

Intravascular ultrasound-guided laparoscopic division of the median arcuate ligament

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ABSTRACT

Median arcuate ligament syndrome is a rare and poorly understood condition that can have a significant impact on the quality of life of patients. Diagnosis is often difficult and delayed because of the need to exclude other pathologic processes. Treatment strategies traditionally involve open or laparoscopic division of the median arcuate ligament, with or without vascular reconstruction. This report portrays a case of median arcuate ligament syndrome with compression of two visceral arteries and distal embolic complications. A novel hybrid technique is described using intravascular ultrasound technology to aid in laparoscopic median arcuate ligament division. This allowed real-time intravascular visualization of the compressive segment, guided release of the ligament fibers, and demonstrated confirmation of decompression. (*J Vasc Surg Cases and Innovative Techniques* 2020;6:147-51.)

Keywords: Vascular surgery; Intravascular ultrasound; Median arcuate ligament syndrome

Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome, is a rare condition caused by compression of the celiac axis by the fibrous arch of the median arcuate ligament to form the diaphragmatic crura. This is seen anatomically in 10% to 24% of the population but is symptomatic in only a small proportion (2/100,000 patients).¹⁻³ Symptoms most commonly affect young women between 20 and 40 years of age; it typically is manifested with nonspecific postprandial abdominal pain, vomiting, and weight loss.^{1,2} Diagnosis is challenging and often made after exclusion of other causes. Traditionally, open surgical division of the median arcuate ligament has been the mainstay of treatment.^{4,5} In recent years, laparoscopic decompression of the celiac artery has become increasingly reported in practice. The emergence of new technologies also allows more novel approaches to surgical treatment. We present a novel technique in which real-time intravascular ultrasound (IVUS) was used to guide laparoscopic division of the median arcuate ligament. The patient's consent has been obtained for this publication.

CASE REPORT

Our patient is a 42-year-old, otherwise healthy woman who was found to have had a spontaneous partial splenic infarct approximately 2 years ago after presenting with acute abdominal pain. Computed tomography angiography (CTA) revealed a celiac axis thrombus immediately distal to an area of focal compression. She was prescribed anticoagulation, yet extensive cardiac and hematologic investigations for arrhythmias and thrombophilic disorders yielded no clear cause.

The patient remained asymptomatic until recently, when she began to experience exercise-induced abdominal pain. Although she initially denied postprandial pain, this developed rapidly within the following weeks and led to significant intolerance of oral intake and consequent weight loss. Repeated CTA demonstrated compression of the celiac axis and resolution of the initial thrombus, and she was referred to a vascular surgeon with suspected MALS.

The patient was admitted for further investigation and treatment of her symptoms. Findings on gastroscopy were normal, and abdominal ultrasound examination found gallbladder sludge without evidence of cholelithiasis or cholecystitis. In the presence of normal blood results, this was deemed by the general surgical team to be an unlikely cause of the patient's presentation. Subsequently, mesenteric digital subtraction angiography (DSA) was performed with breathing maneuvers, which clearly demonstrated significant stenosis of the origin of the celiac axis during full inspiration and complete occlusion in all other phases of respiration (Fig 1). Moreover, the angiogram also demonstrated 50% stenosis of the superior mesenteric artery (SMA) during respiration.

This confirmed a diagnosis of MALS, and following multidisciplinary discussion between the vascular surgery, general surgery, and hematology teams, a decision was made to proceed with combined endovascular and laparoscopic surgery using IVUS technology to guide release of the median arcuate ligament 4 days later. The rationale for using IVUS was to add a further degree of

