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CASE REPORT | SMALL BOWEL

Often Overlooked Diagnosis: Median Arcuate Ligament Syndrome as a Mimicker of Crohn's Disease

Erica Becker, MD, MPH[1], Turab Mohammed, MD[1], and John Wysocki, MD[2]

[1]*Department of Internal Medicine, University of Connecticut Health Center, Farmington, CT*
[2]*Department of Gastroenterology, Hartford Hospital, Hartford, CT*

ABSTRACT

Median arcuate ligament syndrome occurs when the celiac artery and/or the celiac plexus nerves is compressed by the median arcuate ligament during expiration causing a variety of gastrointestinal symptoms. Here, we present a case of median arcuate ligament syndrome in a persistently symptomatic 35-year-old man that presented as a mimicker of Crohn's disease. Symptomatology, computed tomography angiography, and abdominal ultrasound Doppler were consistent with celiac artery compression syndrome. After surgical decompression of the ligament and removal of the celiac ganglion, he reported a definitive relief of abdominal pain and resolution of symptoms.

INTRODUCTION

Median arcuate ligament syndrome (MALS) is a rare clinical condition in which the celiac trunk is compressed by the median arcuate ligament during expiration. MALS often presents as a triad of symptoms: postprandial abdominal pain, bowel function disorder, and weight loss.[1] It is more likely to affect young, thin adult females (F:M ratio of 4:1).[1] The range of patients with celiac compression has been reported between 10% and 24%.[2] However, the overall incidence of symptomatic celiac artery compression syndrome is about 2 per 100,00 patients.[1,2] Although often considered a relatively benign condition, MALS can occasionally mimic life-threatening causes of abdominal pain and present with a variety of nonspecific clinical symptoms, thus calling for an extensive workup in patients with this syndrome.[3,4] In addition, diagnosis can be further delayed because of commonalities of the symptom set. This is a case report of MALS in a patient who does not fit the typical epidemiology and mimicked Crohn's disease (CD) by causing jejunitis.

CASE REPORT

A 35-year-old non-Hispanic white man presented with a 5-year history of nausea with intermittent vomiting, diarrhea with urgency, 90-lbs weight loss, and malnutrition secondary to severe postprandial epigastric pain. Initial workup began several years earlier and before referral to our center when he presented with chronic diarrhea. Endoscopy and colonoscopy with biopsies, stool studies, fecal fat, and abdominal ultrasound were all negative. The patient had video capsule endoscopy by an outside provider that showed scattered ulcers with surrounding exudates in full circumference of the lumen in the middle and proximal jejunum. Lewis Score used to classify the severity of mucosal inflammation was 1,800 in our patient. (A Lewis Score of >790 corresponds to moderate or severe disease). Minimal inflammation was noted in the distal jejunum. He was followed in the gastroenterology outpatient clinic and continued to undergo workup. His complete blood count, comprehensive metabolic panel, thyroid panel, cortisol level, serum amylase, and lipase, serum inflammatory markers, antinuclear antibody, stool calprotectin, pancreatic elastase-1, and prealbumin, routine stool studies, fecal fat, *Helicobacter pylori,* celiac disease panel, and biopsies of the esophagus, stomach, and duodenum were nonrevealing.

Repeat endoscopic evaluations (esophagogastroduodenoscopy/colonoscopy/capsule endoscopy) demonstrated evidence of nonspecific small bowel enteritis with moderate/severe ulceration. He also reported new-onset oral ulcers, and magnetic resonance enterography revealed mild small bowel wall thickening and delayed enhancement of the distal ileum, all suggestive of CD. He was

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Correspondence: Erica Becker, MD, MPH (erica_bckr@yahoo.com).

