Case Reports

Median Arcuate Ligament Syndrome

Confirmed with the Use of Intravascular Ultrasound

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Median arcuate ligament syndrome, a rarely reported condition, is characterized by postprandial abdominal pain, nausea, vomiting, and weight loss. Its cause is unclear. We present the case of a 45-year-old woman who had intermittent chronic positional abdominal pain without weight loss. Magnetic resonance angiograms and computed tomograms revealed stenosis of the celiac artery. Ostial compression was confirmed on catheter angiographic and intravascular ultrasonographic images. Intravascular ultrasound revealed far greater stenosis than did the initial imaging methods and confirmed a diagnosis of median arcuate ligament syndrome. In lieu of surgery, the patient underwent a celiac ganglion block procedure that substantially relieved her symptoms.

To our knowledge, this is the first report of the use of intravascular ultrasound in the diagnosis of median arcuate ligament syndrome. We recommend using this imaging method preoperatively in other suspected cases of the syndrome, to better identify patients who might benefit from corrective surgery. ***(Tex Heart Inst J 2014;41(1):57-60)***

Key words: *Abdominal pain/etiology; arterial occlusive diseases/diagnosis/pathology/physiopathology; celiac artery/pathology/physiopathology/ultrasonography; constriction, pathologic; diagnostic imaging; ligaments/pathology; mesenteric vascular occlusion/complications/etiology; ultrasonography, intravascular*

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Median arcuate ligament syndrome (MALS), also called celiac artery compression syndrome, is a rarely reported condition characterized by postprandial abdominal pain, nausea, vomiting, and weight loss. The cause of these symptoms has not been determined, and different anatomic explanations have been formulated.[1] Because the cause of MALS is unclear and symptoms can persist even after surgical intervention, its diagnosis can be controversial.[2] Advances in imaging techniques and minimally invasive studies have created a niche for MALS in the differential diagnosis of chronic abdominal pain of unclear origin.[3]

We present the case of a woman who had celiac artery compression and stenosis with an atypical clinical presentation and evidence of compression upon expiration.[3] The stenosis was initially detected by means of computed tomography (CT) and magnetic resonance angiography (MRA) and was confirmed by means of catheter angiography and intravascular ultrasound (IVUS). Besides the case of the patient, we discuss the usefulness of IVUS in the diagnosis of MALS.

Case Report

In November 2011, a 45-year-old obese woman with a 5-year history of intermittent positional abdominal pain and nausea presented at our institution for evaluation, after results of MRA were deemed abnormal. The patient's pain was localized in the epigastric area and was aggravated by bending over, sitting, and doing housework. Despite postprandial abdominal pain that reportedly occurred about half the time, she had gained 40 lb in 7 or 8 months from eating mostly bland, high-calorie food. She reported severe constipation, 3 episodes of pancreatitis since 2008, a history of gastroesophageal reflux disease, and a sliding hiatal hernia. Her medical history included remote deep vein thrombosis and pulmonary embolism, and a cholecystectomy in 2002. She was heterozygous for factor V Leiden, had a prothrombin *20210* mutation, and had a *MTHFR* gene variant. She had undergone esophagogastroduodenoscopy, colonoscopy, MRA studies in 2009 and 2011, and multiple CT scans (Fig. 1). She was currently taking warfarin, omeprazole, atenolol, amitriptyline, temazepam, propantheline bromide, and paroxetine.

Physical examination revealed an overweight patient and nothing else unusual. The patient's most recent MRA and CT results suggested mild tortuosity and a 40% to 50% stenosis at the origin of the celiac artery. A new duplex ultrasound study of