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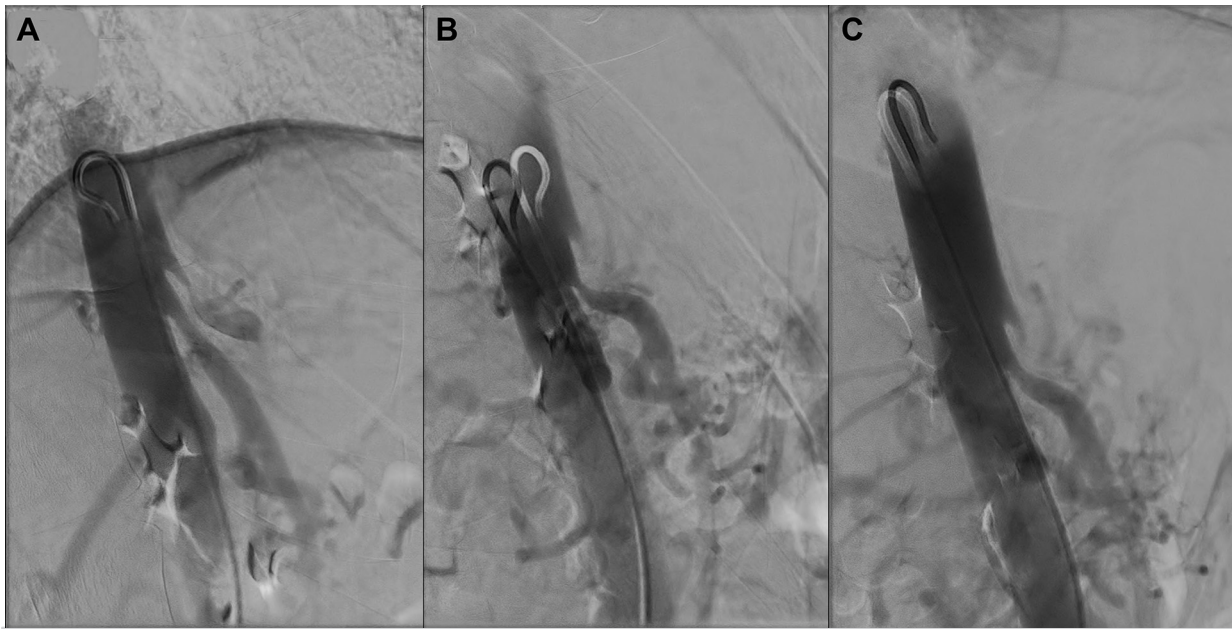


Fig 1. Digital subtraction angiography (DSA) of the aorta and mesenteric arteries. **A**, During deep inspiration, significant stenosis of the celiac artery is seen. **B**, During deep expiration, complete occlusion of the celiac artery is seen with >50% stenosis of the superior mesenteric artery (SMA). **C**, During normal respiration, complete occlusion of the celiac artery is seen with 50% stenosis of the SMA.

