Supradiaphragmatic origin of the celiac trunk leading to median arcuate ligament syndrome with superior mesenteric artery involvement

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ABSTRACT

Median arcuate ligament (MAL) syndrome (MALS) is a rare condition caused by compression of the celiac artery by the MAL. Symptoms include abdominal pain, nausea, and weight loss. Rarely, the MAL can compress both the celiac artery and the superior mesenteric artery (SMA). We describe the case of a young man with MALS involving the celiac artery and SMA. Laparoscopic release of the MAL was performed, and the patient had resolution of his symptoms at 6 months of follow-up. A review of the literature identified only six cases of MALS involving the SMA and celiac artery, making this a rare occurrence. (J Vasc Surg Cases Innov Tech 2024;10:101315.)

Keywords: Dunbar syndrome; MAL; MALS; Median arcuate ligament; Median arcuate ligament syndrome; Supradiaphragmatic celiac trunk; Supradiaphragmatic SMA; Two-vessel MALS

Median arcuate ligament (MAL) syndrome (MALS) is a rare condition caused by MAL compression of the proximal celiac artery. The condition presents with postprandial abdominal pain, nausea, vomiting, food aversion, and weight loss. Given the nonspecific clinical features, MALS represents a diagnostic challenge. It typically requires extensive workup to rule out other causes of abdominal pain combined with imaging findings of vessel compression by the MAL.[1] Treatment is surgical decompression of the MAL to release the compressed vessel. Rarely, compression of both the superior mesenteric artery (SMA) and celiac artery can occur in patients with an unusually high origin of both vessels. This is an uncommon presentation that has only been described in a few cases in the literature. We present the case of a 19-year-old man with symptoms of abdominal pain, nausea, vomiting, and weight loss from compression of the celiac artery and SMA. We additionally present a comprehensive review of cases with similar two-vessel compression reported in the literature. The patient provided written informed consent for the report of his case details and imaging studies.

CASE REPORT

A 19-year-old man with no significant medical or surgical history presented with 1 year of postprandial abdominal pain, nausea, vomiting, diarrhea, and a 20-lb weight loss due to food aversion. Before surgical consultation, he underwent an extensive gastrointestinal workup that included laboratory and endoscopic investigations, which demonstrated no evidence of inflammatory bowel disease or chronic disease to explain his symptoms. The laboratory workup included deamidated gliadin antibodies, immunoglobulin G, immunoglobulin A, immunoglobulin M, tissue transglutaminase, C-reactive protein, sedimentation rate, folate, vitamin D, vitamin B_{12}, human immunodeficiency virus, zinc, complete blood count, and comprehensive metabolic panel. All values were within normal limits. The findings from esophagogastroduodenoscopy with tissue biopsy, colonoscopy, capsule endoscopy, and computed tomography (CT) enterography were unremarkable. CT angiography showed an unusual supradiaphragmatic origin of the celiac axis and severe stenosis of the SMA by the MAL (Fig 1 and Supplementary Video 1, online only). The estimation by CT showed 25% stenosis of the celiac artery at the crossing of the hemidiaphragm and 50% to 60% stenosis of the SMA. Doppler ultrasound confirmed compression of the celiac artery by the MAL. Also, the peak systolic velocities were elevated on expiration (peak systolic velocity: SMA, 521 cm/s; celiac artery, 415 cm/s; Supplementary Video 2, online only). The decision was made to perform laparoscopic exploration and release of the MAL and complete neurolysis of the celiac plexus. The procedure was performed by a general surgeon trained in minimally invasive surgery, with vascular surgery on standby for potential intraoperative complications. The procedure was conducted under general anesthesia with the patient placed in a modified lithotomy position in stirrups. A five-port technique was used, with the trocar sites highlighted in Fig 2. Intraoperative findings confirmed compression of the celiac axis and SMA by the MAL, which was

