Case Report

Dunbar syndrome: An unusual cause of chronic postprandial abdominal pain

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Abstract

Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome, is a rare condition in which the celiac artery is compressed by the median arcuate ligament of the diaphragm. We hereby report a case of a 48-year-old female presenting with long-standing abdominal pain and ninety-pound weight loss who was found to have median arcuate ligament syndrome after extensive workup.

Introduction

MALS classically presents with a triad of postprandial pain, weight loss and bruit in the epigastrium [1]. The pathophysiology of abdominal pain in cases of MALS is not entirely understood, though presumed to be ischemic. Ligamentous compression of the celiac trunk can be asymptomatic in around 10%-20% of patients, with radiographic evidence of compression, yet with no symptoms [2]. Some degree of neural compression of the celiac plexus is postulated to be also a part of the pathophysiology of abdominal pain in MALS [3].

Case presentation

We present the case of a 48-year-old female initially presented to the emergency room with chronic abdominal pain and an unintentional 30-pound weight loss for five months. The pain was described as postprandial, localized at the epigastrium, burning in nature, with a pain score of 10/10 and associated with sitophobia. The patient denied any vomiting, diarrhea, or constipation. Esophagogastroduodenoscopy showed patchy gastropathy—biopsy was negative for malignancy and positive for Helicobacter pylori. She was treated with a course of triple therapy and underwent colonoscopy with normal findings. She returned to our clinic the following year with similar symptoms and had lost a total of 90 pounds. Further investigations including tissue transglutaminase antibodies, HIV, and TB were negative. A repeat esophagogastroduodenoscopy showed small hiatal hernia-repeat biopsy was negative for recurrence of Helicobacter pylori infection. Further investigation was performed for assessment of the weight loss with fluoroscopic upper gastrointestinal series with small bowel follow through showing heterogenous fundus with nodularity with no small bowel abnormalities. CT angiography (CTA) showed a severe narrowing of the trunk of

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Fig. 1 – Axial abdominal CTA image demonstrates narrowing of the proximal celiac axis (red arrow). There is minimal post stenotic dilatation, characteristic of median arcuate ligament syndrome. (Color version available online.)

Fig. 2 – Sagittal image of a CTA of the abdomen demonstrates acute angulation and narrowing of the proximal celiac axis. There is minimal post stenotic dilatation, which overall creates a “hooked” appearance (red arrow) that is characteristic of median arcuate ligament syndrome.

celiac artery with poststenotic dilatation secondary to compression by the crus of the diaphragm (Figs. 1 and 2) that in view of the clinical scenario lead to the diagnosis of MALS. The patient was offered arteriography and possible decompression of celiac artery but declined the intervention. Her symptoms continue to be managed with repeated visits to the emergency department for abdominal pain.

Discussion

The median arcuate ligament (MAL) is a fibrous arch that connects the diaphragmatic crura to form the anterior margin of the aortic hiatus [4]. The location of the MAL is exceedingly variable [5] and it may indent upon and cause downward angulation of the celiac trunk, which can be a nonobstructive anatomic variant or result in mesenteric ischemia. Dunbar et al. linked the anatomic anomaly with clinical symptoms of intestinal angina in 1965 [6], hence linking the disease to his name.

MALS is a rare [7] and often difficult diagnosis in view of nonspecific presenting symptoms such as postprandial abdominal pain, nausea, vomiting, and weight loss.

The exact pathophysiology of the disease is not fully understood, primarily attributed to external compression of the celiac artery by an abnormally low-lying median arcuate ligament. The compression worsens with expiration as the diaphragm moves caudally worsening the compression of the celiac trunk. This leads to visceral ischemia and postprandial abdominal pain. Chronic abdominal pain is also postulated to occur from overstimulation of the celiac ganglion. Sustained compression of the celiac axis may lead to changes in vascular layers such as intimal hyperplasia, proliferation of elastic fibers in the media, and disorganization of the adventitia [8].

Physical examination may reveal an abdominal bruit. Patients often undergo a battery of gastrointestinal evaluation including endoscopy/colonoscopy, motility studies, and abdominal imaging before the diagnosis of MALS is considered [9].

MALS is diagnosed with CTA which demonstrates a characteristic focal narrowing of the proximal celiac axis with a “hooked” appearance caused by the inferior displacement of the celiac artery by the MAL, most optimally noted on sagittal views [10]. Because the MAL is attached to the diaphragmatic crura, the position of the MAL and subsequently the degree of compression of the celiac axis changes during different phases of respiration. Imaging is best acquired during the end-inspiratory phase, where true compression can be identified, since cranial displacement of the diaphragm during end-expiration can cause transient narrowing of the celiac axis, with a false positive impression [11]. Ancillary findings such as poststenotic dilatation and collateral formation may be present, and aids in diagnosis. CTA may also identify concomitant vascular abnormalities such as anatomic abnormalities or mesenteric thrombosis/atherosclerosis. Duplex ultrasound showing a peak systolic flow greater than 200cm/s can be used as screening for MALS, however may not clearly identify the cause as MALS compression [10]. MALS is managed by celiac decompression via release of median arcuate ligament guided by intraoperative duplex ultrasound, which can be done laparoscopically [12]. If the celiac artery flow abnormality on Doppler ultrasound persists after MAL release, angioplasty and stenting can be proposed. [13]
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