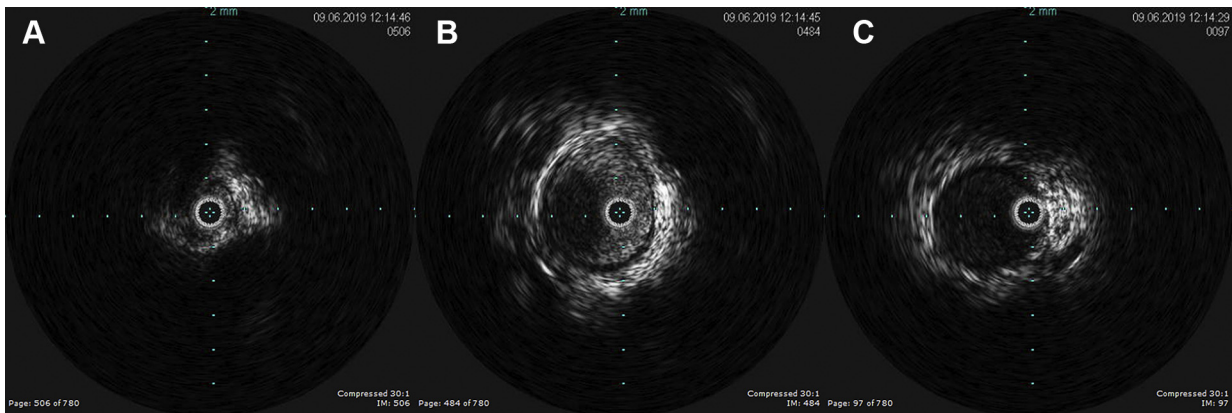


Fig 2. Intravascular ultrasound (IVUS) images of the celiac axis intraoperatively. **A**, At the point of complete occlusion of the celiac artery. **B**, Immediately distal to the point of celiac axis occlusion, there is an area of slow vascular flow/thrombus as demonstrated by the heterogeneous intraluminal appearance. **C**, Post-stenotic dilation of the celiac axis.

visualization to the operation, in particular a luminal representation of the exact location that required extrinsic decompression.

In a Philips hybrid operating suite (Philips Healthcare, Best, The Netherlands), the patient was positioned on the angiography table in lithotomy position with her left arm abducted for vascular access. Her brachial artery was punctured under ultrasound guidance and a 6F sheath inserted. Mesenteric DSA was performed, and the celiac axis was accessed using a 0.018-inch guidewire and corresponding IVUS catheter. IVUS measurements demonstrated a 2-cm length of complete occlusion of the celiac axis with post-stenotic dilation (Fig 2) secondary to extrinsic compression from the median arcuate ligament. Five-port laparoscopic access was gained into the

abdomen, and the lesser sac was opened. The left gastric artery was identified and traced to its origin to the celiac axis. Celiac splanchnic nerves were split, and the ganglion was excised. The fibers of the median arcuate ligament were gradually divided cephalad toward the aorta guided by IVUS visualization of the celiac axis (Fig 3). During the procedure, on laparoscopic visualization, there seemed to be adequate release of the arcuate ligament; IVUS was able to demonstrate remnant compression and its exact location in relation to the origin of the celiac artery. This information helped guide further clearance of bands of the median arcuate ligament by laparoscopic division and allowed confirmation of the complete release of the vessel. A subsequent increase in cross-

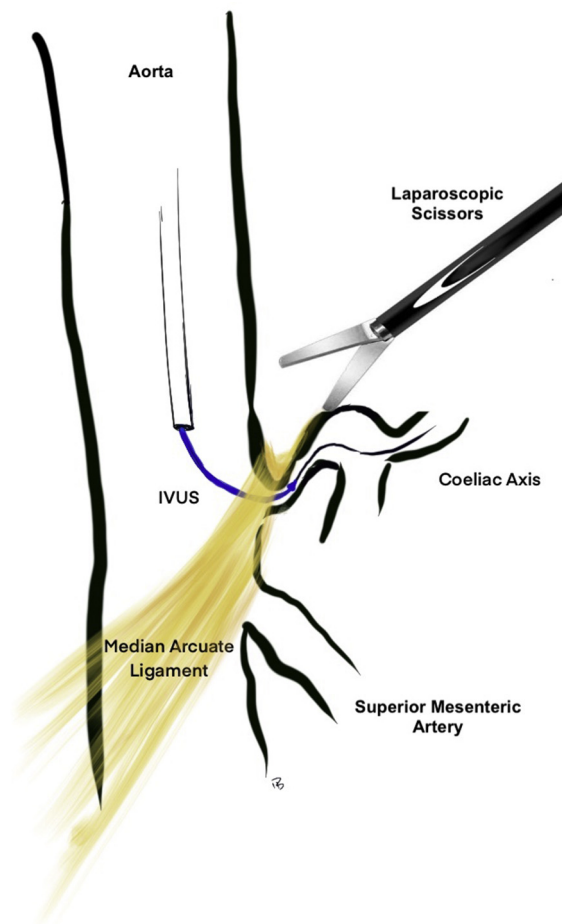


Fig 3. Schematic illustrating the technique of laparoscopic release of the median arcuate ligament using intravascular ultrasound (IVUS) guidance.