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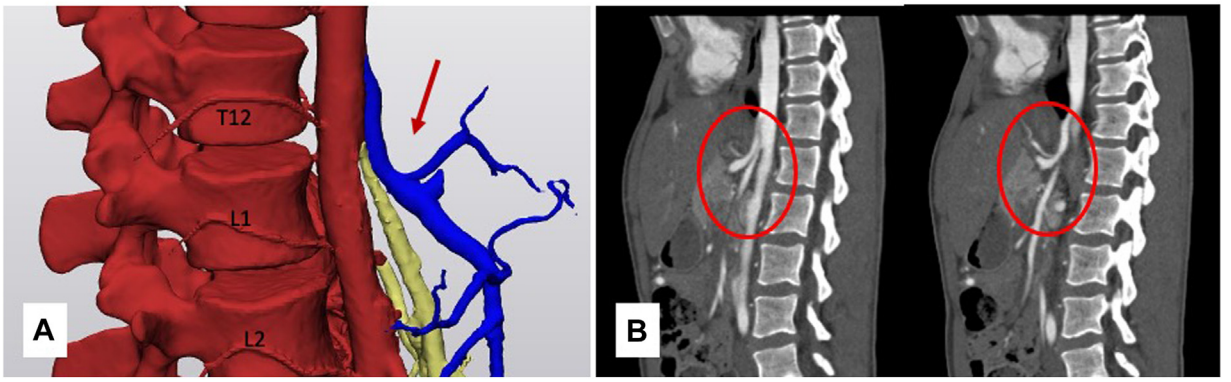

Fig 1. Three-dimensional rendering and computed tomography (CT) angiography of the celiac axis and superior mesenteric artery (SMA). **A,** Mimics Innovation Suite software (Materialise NV) was used to create a digital model from the CT angiogram. *Blue* indicates the celiac artery and its bifurcations; *yellow,* the SMA; and *red arrow,* downward compression of the celiac trunk and SMA by the hemidiaphragm. **B,** CT angiogram of the celiac axis and SMA showing 25% stenosis of the celiac artery as it crosses the region of the hemidiaphragm and 50% to 60% stenosis of the SMA.

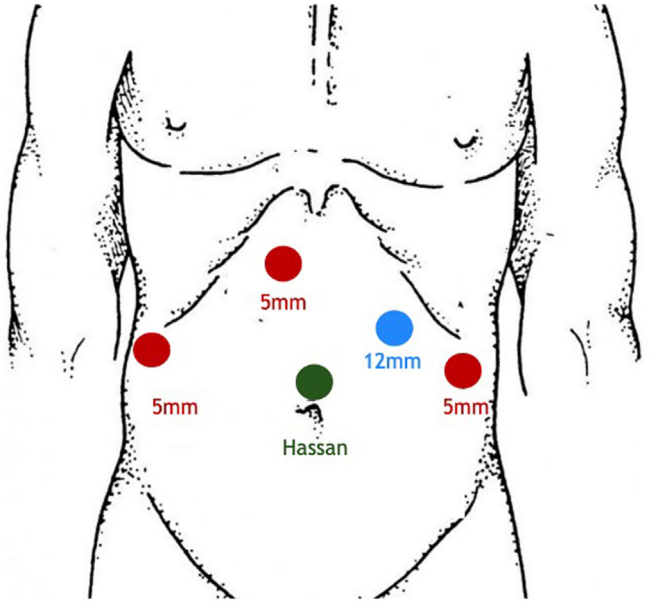

Fig 2. Diagram showing trocar positions intraoperatively, including the Hassan port, supraumbilical, 12 cm from the xiphoid process; three 5-mm trocars located at the intersection of the patient's right subcostal and anterior axillary line, right subcostal and right parasternal line, and left subcostal and anterior axillary line; and a 12-mm trocar placed at the left subcostal midclavicular line.

released, with complete lysis of the celiac plexus (Fig 3 and Supplementary Video 3, online only). The patient's postoperative course was uneventful, and he was discharged on postoperative day 1. The patient declined any additional testing after surgery. At 2 weeks of follow-up, he had experienced significant improvement in his symptoms. At 6 months of follow-up, he had gained 20 lb and remained symptom free.