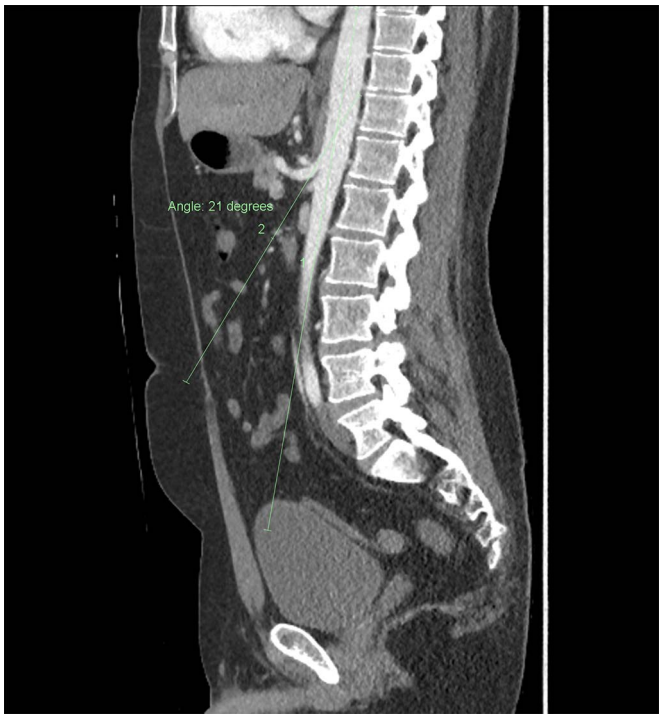

Figure 1. Contrast-enhanced computed tomography showing stenosis and acute angulation at origin of the celiac artery commonly associated with MALS.

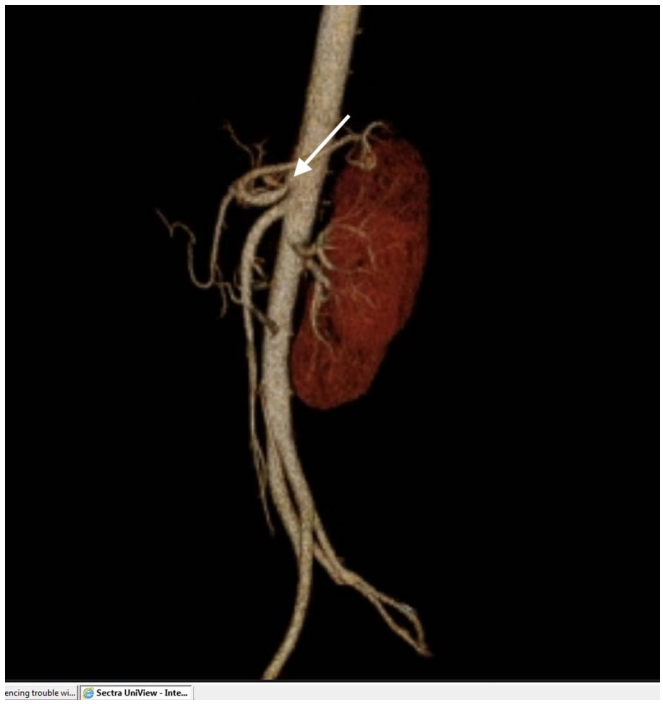

Figure 2. Magnetic resonance angiography 3D reconstruction showing compression of the celiac artery at its origin from abdominal aorta.

initially started on infliximab, but it was stopped about 1 year later because it was not well tolerated. Repeat video capsule endoscopy 1 year later after infliximab was initiated and while he was continued on prednisone showed no evidence of inflammation, yet all the clinical symptoms persisted as before endoscopic evaluation and overall health continued to deteriorate. Importantly, biopsy specimens were not obtained from the area of inflammation to confirm the diagnosis of CD, given the clinical picture and radiographic evidence.

On further workup, a gastric emptying study showed normal emptying. Hepatobillary iminodiacetic acid scan demonstrated biliary dyskinesia for which he underwent cholecystectomy, but his symptoms continued to progress. He required total parenteral nutrition for at least a year because his postprandial epigastric pain and weight loss had become severe. He was also following up in the outpatient pain clinic for the intractable abdominal pain.

