

Celiac Artery Compression Syndrome

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Abstract

Celiac artery compression syndrome, Dunbar syndrome, or median arcuate ligament syndrome. They are different names for one syndrome. It is a rare syndrome that can present with recurrent abdominal pain. The condition results from the compression of the celiac artery by the median arcuate ligament which is a fibrous band of the diaphragm [1]. The etiology of celiac artery compression syndrome is still not very clear. Not every patient with this anatomic compression develops symptoms.

Keywords: Celiac Artery; Median Arcuate Ligament; Mesenteric Ischemia

Introduction

The median arcuate ligament is a fibrous band that connects the two medial borders of the diaphragmatic crura near the level of the 12th thoracic. The level of Insertion of the diaphragm is influenced by some congenital factors or the origin of the celiac artery. This is supported by a small series of studies in families. This type of compression is believed to cause chronic intermittent mesenteric ischemia [2,26].

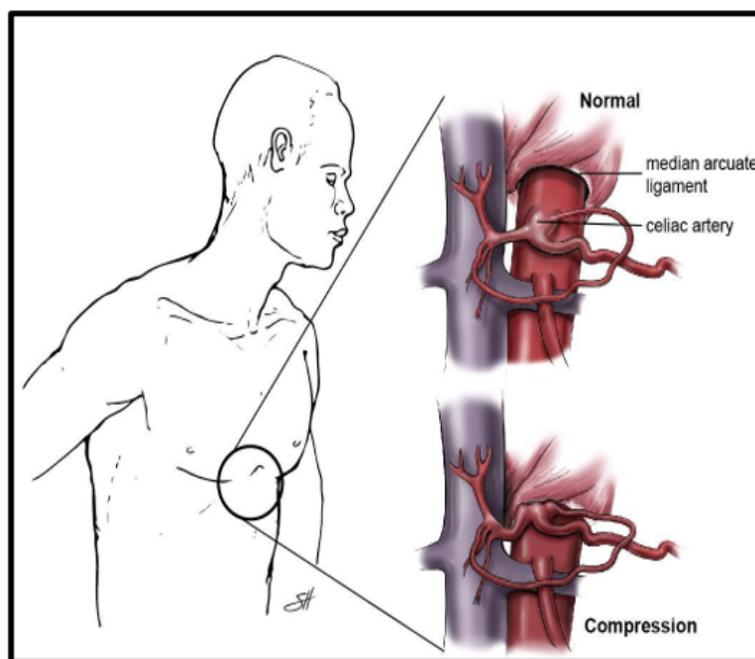


Figure 1: Illustration by Scott Holmes, CMI, Michael E. DeBakey Department of Surgery.

Signs and symptom

Presentation is classically complaining of postprandial, dull, crampy epigastric pain within 1 - 2 hours after eating. Especially large fatty meals. The pain can be of variable intensity and location. Symptoms are often progressive and may present with acute mesenteric ischemia. The diagnosis of chronic mesenteric ischemia is usually made in patients with unexplained chronic abdominal pain, weight loss, and decreased appetite [5].

Investigations

The diagnosis of Chronic mesenteric Ischemia (CMI) is based on:

- The triad of clinical symptoms; postprandial epigastric pain, weight loss, and abdominal bruit.
- Radiological evaluation of the mesenteric vasculature,
- Functional assessment of mucosal ischemia.

Celiac artery compression syndrome is a diagnosis of exclusion. Complete workup of this condition may include simple tests to rule out other etiologies. These can include:

- Colonoscopy,
- Liver, pancreas and gallbladder ultrasound,
- Upper gastrointestinal endoscopy,
- Conventional visceral angiography,
- Doppler ultrasound,
- Magnetic resonance imaging, and
- Computerized tomography angiography (CTA) [12,22]
- Lab workup also likes; CBC, LFTs, serum amylase and lipase, CRP, and testing of certain antibodies such as the anti-smooth muscle antibody. Several diagnostic modalities can be categorized as non-invasive and invasive modalities.

