or laparoscopic surgery (*5*). Compared to laparotomy, laparoscopic surgery may reduce surgical trauma and patient hospitalization, improve the safety of the surgery, and ultrasound could be used to assist in confirming the opening of the celiac trunk (*26,27*). In addition, laparoscopic surgery could be performed with the da Vinci Surgical System as a form of robotic-assisted surgery (*28,29*), with improved surgical sensitivity and a wider visual field in comparison to conventional laparoscopic procedures.

Jimenez *et al*. (*5*) reviewed the English literature on MALS surgery and laparoscopic surgery between 1963 and 2012 and they analyzed postoperative outcomes in 400 patients, procedure details, and intraoperative and postoperative complications. The procedure was mainly the release of the median arcuate ligament; the celiac ganglia were removed or blood flow in the celiac trunk was restored in some patients. Results indicated that 85% (339/400) of patients had immediate postoperative relief of symptoms, and the rate of symptom recurrence was 6.8% (19/279) in patients who underwent a laparotomy and 5.7% (7/121) in those who underwent laparoscopic surgery. Restoring blood flow was not more likely to relieve symptoms compared to simple release of the median arcuate ligament; the incidence of complications was 11.6% for laparoscopy

and 6.5% for open surgery. The common complications of laparoscopy mainly included bleeding and pneumothorax, postoperative complications included pancreatitis and gastroparesis (in 1 patient each), and laparoscopy had to be converted to open surgery in 9.1% of patient (11/121) due to bleeding. The primary complications of open surgery included post-operative vascular thrombosis, stroke, and gastroesophageal reflux. There were no procedure-related deaths due to either procedure. Laparoscopic surgery is performed at the authors' facility to treat MALS, and patient prognosis is good (Figure 2).

Reilly *et al.* (*30*) analyzed factors that may affect the outcomes of surgery for celiac trunk compression, and results suggested that factors for a better prognosis included postprandial abdominal pain (81% cure rate), an age between 40 and 60 years (77% cure rate), weight loss greater than 20 pounds (67% cure rate), and no history of mental illness or alcohol abuse. However, this study can only be used as reference due to the small sample, the fact that multivariate statistical analysis was not performed, different procedures performed, and different definitions of "cured." The key to a good prognosis is the identification of "true" patients and the development of appropriate treatments. Given neurogenic involvement in MALS, Sultan *et al.* (*31*) contended that the abdominal nerve plexus could be routinely removed in addition to release of the ligament. Additional clinical experience is needed to devise an "Optimal Treatment." If an aneurysm has formed in a collateral, it might rupture, so the pathogenesis of MALS needs to be quickly determine and treatment needs to be performed as soon as possible.

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