Attention was turned to more uncommon causes of abdominal pain. Computed tomography angiography showed a downward trajectory of the celiac axis with narrowing of the origin to approximately 50% (Figure 1). Magnetic resonance angiography 3D reconstruction also showed compression of the celiac artery at its origin from abdominal aorta (Figure 2). Abdominal ultrasound Doppler revealed a patent celiac artery with turbulent, elevated velocities of 348 cm/seconds in the proximal segment with normal expiration (Figure 3A). Waveforms normalized with inspiration and in the upright position (Figure 3B). These findings were consistent with compression of the celiac artery.

The patient underwent diagnostic celiac plexus block and was noted to have a positive response to the injection. He later underwent surgical decompression of the median arcuate ligament and removal of the celiac ganglion. He reported a definitive relief of abdominal pain, nausea, and vomiting. Total parenteral nutrition was stopped. He was gaining weight after procedure and weaning off the pain medications. Six months after intervention, the patient remained off immunosuppressive therapy, and repeat enteroscopy and imaging studies did not reveal any signs of inflammatory lesions.

DISCUSSION

During 1917, Lipshutz first described the compression of the celiac artery, in 1963, Harjola described MALS, and 2 years later, Dunbar et al published the first clinical study describing MALS.[5] It is often a diagnosis of exclusion which can be further delayed in individuals believed to have underlying abdominal disease. A high index of suspicion is needed among patients with persistent symptoms despite adequate treatment of other more common abdominal disease because MALS can present as a mimicker of more common abdominal disease. The diagnosis of MALS can be further complicated because there is a large proportion of patients who have celiac artery compression, but do not present with symptoms. Hence, it is important that other common causes with similar symptoms are evaluated before

