CLINICAL VIGNETTE

Median Arcuate Ligament Syndrome – An Uncommon Cause of Abdominal Pain

Mindy Goh, DO and Pinting Chen, MD

Case

A 44-year-old female presented to her primary care physician's office with generalized abdominal pain and swelling in her abdomen and legs. Her prior medical history includes hypertension, rheumatoid arthritis, Raynaud's and prior cholecystectomy and appendectomy. CT of the abdomen and pelvis found a 17 mm nodular density in the greater curvature of gastric antrum. Both abdominal ultrasound and lower extremity ultrasounds were unremarkable, and a pelvic ultrasound noted an 18 mm subserosal fundal fibroid with normal ovaries. She was given furosemide and her abdominal and leg swelling improved after a few doses. However, over the next two weeks she developed worsening epigastric postprandial abdominal pain with inability to tolerate oral intake other than plain crackers. Her pain was associated with occasional nausea, nonbloody and nonbilious vomiting, and a 6-pound weight loss. She also reported increasing acid reflux over the last 6 months with some benefit from daily pantoprazole 40mg. She did not drink alcohol. Labs including CBC, CMP and lipase were unremarkable. Upper endoscopy noted LA Grade A esophagitis, patchy gastropathy, and several small gastric body polyps. Her pain continued to worsen and she lost about 20 pounds with inability to tolerate oral intake. MRA showed narrowing of the celiac artery with a J-shaped appearance at the level of the median arcuate ligament with post stenotic dilatation, most compatible with median arcuate ligament syndrome. (Figure 1, sagittal plane. Figure 2, coronal plane). Given continued inability to tolerate oral intake, a PICC was placed for TPN (total parenteral nutrition). She underwent a laparoscopy with median arcuate ligament release and celiac plexus neurolysis. By post-op day 3, she was able to tolerate PO intake and was discharged and weaned off TPN.

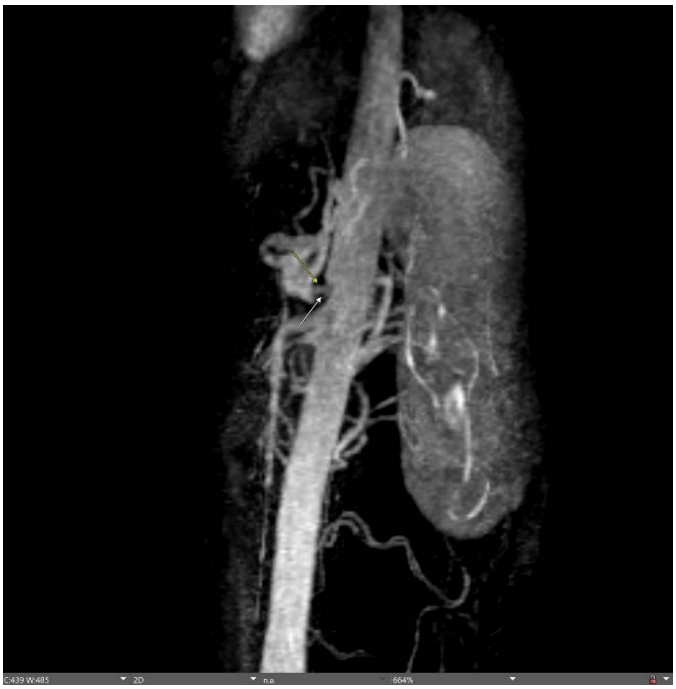

Figure 1. Sagittal plane.

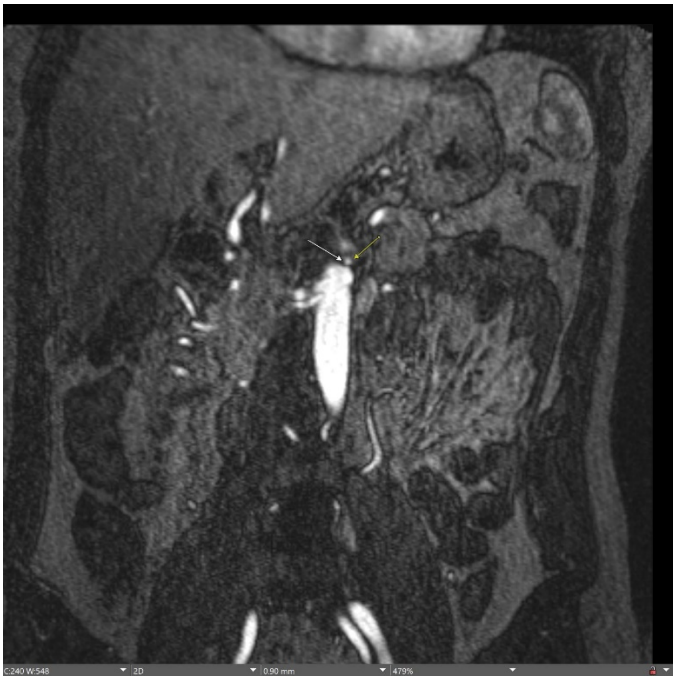

Figure 2. Coronal plane.

Discussion

Median arcuate ligament syndrome (MALS) is chronic abdominal pain related to compression of the celiac artery from the median arcuate ligament of the diaphragm.[1] It is more common in women, with a higher incidence between the third and fifth decades.[1] The pathophysiology is controversial and poorly understood, but thought to be related to vascular and neuropathic pathways.[2] While the celiac artery typically branches off the aorta under the median arcuate ligament, this can vary widely. Higher or lower origins can result in compression, compromising blood flow to the abdominal organs. Variation in anatomy does not always lead to compromise in blood flow due to superior and inferior mesenteric arteries collaterally. It is also hypothesized that the median arcuate ligament may also compress the celiac plexus, which is often adjacent to the celiac artery origin, causing neuropathic pain.[2]

Patients with MALS may present with variable symptoms including postprandial abdominal pain, bloating, diarrhea, nausea, vomiting, and unintentional weight loss.[2-4] The incidence of MALS is approximately 0.02%, and its rarity often makes it a diagnosis of exclusion.[1] It should be considered in the differential diagnosis of unexplained abdominal pain and weight loss. An extensive gastrointestinal evaluation is typically performed to rule out more common causes of a patient's symptoms. These include esophagogastroduodenoscopy, colonoscopy, motility studies, imaging studies, and laboratory testing prior to arriving at this diagnosis, as seen in our patient. Patients with MALS often have unremarkable EGDs, colonoscopies, abdominal ultrasounds, but have celiac artery narrowing on cross sectional vascular imaging such as duplex ultrasound, or CT or MR angiography.[3]

The mainstay of treatment of MALS is surgical with laparoscopic release of the median arcuate ligament. Conservative measures such as dietary modifications and pain control can help with symptoms until definitive surgery.[1,4]

Laparoscopic release of the median arcuate ligament alleviates external compression on the celiac artery, restoring normal blood flow to the abdominal organs.[2] Dissection of the celiac plexus is also often performed given evidence of the neuropathic contribution to symptoms.[3]

Conclusion

Median arcuate ligament syndrome, although rare, should be considered in the differential diagnosis when evaluating patients with symptoms similar to mesenteric ischemia. Diagnosis is established with vascular imaging that demonstrates compression of the celiac artery by the MAL. Definitive treatment involves surgical intervention to release the compression.

REFERENCES

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