Radiology Case Reports 15 (2020) 1747–1749

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Dunbar syndrome: An unusual cause of chronic postprandial abdominal pain

Jonathan Li, Xin Yee Tan, Ayah Megahed[1,], Angela Evangelista*

Department of Internal Medicine, NYC Health + Hospitals/Lincoln, 234 East 149 St, Bronx, NY 10451, USA

ARTICLE INFO

Article history:
Received 8 May 2020
Revised 28 June 2020
Accepted 4 July 2020

Keywords:
Coeliac axis
Weight loss
CT angiography

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

* Corresponding author.
E-mail address: ayahmugahid@gmail.com (A. Megahed).
[1] Present corresponding author address: 1080 New Haven Avenue, Apt 95, Milford, CT, 06460.

https://doi.org/10.1016/j.radcr.2020.07.016

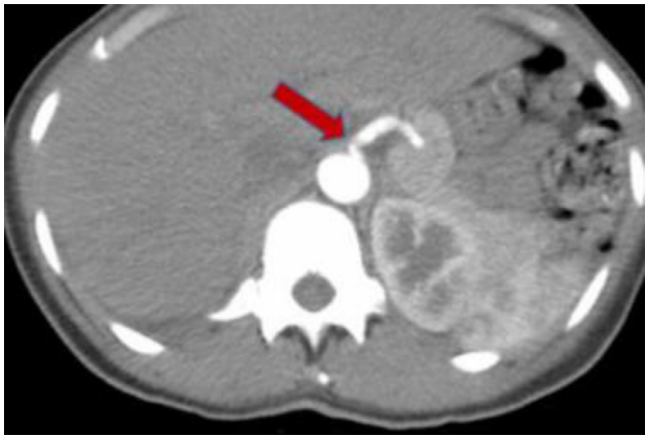

Fig. 1 – Axial abdominal CTA image demonstrates narrowing of the proximal celiac axis (red arrow). There is minimal post stenotic dilatation, characteristic of median arcuate ligament syndrome. (Color version available online.)

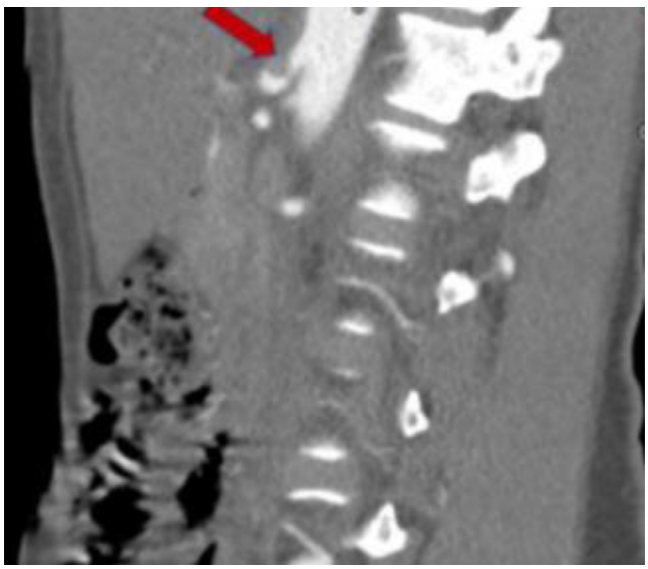

Fig. 2 – Sagittal image of a CTA of the abdomen demonstrates acute angulation and narrowing of the proximal celiac axis. There is minimal post stenotic dilatation, which overall creates a "hooked" appearance (red arrow) that is characteristic of median arcuate ligament syndrome.

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 MAL 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]

REFERENCES

[1] Harjola PT. A rare obstruction of the coeliac artery: report of a case. Ann Chir Gynaecol Fenn 1963;52:547–50.
[2] Sturiale A, Alemanno G, Giudici F, Addasi R, Bellucci F, Tonelli F. Median arcuate ligament syndrome in a patient with Crohn's disease. Int J Surg Case Rep 2013;4:399–402.
[3] Brandt LJ, Boley SJ. Celiac axis compression syndrome. Am J Dig Dis 1978;23:633–64.
[4] Lambda R, Tanner DT, Sekhon S, McGahan JP, Corwin MT, Lall CG. Multidetector CT of vascular compression syndromes in the abdomen and pelvis. Radiographics 2014;34:93–115.
[5] Levin DC, Baltaxe HA. High incidence of celiac axis narrowing in asymptomatic individuals. Am J Roentgenol Radium Ther Nucl Med 1972;116:426–9.
[6] Dunbar JD, Molnar W, Beman FF, Marable SA. Compression of the celiac trunk and abdominal angina. Am J Roentgenol Radium Ther Nuc Med 1965;95:731–44.
[7] You JS, Cooper, Nishida M, Matsuda S, Murariu D E. Treatment of median arcuate ligament syndrome via traditional and robotic techniques. Hawaii J Med Public Health 2013;72:279–81.
[8] A-Cienfuegos J, Rotellar F, Valentí V, et al. The celiac axis compression syndrome (CACS): critical review in the laparoscopic era. Rev Esp Enferm Dig 2010;102(3):193–201.
[9] Kyle Bennett DO, Andrew Rettew DO, Bilal Shaikh DO, Suzanne Supplee DO, Alweis Richard. An easily overlooked cause of abdominal pain. J Community Hosp Intern Med Perspect 2014;4(5):25083.
[10] Horton KM1, Talamini MA, Fishman EK. Median arcuate ligament syndrome: evaluation with CT angiography. Radiographics 2005;25(5):1177–82.
[11] Lee VS, Morgan JN, Tan AG, Pandharipande PV, Krinsky GA, Barker JA, et al. Celiac artery compression by the median arcuate ligament: a pitfall of end-expiratory MR imaging. Radiology 2003;228:437–42.
[12] Roayaie S, Jossart G, Gitlitz D, Lamparello P, Hollier L, Gagner M. Laparoscopic release of celiac artery compression syndrome facilitated by laparoscopic ultrasound scanning to confirm restoration of flow. J Vasc Surg 2000;32:814–17.
[13] Duffy AJ, Panait L, Eisenberg D, Bell RL, Roberts KE, Sumpio B. Management of median arcuate ligament syndrome: a new paradigm. Ann Vasc Surg 2009;23:778–84.