Ultrasound shows an elevated celiac artery peak systolic velocity with deep expiration greater than 200 cm/s and a deflection angle greater than 50 degrees. Ultrasound examination is an important method in diagnosing visceral ischemia of atherosclerotic origin, as well as constriction related to the compression of the celiac trunk. In the case of compression of the celiac trunk by the median arcuate ligament of the diaphragm, a needed regular examination procedure is to normalize the flow velocity, i.e. reduce the peak systolic velocity > 200 cm/s, and end-diastolic velocity (EDV) >55 cm/s during inspiration. Additionally, the following findings may also be found: the abnormal origin of the celiac artery, flow reversal in the hepatic artery, and lowering of velocity in the celiac artery when the patient stands upright [8,23].

Conventional visceral angiography shows partial to complete stenosis of the celiac artery with possible post-stenotic dilation and retrograde filling of the celiac artery. CTA shows compression of the celiac axis with focal stenosis and post-stenotic dilation also. The difference in perfusion of the celiac artery during inspiration and expiration is one of the characteristic findings in this syndrome [3,24].

Abdominopelvic vascular compression syndromes characterized by either:

External compression of vessels by adjacent anatomical structures (i.e., median arcuate ligament syndrome, nutcracker syndrome) or Compression of the viscera by adjacent vessels (superior mesenteric artery syndrome, obstruction of ureteropelvic junction, ureteral vascular compression, portal biliopathy).

These syndromes can be accidentally discovered in asymptomatic patients on imaging examinations performed for other indications.

Although computed tomography (CT) is an accurate noninvasive technique for their detection, the diagnosis remains challenging due to the different non-specific clinical presentation and often missed imaging features. Dynamic imaging may be performed in order to evaluate patients with inconstant symptoms manifesting in a specific position. The purposes are to review the CT imaging findings, correlating with anatomical variants [7].

Treatment

Revascularization indications include abdominal pain and weight loss to prevent future bowel infarction. Surgical techniques can be open surgery or endovascular revascularization.

The endovascular approach consists of angioplasty with or without stenting with a higher rate of restenosis. However open revascularization has a longer recovery period. Patients with atherosclerosis should receive treatment to avoid the risk of cardiovascular events, treatment of hypercholesterolemia, hypertension, diabetes, antiplatelet medications, and smoking cessation counsel. Routine follow up is mandatory to identify restenosis. Out of 161 patients with endovascular angioplasty, 27 developed restenosis and required open repair had significantly high rates of aortic occlusive disease. Open surgery includes aortomesenteric and/or celiac bypass grafting, endarterectomy, and mesenteric reimplantation. The choice of procedure depends primarily on patient anatomy. The most common open surgical procedure is antegrade aorto-mesenteric/ aorto-celiac bypass [4].

Laparoscopic or open surgical decompression are the only treatment options in Median arcuate ligament syndrome MALS [6,25]. A common procedure involves the separation of the ligament fibers and other surrounding tissues around the beginning of the celiac trunk. This can be done by either laparotomy or laparoscopic surgery. The prognosis is good, with a cure rate of approximately 80%. Laparoscopic and open ligament release, celiac artery revascularization, and celiac ganglionectomy, may provide symptom relief in most of the patients diagnosed with MALS [10,11].

Stenotic Celiac trunk resection and the continuity of the vascular axis establish using a splenic artery patch. The postoperative course usually with rapid recovery and patients presents with adequate hepatic blood flow on the first day postoperatively [9].

Case Report

A 68-year-old Caucasian woman, with a past medical history of hypertension, atherosclerosis of the aorta, and hyperlipidemia, presented to the clinic with a one-year history of intermittent right and left lower quadrant abdominal pain. She had associated change in bowel habits (alternating constipation and diarrhea), weight loss, depressed mood, and joint pain. No aggravating or relieving factors were mentioned. Her physical examination, vital signs, and blood works were noted to be unremarkable.