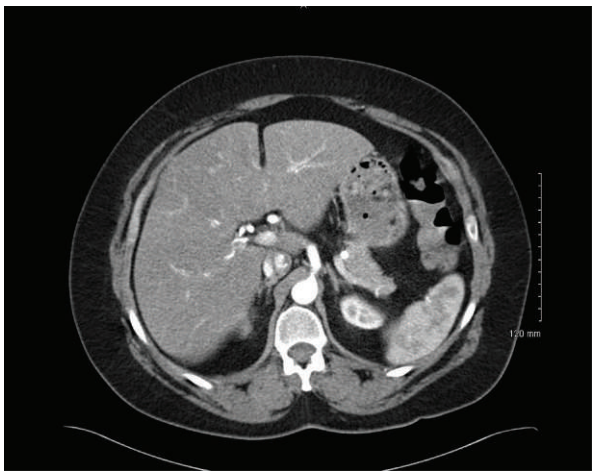

Fig. 1 *Axial computed tomographic angiogram from February 2011 shows narrowing of the proximal celiac artery.*

the abdominal arteries revealed normal flow velocities and no evident obstruction of the celiac and superior mesenteric arteries at rest.

Because MALS was suspected, the patient underwent celiac angiography. Multiple orthogonal views showed narrowing of the celiac trunk (Fig. 2) and compression of the celiac artery by the median arcuate ligament (MAL) upon expiration. We performed dynamic (inspiratory and expiratory) pressure-gradient evaluation with use of a Radi PressureWire® (St. Jude Medical, Inc.; St. Paul, Minn) without adenosine infusion; results showed a gradient of 7 mmHg. Using an Eagle Eye® Platinum Catheter (Volcano Corporation; San Diego, Calif), we then performed IVUS. This revealed substantial luminal stenosis—a minimal lumen diameter of 2.9 mm at the origin of the celiac artery with thickening of soft tissue on the superior surface of the vessel, and a 9.5-mm diameter at a distal reference point. This correlated with a nonatherosclerotic luminal narrowing of 72.2% (Fig. 3).

The patient was referred for surgical correction; however, she was a poor candidate because of her comorbidities and lack of weight loss. Instead, she underwent an endoscopic celiac ganglion block procedure with 5 mL of 0.25 bupivacaine and 80 mg of triamcinolone, which substantially relieved her symptoms. The relief lasted for approximately one week, and the symptoms gradually returned.

Discussion

We searched MEDLINE for "median arcuate ligament syndrome" and "celiac artery compression syndrome," and the MeSH database for "artery occlusive disease" and "celiac artery." We found no articles describing the use of IVUS as a diagnostic tool in MALS.