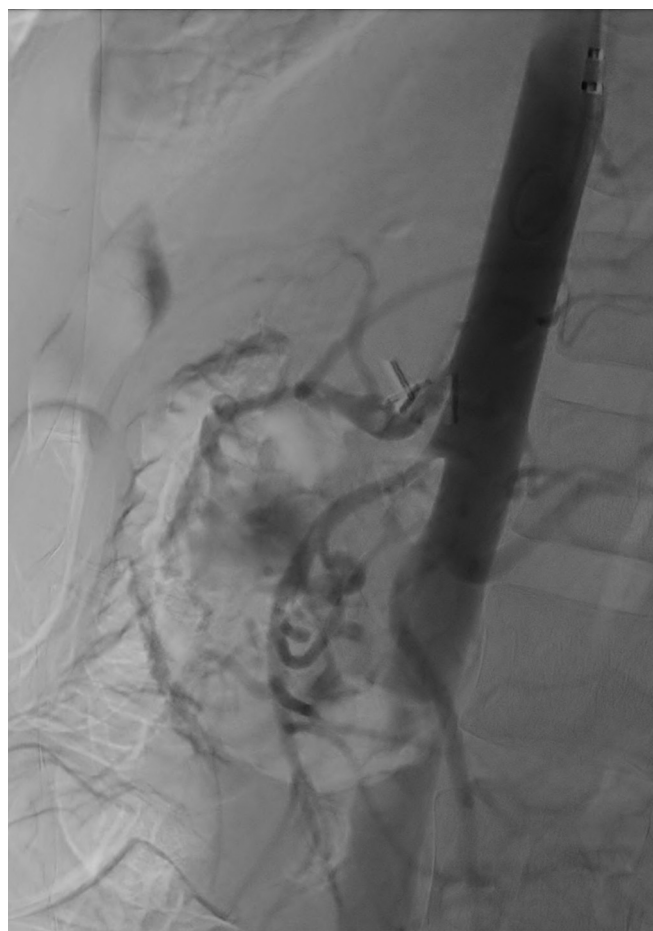


Fig 4. Digital subtraction angiography (DSA) of the aorta and mesenteric arteries after intravascular ultrasound (IVUS)-guided laparoscopic median arcuate ligament release illustrating residual stenosis of the celiac artery secondary to chronic fibrosis.

sectional area across the point of obstruction was seen in IVUS, although $>50\%$ stenosis of the artery remained, probably secondary to chronic fibrosis (Fig 4). The compressive stenosis of the SMA was also seen to be improved on DSA (Fig 4).

After release of the median arcuate ligament, despite experiencing overall improvement in her abdominal pain, the patient continued to report mild pain postprandially. A decision was made to proceed with further endovascular intervention. Several days later, the patient underwent endovascular stent insertion across the area of residual stenosis in the celiac axis with a 7- \times 19-mm Omnilink Elite (Abbott Vascular, Abbott Park, Ill) balloon-expandable stent. Postintervention DSA demonstrated a widely patent celiac artery and excellent perfusion of distal organs (Fig 5).

The patient was discharged after several days of observation and returned for follow-up assessment. She reported significant improvement in her symptoms and was now able to tolerate food without postprandial pain as well as to return to her routine exercise regimen. Interval ultrasound and CTA at 4 weeks demonstrated a widely patent celiac artery stent.

DISCUSSION

MALS, a rare condition with nonspecific symptoms, is often a diagnostic challenge for clinicians. The underlying pathophysiologic mechanism is controversial and has been heavily debated in the literature. It is most commonly postulated to be due to demand ischemia secondary to external compression of the celiac axis by the crural fibers of the median arcuate ligament, particularly in individuals who have an unusually high celiac axis origin or low insertion of the median arcuate ligament.^{1,6,7} Alternatively, there are also hypotheses about neurogenic mechanisms.^{7,8} Left untreated, individuals with MALS may suffer complications such as gastroparesis and arterial aneurysms.^{9,10} Visceral thromboembolic events are rare sequelae, and interestingly, splenic infarcts as a result of MALS, as seen in this case, have not been previously described in the literature.

