

CLINICAL IMAGING

An easily overlooked cause of abdominal pain

Kyle Bennett, DO*, Andrew Rettew, DO, Bilal Shaikh, DO,
Suzanne Supplee, DO and Richard Alweis, MD FACP

Department of Internal Medicine, Reading Health System, West Reading, PA, USA

A 63 year old female presented to the emergency department with a several month history of intermittent right upper quadrant abdominal pain, early satiety with loss of appetite, and an unintentional weight loss of 8. She underwent a battery of tests that returned negative and subsequently sent for Computed tomographic angiography (CTA) of the abdomen revealing luminal stenosis of the proximal 1 cm of the celiac axis estimating 90% occlusion and a patent SMA and IMA with, typical for median arcuate ligament syndrome. The symptoms, diagnosis, and treatment options are discussed.

Keywords: *Median arcuate ligament syndrome (MALS); postprandial pain; unintentional weight loss*

*Correspondence to: Kyle Bennett, Department of Internal Medicine, Reading Health System, 6th Avenue, Spruce Street, West Reading, PA 19611, USA, Email: kyle.bennetto@readinghealth.org

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A 63-year-old female with a history of irritable bowel syndrome, anxiety, and depression presented to the emergency department with a several month history of intermittent right upper quadrant abdominal pain, early satiety with loss of appetite, and an unintentional weight loss of 8 pounds. The pain was described as sharp, worse during and after meals, and progressively worsening. She denied symptoms of nausea, vomiting, changes in bowel habits, or hematochezia. She was admitted and had an esophagogastroduodenoscopy for these symptoms the following morning, which showed mild gastric and esophageal inflammation. Celiac serology and *Helicobacter pylori* studies returned negative. Diagnostic evaluation by right upper quadrant ultrasound, EKG, electrolytes, liver function tests, amylase, lipase, and complete blood counts were also unremarkable. Computed tomography of the abdomen and pelvis only identified mild edema of the duodenal bulb, possibly representing duodenitis. She was discharged on lansoprazole and dicyclomine daily for the following month and asked to follow-up with her primary care physician in the outpatient setting. She returned to the outpatient clinic with continued postprandial pain, weight loss, and subjective worsening with an unremarkable abdominal examination. With no improvement, she underwent a gastric emptying study that was normal. Doppler ultrasound was performed with inconclusive results. The patient was subsequently sent for computed tomographic angiography (CTA) of the abdomen revealing luminal stenosis of the proximal 1 cm of the celiac axis estimating 90% occlusion, typical for median arcuate ligament syndrome (MALS),

and a patent SMA and IMA (Fig. 1). The patient was referred to surgery, but ultimately opted to not have the procedure performed based on the risks of surgery outweighing the benefits.

MALS is characterized by classic symptoms including postprandial epigastric crampy pain, nausea, vomiting, bloating, and weight loss which can mimic mesenteric ischemia. The etiology of MALS is incompletely understood and likely multifactorial. However, it is widely accepted that it starts anatomically when the median arcuate ligament and the origin of the celiac artery are abnormally close in proximity. This, in turn, can result in a combination of ischemic changes from external compression of the celiac artery and damage to the celiac nerve plexus resulting in neuropathic pain causing symptoms of the disease (1, 2). Typically, patients are young, thin females aged between 40 and 60 presenting with the aforementioned symptoms prevalent for a duration of 3 months-10 years (3, 4). Doppler ultrasound is an effective screening test and detailed imaging techniques such as CTA, MRA, and direct catheter angiography are considered the 'gold standard' for diagnosis. Management is aimed at restoring normal blood flow in the celiac axis and eliminating neural irritation produced by the celiac ganglion fibers (3). Laparoscopic and robotic surgical median arcuate ligament release has replaced open surgical management as the mainstay of treatment. The procedure includes division of the restrictive fibrous ligament with or without celiac ganglionectomy (4). If persistent celiac stenosis remains after surgical intervention, it is recommended the patient undergo endovascular approaches,

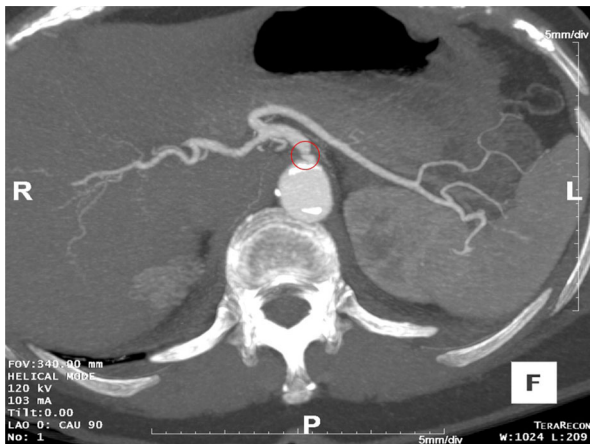


Fig. 1. Computed tomographic angiography (CTA) of the abdomen revealing luminal stenosis of the proximal 1 cm of the celiac axis estimating 90% occlusion, typical for median arcuate ligament syndrome, and a patent SMA and IMA.

including angioplasty and stenting (2, 5). In summary, MALS can be difficult to diagnose based on vague and non-specific symptoms and the disease should remain on the differential after extensive work-up of chronic intestinal ischemia returns negative.

References

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