DISCUSSION

MALS, also known as Dunbar syndrome and celiac compression syndrome, is an uncommon vascular disorder. The MAL is a fibrous arch connecting the right and left crura of the diaphragm at the level of the aortic hiatus (T12-L1). The MAL traverses anterior to the aorta and is usually cranial to the celiac trunk.[2] The syndrome is thought to be caused by vascular compression exerted by the MAL in patients with an abnormally low insertion of the diaphragm or an abnormally high origin of the celiac trunk from the aorta.[3] The celiac ganglion, which lies anterior to the aorta and is composed of afferent fibers from the upper abdominal viscera and sympathetic fibers from the greater and lesser splanchnic nerves, can also be compressed, contributing to the resulting symptoms.[1] The amount of compression can vary during the respiratory cycle: with expiration, the aorta and celiac artery move superiorly, and the MAL moves dorsally, causing compression of the visceral vessels or complete arterial occlusion.[3]

Most commonly, patients with MALS experience compression of the celiac artery alone. However, a few cases have been reported in the literature, including the present patient, with the MAL compressing both the celiac artery and the SMA. A review of the literature is summarized in the Table, including, to the best of our knowledge, six reported cases of MALS with combined celiac artery and SMA involvement. Studies have reported triple entrapment syndrome with MAL compression of the celiac trunk and SMA and a third entrapment resulting from the severely acute angle of the SMA compressing the duodenum.[8,9]

Symptoms of MALS are classically described as postprandial abdominal pain, nausea and vomiting, food aversion, and weight loss. In addition, some studies

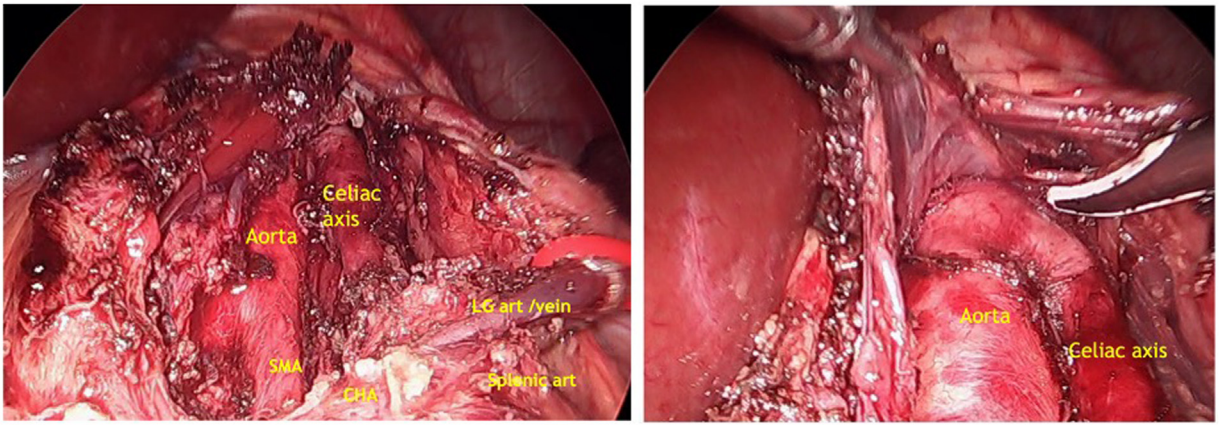

Fig 3. Intraoperative findings. *CHA,* Common hepatic artery; *LG art/vein,* left gastric artery/vein; *SMA,* superior mesenteric artery.