The diagnosis of MALS is challenging as there are no universal diagnostic criteria, and it requires exclusion of other abdominal diseases. Commonly, a combination



Fig 5. A, Digital subtraction angiography (DSA) showing position of a 7- × 19-mm Omnilink Elite (Abbott Vascular, Abbott Park, Ill) balloon-expandable stent within the origin of the celiac axis. **B,** DSA after stent insertion demonstrating a widely patent celiac artery and excellent perfusion of distal organs despite a degree of vasospasm of the common hepatic artery.

of Doppler ultrasound, CTA, and magnetic resonance angiography is used to preliminarily characterize the location and degree of celiac compression.^{1,7} Invasive mesenteric DSA, with multiple orthogonal views, allows dynamic visualization of the arterial compression using respiratory maneuvers.^{2,7} In this case, 50% stenosis of the SMA was seen in addition to occlusion of the celiac axis. This concurrent compromise of both foregut and midgut blood flow in MALS has been scarcely reported in the literature^{9,10} and may be an important indicator for greater disease severity. Given the difficulties in diagnosing MALS, involvement of both the SMA and celiac axis may represent a more specific finding in the diagnosis of this condition.

Currently, there are only studies, both published in 2014, describing the use of IVUS technology in the diagnosis of MALS.^{11,12} Both reported superior quantification of the exact percentage of celiac stenosis on IVUS using cross-sectional area measurements compared with conventional DSA.^{11,12}

The aim of MALS treatment is to relieve the celiac artery compression. This is traditionally achieved through surgical division of the median arcuate ligament by an open laparotomy.^{1,3,13} Neurolysis and excision of the celiac plexus are also usually performed to address any neuropathic process.⁷ After decompression, vascular reconstruction is often necessary because of structural changes in the vessel secondary to chronic compression leading to intimal hyperplasia and persistent stenosis.^{1,6} Options include endovascular angioplasty or stenting of

the stenotic segment, arterial bypass, and reimplantation of the vessel. Laparoscopic release of the median arcuate ligament has become increasingly prevalent in practice, with the associated benefits of reduced morbidity and shorter recovery time.^{4,5,14} However, there is the possibility of incomplete ligament release resulting in persistent symptoms, with recurrence of symptoms reported in between 5.7% and 30% in the literature.^{4,5,14,15}

This case report is the only one in the published literature that describes the use of IVUS technology to guide laparoscopic division of the median arcuate ligament. This not only allowed precise localization of the point of arterial compression but also provided real-time feedback regarding the success of the release. There were several instances during the procedure when the artery was thought to be completely freed but significant compression was still seen on IVUS. Exploration revealed additional fibrous bands that required further division. Immediate confirmation of the successful decompression of the celiac artery was possible using IVUS, which demonstrated significant improvement at the site of previous complete obstruction. Endovascular stent insertion was later performed as an adjunct to treat residual stenosis secondary to chronic compression. Despite this, the use of IVUS was highly beneficial to the initial operation and contributed to improving surgical accuracy and ease.

CONCLUSIONS

MALS is a distressing and debilitating condition with various postulated causes. Diagnosis is often difficult

because of the need to exclude other pathologic processes, and investigations can often be inconclusive. Concurrent involvement of both the celiac axis and SMA, as seen in this case, may represent a more specific finding and be important in advancing nuances around diagnosis. Because of the rarity of this disease, there is a paucity of evidence regarding the best approach to treatment. The development of IVUS technology allows real-time guidance of laparoscopic release of the median arcuate ligament, offering immediate confirmation of successful decompression. This, coupled with endovascular reconstruction, presents a hybrid, minimally invasive approach to the treatment of MALS. The use of IVUS is a novel and previously unpublished technique that the authors believe should be considered in all cases of median arcuate ligament release.

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