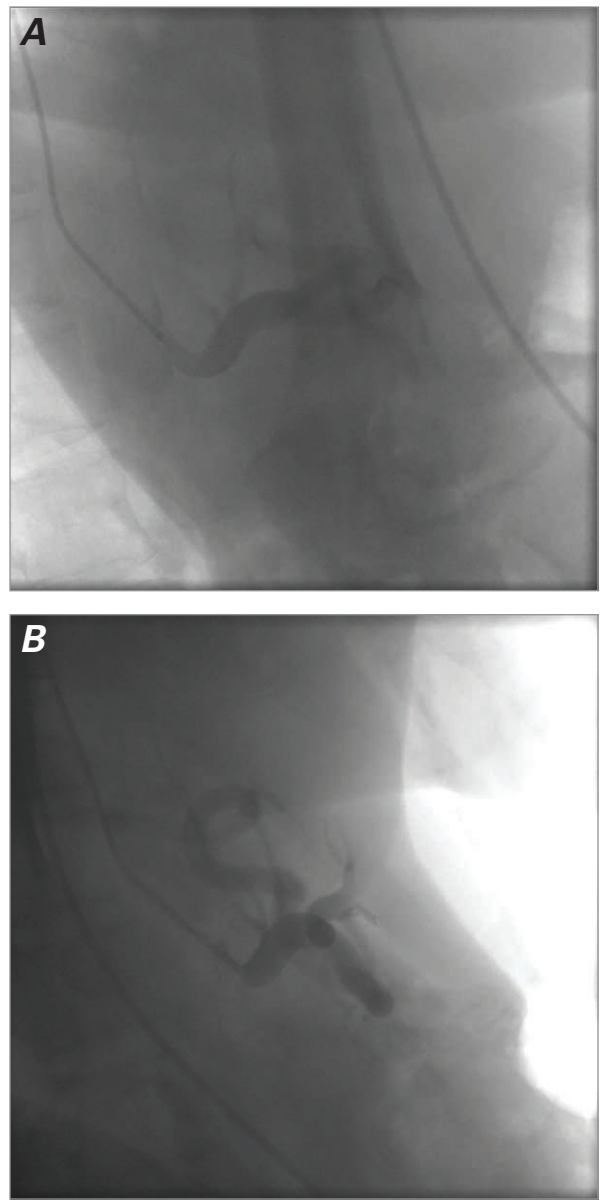

Fig. 2 *Angiograms show a multipurpose catheter engaging the celiac artery via radial access. Ostial compression was evident in the celiac artery and worsened upon expiration.* ***A****) There is tenting of the celiac artery upon inspiration, with a patent vessel.* ***B****) During the expiratory phase, apparent compression of the celiac artery causes severe stenosis.*

Median arcuate ligament syndrome was first described in 1963, in a patient whose mesenteric ischemia was thought to be caused by extrinsic compression of the celiac artery.[4] In 1965, Dunbar and colleagues[5] reported on 15 patients with MALS who underwent surgical division of the MAL; this procedure successfully decompressed the artery, restored blood flow, and relieved abdominal pain. Subsequent theories of the pathophysiology of MALS have involved various anatomic and functional effects that surrounding structures might have on the celiac trunk.[1]

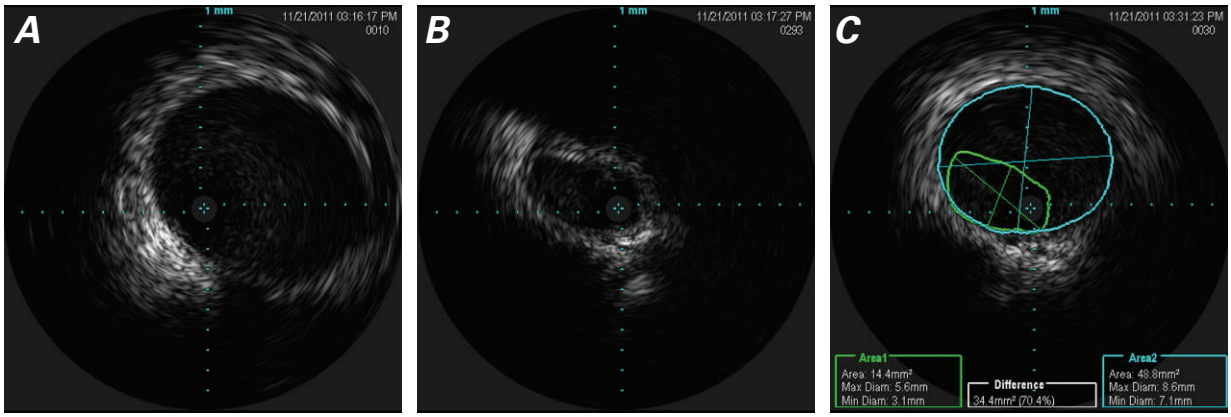

Fig. 3 *Intravascular ultrasonographic images of the celiac artery show* ***A****) the distal reference portion of the celiac artery;* ***B****) proximal hypoplasia of the celiac vessel with no intimal plaque, and hyperechoic fibrous tissue in the adventitia (right lower quadrant of image); and* ***C****) 72.2% luminal narrowing.*

Among proposed pathophysiologic mechanisms is mesenteric ischemia that is caused by extrinsic compression from the MAL or a fibrotic celiac ganglion.[6] A neurogenic process might result directly from sympathetic pain or local irritation, or indirectly from splanchnic vasoconstriction.[7] Exercise-induced MALS could result from reduced splanchnic blood flow during physical activity.[8] It is generally accepted that the classic presentation of MALS is caused by mesenteric ischemia, which initiates the recurrent postprandial symptoms of nausea, vomiting, and weight loss, with aversion to food. We infer that atypical clinical presentations, such as our patient's, could be explained by neurogenic involvement rather than a purely ischemic process. Because of the extensive collateralization of the mesenterium from the celiac, superior mesenteric, and inferior mesenteric arteries, it is controversial to conclude that isolated compression or stenosis of the celiac artery causes mesenteric ischemia.