Table. Summary of clinical features from previous reports of median arcuate ligament (MAL) syndrome (MALS) with celiac trunk and superior mesenteric artery (SMA) involvement

			Presenting symptoms									
Investigator	Age, years	Sex	AP	DFI	WL	Other	Anatomic details	Treatment: surgical decompression of MAL	Surgical approach	Additional surgery	Postoperative course	Symptom resolution
Curl et al,[4] 1970	39	F	Yes	Yes	Yes (10 lb ≤3 months)	Constipation	Celiac artery and proximal SMA compressed against aorta by MAL	Yes	Transabdominal	Right segmental renal artery aneurysmectomy	Uncomplicated	Yes
Lawson et al,[5] 1984	37	M	Yes	Yes	Yes (25 lb ≤4 months)	No	Total occlusion of origin of celiac axis and tight stenosis of SMA	Yes	Transabdominal	No	Uncomplicated	Yes
Stein et al,[3] 2011	40	M	Yes	No	Yes (60 lb ≤1 year)	No	Tendinous band over celiac artery and SMA	Yes	Retroperitoneal	No	Uncomplicated	Partially; intermittent abdominal pain, nausea, vomiting at 1 year of follow-up
Doyle et al,[6] 2012	26	M	Yes	Yes	Yes (40 lb ≤6-8 months)	No	Thick band compressing celiac artery and SMA	Yes	Retroperitoneal release and SMA bypass	Retrograde aorto—celiac and aorto—SMA bypass	Readmitted 4 months postoperatively with stenosis of aorto—SMA bypass; treated with balloon angioplasty	Yes
Lee et al,[2] 2016	47	F	Yes	No	No	Nausea; diarrhea	Adhesions encasing celiac artery and SMA	Yes	Retroperitoneal	Limited endarterectomy and patch angioplasty of SMA	Uncomplicated	Yes
Shao et al,[7] 2020	46	M	Yes	No	Yes (37 lb ≤1 year)	Nausea	Ligamentous compression of SMA and celiac artery	Yes	Laparoscopic release of MAL and SMA stenting with percutaneous brachial access	No	Uncomplicated	Yes

AP, Abdominal pain; *DFI,* decreased food intake; *F,* female; *M,* male; *WL,* weight loss.

have described an epigastric bruit on physical examination.[4] Most often, MALS affects women aged 20 to 40 years.

MALS is typically a diagnosis of exclusion combined with imaging findings of vessel compression by the MAL.[10] Symptoms can vary between patients, and other causes of chronic abdominal pain must be ruled out.[2] Historically, the original report of MALS was diagnosed using visceral angiography with a finding of narrowing of the celiac artery.[11] In recent years, CT angiography has emerged as the imaging modality of choice, because it allows for three-dimensional reconstruction of the compressed vessel and direct visualization of the MAL.[7] Imaging findings of a "hooked" appearance of the celiac artery by the ligament aid in distinguishing MALS from atherosclerotic stenosis.[7]

The mainstay treatment of MALS is surgery. Treatment consists of division of the MAL and crus of the diaphragm, followed by celiac ganglion neurolysis.[12] It is still unclear how much of a role celiac ganglion lysis plays in the symptomatic relief of MALS. A retrospective cohort study of 81 patients with clinical symptoms of MALS showed that although only 22% of patients had confirmed celiac artery compression found on imaging, 86% had symptom relief after percutaneous celiac plexus block.[6] In cases of persistent stenosis after MAL release or unresolved symptoms, additional revascularization with angioplasty or bypass of the celiac artery can be performed.[13]

Traditionally, the surgical approach for MAL release has been transabdominal and retroperitoneal, with a switch in recent years to laparoscopy and robotic-assisted approaches.[1,3] In a recent study by Kazmi et al,[13] laparoscopic transperitoneal decompression of MALS was found to provide most patient with persistent relief of symptoms. Of 52 patients, 90% of patients had achieved symptom relief either completely or partially at 3 to 6 months of follow-up. Moreover, a laparoscopic approach resulted in a shorter operation time and hospital length of stay, with most patients discharged from the hospital the day after surgery.[5]

CONCLUSIONS

MALS is a rare condition that is often challenging to diagnose. We presented an unusual case of MALS with involvement of both the celiac artery and the SMA and present a comprehensive review of similar cases reported in the literature. Although uncommon, it is important to remember the differential diagnosis of MALS when encountering patients with nonspecific abdominal symptoms and an unexplained etiology.

DISCLOSURES

None.

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