A CT Abdomen and Pelvis with oral and IV contrast was ordered, and the scan showed a long segment of abnormal wall thickening, with minimal fat stranding affecting the left hemicolon. These findings were consistent with colitis of the left colon, and the patient was referred to a gastroenterologist for further evaluation and appropriate treatment.

Two months after treatment, the patient was represented with a history of right-sided epigastric pain. The pain was not related to her diet, and she denied accompanying symptoms of weight loss, diarrhea, fever, melena, or hematochezia. A CT angiogram was ordered which showed compression of her celiac artery by the arcuate ligament. Of note, her superior and inferior mesenteric arteries were patent. The gastroenterologist was consulted and suggested that the patient be referred to a vascular surgeon, to ensure that there was no vascular compromise to her colon.

The vascular surgeon discovered that the patient had scar tissue compressing her celiac artery, likely accounting for her symptoms. The blood flow to her colon was intact, and thus surgical intervention was not warranted. The patient was managed conservatively with lifestyle modifications, and her condition improved.

Discussion

Celiac Artery Compression Syndrome is a rare disorder that was first observed in 1917 by Benjamin Lipchitz [13]. Pekka-Tapani Harjola and Samuel Marable also described this syndrome in 1963 and 1965 respectively [14,15]. Hence, Celiac Artery Syndrome is also known as Harjola-Marable syndrome and Marable syndrome.

The prevalence of Celiac Artery Compression Syndrome is estimated to be between 10-24%. It is more common in woman who is between the ages of 20- 40 yrs old, with the female to male ratio being 2-3: 1 [16]. This disorder occurs as a result of abnormal migration of cells during embryogenesis, resulting in the median arcuate ligament crossing anterior to the celiac artery. If this happened, the compression of the celiac artery by the median arcuate ligament chance increases.

Most patients with this syndrome are asymptomatic. However, when individuals are symptomatic, they typically present with epigastric pain made worse by eating food, anorexia, bloating, vomiting, weight loss, and exercise intolerance. Additionally, an abdominal bruit may be heard on physical examination [17]. The patient in the above case admitted to only experiencing epigastric pain. Given her age and absence of additional supporting symptoms, it is fair to say this patient had an atypical presentation of Celiac Artery Compression Syndrome. Patient was treated conservatively and showed improvement.

Individuals who have evidence of a vascular compromise to their bowel, or those who have failed conservative treatment can be managed surgically. The surgical management involves dividing the median arcuate ligament, thus decompressing the celiac artery. The surgery can be open or laparoscopic surgery [16,19].

Juan Carlos Jimenez, Michael Harlander-Locke, and Erik P Dutson conducted a retrospective study that was published on June 29, 2012, looking at open versus laparoscopic surgical treatment of Celiac Artery Compression Syndrome. There were 400 patients included in the study, and they found that 85% of patients reported immediate postoperative relief of their symptoms (laparoscopic and open surgery). Furthermore, 5.7% of the patients who had laparoscopic surgery, compared to 6.8% of the patients who had open surgery experienced "late recurrence of their symptoms" [21].

Conclusion

Celiac Artery Compression Syndrome is a clinically rare disorder caused by compression of the celiac artery by the median arcuate ligament. Symptoms of this disorder include epigastric pain made worse by eating, nausea, exercise intolerance, bloating, weight loss, and vomiting. Most patients with this disorder are asymptomatic. Individuals who develop symptoms can be managed conservatively or with surgery.