The diagnosis of MALS is challenging and is one of exclusion, especially in the absence of typical symptoms. Suspicion of the condition warrants further invasive studies and raises the possibility that surgical intervention will be necessary. After other causes of abdominal pain have been excluded by initial imaging and endoscopic results, further studies—such as abdominal duplex ultrasound, CT angiography, and MRA of the abdomen—should be performed.[9] If an abnormality is found and mesenteric vascular disease is suspected, angiography with pressure-gradient measurement is warranted.[9] Our patient had already consulted a gastroenterologist and undergone esophagogastroduodenoscopy and colonoscopy.

In our patient, the CT and MRA evidence of celiac artery stenosis correlated in general with the IVUS evidence of stenosis and a measurable pressure gradient. The severity of the gradient is merely to be noted, because there are currently no established pressure gradients to correlate with symptomatic improvement in MALS after revascularization. However, a stenosis greater than 50% is generally considered to be clinically important when detected on vascular imaging.[10] The initial imaging methods showed a stenosis of possible significance in our patient (40% to 50%), whereas IVUS detected an indisputably severe stenosis (72.2%). In addition, a hyperechoic area was noted in the periadventitia, which could correlate with the description by Lindner and Kemprud[11] in which 15% of pathologic specimens showed a calcified plaque on the ventral surface of the vessel. The comparative sensitivity and specificity of these imaging methods could warrant study.

Anatomic variations that cause indentation of the celiac artery by the MAL are seen in the general population and might not be clinically important. In an autopsy study of 75 specimens,[11] the celiac artery originated at or above the arcuate ligament in 33% of the specimens; this was thought to be due to a lower and more anterior placement of the ligament. Isolated compression during expiration can be detected angiographically in 13% to 50% of patients, most of whom have no symptoms. Compression considered severe enough to compromise blood flow reportedly occurs in only 1% of patients.[12] Because of the effect that ventilation can have on the celiac artery, a focal narrowing that persists during inspiration might be clinically significant.[13] None of the abovementioned studies defines what degree of compression could be considered pathologic.

A secure preoperative diagnosis is important if clinicians are to decide which patients can most likely benefit from surgical intervention, and various diagnostic approaches have been developed. At the Mayo Clinic, 9 of 9 patients with MALS whose symptoms initially improved after percutaneous celiac ganglion block experienced further improvement after corrective surgery.[14] Gastric tonometry has been used to measure intestinal perfusion and to predict whether surgery might improve

a patient's outcome.[15] Investigators injected a vasodilator into the selectively cannulated superior mesenteric arteries of 8 patients with MALS, forcing blood flow away from the collateral bed and reproducing their symptoms. The test results were positive in 4 of the patients; all 4 improved after surgical MAL decompression, and 3 were asymptomatic.[16] That vasodilator test was performed conveniently during conventional angiography, as can IVUS. Diagnostic techniques for MALS continue to be designed.

Open surgery is the mainstay of treatment for the mechanical compression that defines MALS. Resecting the MAL and celiac ganglion tissue enables concurrent patch angioplasty, aortoceliac bypass, or intraluminal dilation. Newer treatment options include laparoscopic surgery with percutaneous transluminal angioplasty or stenting of the celiac artery. Purely endovascular treatment is not recommended, because stenting alone (without removing the extrinsic compression by the MAL) can lead to suboptimal results and crushing of the stent by the MAL.[17,18] As an alternative to open surgery, an intrathecal morphine pump was implanted in a patient with intractable visceral pain who already had undergone laparoscopic surgery for MALS. The patient was free of symptoms 6 months after the device was implanted.[19]

Individuals who present with atypical pain, no weight loss, and intermittent use of narcotics do not always benefit from surgical repair.[2] Our patient was a poor candidate for surgery: her postprandial symptoms were not consistently present and she had lost no weight. Nevertheless, the absence of these typical characteristics did not exclude the diagnosis of MALS.[13,14]

In cases of suspected MALS, we propose the routine preoperative diagnostic use of IVUS, to clarify which patients might benefit from surgical treatment. The use of IVUS will also help to distinguish patients who have isolated dynamic obstruction from those who have developed a fixed obstruction from chronic irritation by the MAL.

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