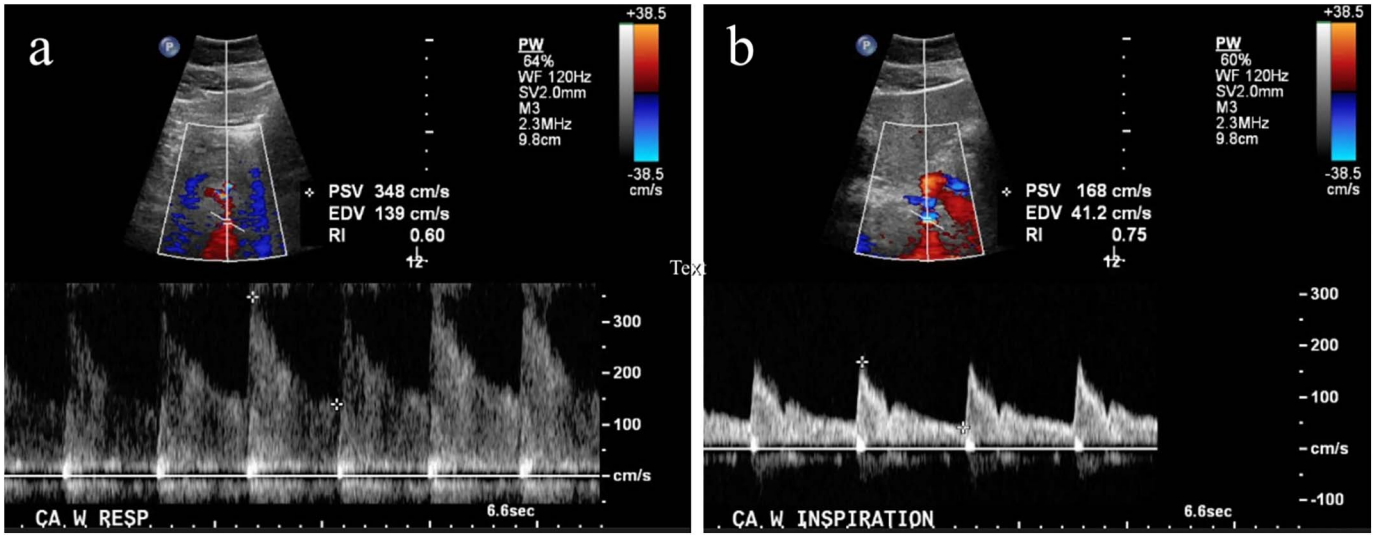

Figure 3. (A) Abdominal ultrasound Doppler showed a patent celiac artery with turbulent, elevated velocities of 348 cm/s in the proximal segment with normal expiration. (B) Waveforms normalized to velocities of 168 cm/s with inspiration and in the upright position.

diagnosing MALS. The diagnosis of MALS significantly relies on reviewing the patient's symptoms, medical history, and correlating imaging findings with patient symptoms and resolution of symptoms with treatment.

Sturiale et al described a case with MALS occurring in a patient with pain refractory to treatment with CD.[1] The case demonstrated that MALS was a diagnosis of exclusion and MALS was only considered when the patient continued to have postprandial abdominal pain after all possible causes including relapse from CD was ruled out. Once MALS was treated, the pain resolved. On the other hand, another case described the diagnosis of MALS in a patient who was later diagnosed with CD after he refused surgical treatment.[6] The symptoms previously believed to be from MALS improved after treatment for CD was started.[6] Our case follows the line similar to the second case described above, but in the opposite direction where we assumed that patients' symptoms were secondary to underlying CD, but instead, it was the MALS that caused the jejunitis and the distressing symptom burden. Also, other authors have described diagnosing MALS in patients with underlying hiatal hernias, gastroesophageal reflux disease, ulcerative colitis, pancreatic cancer, and after pancreaticoduodenectomy.[7–10] In conclusion, among patients with MALS and suspected concurrent abdominal disease, it can be very challenging and difficult to point out the actual cause of patient symptoms. To the best of our knowledge, there are no reports of MALS causing jejunitis.

It is crucial that once MALS in considered, patients undergo relevant diagnostic evaluation. At this time, there is not a consensus to workup; however, mesenteric duplex ultrasound, angiography, and gastric exercise tonometry are all available options.[11] A diagnosis of MALS is demonstrated when mesenteric duplex ultrasound shows increased blood flow velocities of greater than 200 cm/s in the celiac artery and end diastolic velocity greater than 55 cm/s.[12] With deep inspiration, there should be a decreased or normalization of velocities.[12] Angiogram can help to detect the change in shape of the celiac artery during both inspiration and expiratory phases.[12]

Previous studies have demonstrated that certain patient characteristics such as abdominal pain after eating, patients between 40 and 60 years of age, and weight loss of 20 pounds or more indicate a better prognosis after surgical intervention and these characteristics should help determine who needs surgery.[12] Both open surgery and laparoscopic median arcuate ligament release are safe options for decompression, and advantages/disadvantages of each should be considered on a case-to-case basis.[5] During decompression of the artery, the celiac nerves may be simultaneously removed. Short- to medium-term follow-up has shown improvement in symptoms after surgical intervention.[11] Other options for treatment include celiac artery bypass (ie, aortoceliac or aorta-hepatic bypass) and superior mesenteric artery or splenic artery transposition.[2] Second-line treatment would include balloon angioplasty when surgical options are unavailable or unsuccessful.[2]

MALS is an often-overlooked diagnosis, and unfortunately, most patients have had protracted workup and sometimes even surgical procedures before the diagnosis of MALS is even considered. Furthermore, clinicians should consider MALS even in patients diagnosed with other abdominal disease especially among those who do not show adequate response to appropriate treatment as MALS could be mimicking a more common abdominal disease.

DISCLOSURES

Author contributions: E. Becker wrote the manuscript. T. Mohammed and J. Wysocki revised the manuscript for intellectual content. E. Becker, T. Mohammed, and J. Wysocki

approved the final manuscript. J. Wysocki is the article guarantor.

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Informed consent was obtained for this case report.

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