Bibliography

1. Zambrano-Lara M., *et al.* "Median arcuate ligament syndrome as a rare cause of chronic abdominal pain". *Revista de Gastroenterología de México* (2020): 14.
2. Matsuura H., *et al.* "Intermittent Severe Epigastric Pain and Abdominal Bruit Varying With Respiration". *Gastroenterology* 158.4 (2020): e11-e12.
3. Römer C., *et al.* "Assessment of celiac artery compression using color-coded duplex sonography". *Clinical Hemorheology and Microcirculation* (2020).
4. Acosta S. "Epidemiology of mesenteric vascular disease: clinical implications". *Seminar Vascular Surgery* 23 (2010): 4.
5. van Dijk LJ., *et al.* "Clinical management of chronic mesenteric ischemia". *United European Gastroenterol Journal* 7.2 (2019): 179-188.
6. Rubinkiewicz M., *et al.* "Laparoscopic decompression as treatment for median arcuate ligament syndrome". *Annals of the Royal College of Surgeons of England* 97.6 (2015): e96-99.
7. Gozzo C., *et al.* "CT imaging findings of abdominopelvic vascular compression syndromes: what the radiologist needs to know". *Insights Imaging* 11.1 (2020): 48.
8. Elwertowski M and Lechowicz R. "Standards of the Polish Ultrasound Society - update. Ultrasound examination of the visceral arteries". *Journal of Ultrasonography* 15.60 (2015): 85-95.
9. Bacalbasa N., *et al.* "Celiac Trunk Stenosis Treated by Resection and Splenic Patch Reconstruction - A Case Report and Literature Review". *In Vivo* 32.3 (2018): 699-702.
10. Sun Z., *et al.* "Laparoscopic treatment of median arcuate ligament syndrome". *Intractable and Rare Diseases Research* 8.2 (2019): 108-112.
11. Jimenez JC., *et al.* "Open and laparoscopic treatment of median arcuate ligament syndrome". *Journal of Vascular Surgery* 56.3 (2012): 869-873.
12. M Rubinkiewicz., *et al.* "Laparoscopic decompression as treatment for median arcuate ligament syndrome". *Annals of the Royal College of Surgeons of England* 97.6 (2015): E96-e99.
13. Harjola PT. "A rare obstruction of the coeliac artery. Report of a case". *Annales Chirurgiae et Gynaecologiae Fenniae* 52 (1963): 547-550.
14. Dunbar JD., *et al.* "Compression of the celiac trunk and abdominal angina". *American Journal of Roentgenology Radium Therapy and Nuclear Medicine* 95.3 (1965): 731-744.
15. Duran M., *et al.* "Open vascular treatment of median arcuate ligament syndrome". *BMC Surgery* 17 (2017): 95.
16. Duffy AJ., *et al.* "Management of median arcuate ligament syndrome: a new paradigm". *Annals of Vascular Surgery* 23.6 (2009): 778-784.
17. Rubinkiewicz M., *et al.* "Laparoscopic decompression as treatment for median arcuate ligament syndrome". *Annals of the Royal College of Surgeons of England* 97 (2015): 96-99.
18. Duncan AA. "Median arcuate ligament syndrome". *Current Treatment Options in Cardiovascular Medicine* 10.2 (2008): 112-116.
19. Lainez RA and Richardson WS. "Median arcuate ligament syndrome: a case report". *Ochsner Journal* 13 (2013): 561-564.

20. Juan Carlos Jimenez., *et al.* "Open and laparoscopic treatment of median arcuate ligament syndrome". (2012).
21. Manghat NE., *et al.* "The median arcuate ligament syndrome revisited by CT angiography and the use of ECG gating—a single-center case series and literature review". *British Journal of Radiology* 81.969 (2008): 735-742.
22. P T Harjola. "Buffer capacity and acid-base balance of blood effects of blood loss and some infusion fluids on rabbit blood". *Annales chirurgiae et gynaecologiae Fenniae*. Supplementum, Helsinki (1960): 95.
23. P T Harjola. "A rare obstruction of the coeliac artery". *Annales chirurgiae et gynaecologiae Fenniae* 52 (1963): 547-549.
24. J D Dunbar., *et al.* "Compression of the celiac trunk and abdominal angina". *American Journal of Roentgenology* 95 (1965): 731-743.
25. S A Marable., *et al.* "Abdominal pain secondary to celiac axis compression". *The American Journal of Surgery*, New York, April 111.4 (1966): 493-495.
26. S A Marable., *et al.* "Celiac compression syndrome". *American Journal of Surgery* 115 (1968): 97-102.

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