

CASE REPORT

Median Arcuate Ligament Syndrome: A Rare Cause of Epigastric Pain

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ABSTRACT

Aim: To diagnose and aid the treatment of a patient with an unknown cause of epigastric pain.

Background: Harjola first described median arcuate ligament syndrome (aka celiac artery compression syndrome) in 1963. Typically it occurs in young patients (20–40 years age group) and is more common in lean women, presents with epigastric pain and weight loss.

Case description: An 18-year-old male visited the emergency room with a complaint of recurrent episodes of abdominal pain. There was no past or any medical history of any previous illness. On physical examination, the patient was afebrile and had pallor. All the vitals were within normal limits. The abdominal examination did not reveal any significant abnormality. Laboratory investigations (Hb, GBP, TLC) were within normal limits.

Conclusion: Median arcuate ligament syndrome is a rare clinical entity which can be found in normal asymptomatic people. In a few cases, however, celiac axis compression can cause symptoms which can be relieved with laparoscopic surgical decompression.

Clinical significance: The median arcuate ligament syndrome as an entity must be kept in mind when no other cause of abdominal pain is zeroed on even after all the clinical examinations and investigations.

Keywords: Celiac artery compression syndrome, Epigastric pain, Median arcuate ligament syndrome, Radiology.

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BACKGROUND

Harjola first described median arcuate ligament syndrome (aka celiac artery compression syndrome) in 1963. Typically it occurs in young patients (20–40 years age group) and is more common in lean women, presents with epigastric pain and weight loss. A fibrous arch uniting

the diaphragmatic crura on either side of the aortic hiatus called Median arcuate ligament, compresses celiac artery compromising the blood flow and hence leading to symptoms. Normally ligament passes cranially to the origin of the celiac axis, whereas, in some cases, low insertion of ligament causes compression of the celiac axis leading to recurrent abdominal pain. It can be easily diagnosed on CT angiography showing focal narrowing (hooked up appearance) of the celiac axis in the region of compression. Here, we present a rare case of an 18-year-old male patient presenting with recurrent abdominal pain.

CASE DESCRIPTION

An 18-year-old male visited the emergency room with a complaint of recurrent episodes of abdominal pain. There was no past or any medical history of any previous illness. On physical examination, the patient was afebrile and had pallor. All the vitals were within normal limits. The abdominal examination did not reveal any significant abnormality. Laboratory investigations (Hb, GBP, TLC) were within normal limits. The patient was then referred to the Department of radiodiagnosis for ultrasound whole abdomen (USG) which was found to be normal at first two instances. The patient visited again for a color Doppler ultrasound for the aorta, and coeliac trunk which revealed high-velocity flow in the coeliac trunk with aliasing of blood noted at the site of “kink” noted in the coeliac artery just proximal to its origin from the abdominal aorta (Figs 1 to 3).



Fig. 1: USG image demonstrating compression of celiac trunk (kink) proximal to its origin from abdominal aorta

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Fig. 2: Color Doppler image demonstrating aliasing of blood at the site of compression of celiac trunk

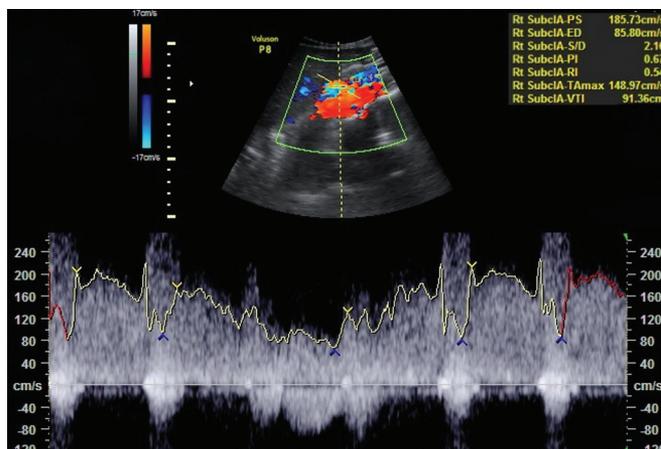


Fig. 3: Doppler image demonstrating high peak systolic flow of celiac trunk at the site of compression



Fig. 4: CECT sagittal image of abdomen in arterial phase demonstrating “kink” in celiac trunk noted proximal to the site of origin from abdominal aorta (arrow)

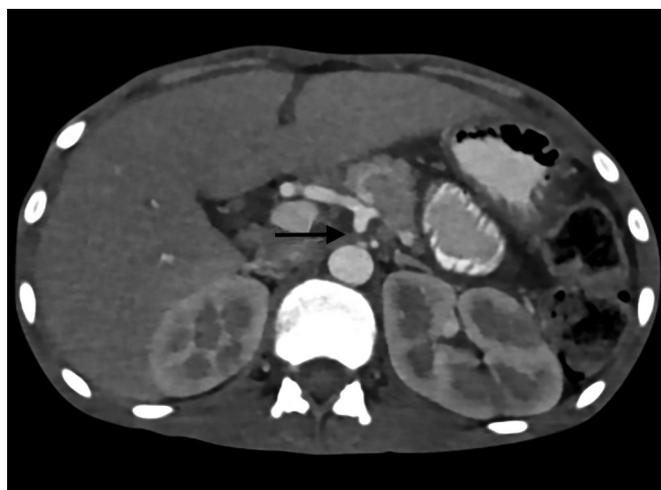


Fig. 5: CECT axial image of abdomen in arterial phase demonstrating compression of celiac trunk by median arcuate ligament at the site of its origin from aorta (arrow)

To confirm the above-mentioned findings, a contrast-enhanced CT scan (CECT) of the abdomen was advised which revealed the characteristic “hooked up” appearance of the coeliac trunk in the sagittal images (Figs 4 and 5). The patient then underwent laparoscopic resection of the median arcuate ligament after which the symptoms were resolved.

DISCUSSION

Median arcuate ligament syndrome (aka coeliac artery compression syndrome) is a rare cause of mesenteric ischemia presenting with weight loss and epigastric pain.¹ It is characterized by both clinical and radiological features. The median arcuate ligament is a fibrous ligament which unites both the diaphragmatic crura across the aortic hiatus. Generally, the ligament passes, at the level of L1 vertebra over the aorta, above the origin of coeliac trunk. In 10 to 20% cases the origin of the ligament is found at a lower level, thereby causing indentation of the proximal part of the coeliac trunk, leading to clinical symptoms.²

However, in the majority of cases, the indentation over coeliac trunk does not produce symptoms because of the release of pressure during inspiration as the coeliac trunk moves more caudally. It is during expiration that the compression of coeliac artery truly manifests.^{3,4}

A combined clinical and radiological approach is required in the diagnosis of median arcuate ligament syndrome.

Radiological Diagnosis

In symptomatic patients, USG of the abdomen is the first investigation to be done, which is usually normal. On color Doppler examination, a characteristic kink is noted at the site where the coeliac trunk is compressed by the overlying median arcuate ligament,⁵ showing aliasing at the site of kinking. The compression site denotes high peak systolic velocity (PSV) at the site of compression. On a CECT scan, the coeliac artery shows the classical “hooked appearance” at the site of compression. This appearance is characteristic to median arcuate ligament

syndrome and thereby helps in distinguishing it from other etiologies causing compression of the coeliac artery, like atherosclerosis. Three-dimensional (3D) CT imaging is also valuable to detect any collateral vascular channels formed due to coeliac artery compression; this helps in determining the hemodynamic significance of the narrowing and is important for planning of surgical procedures.

Treatment

In a few patients who indeed have hemodynamic and clinical symptoms, laparoscopic separation of the ligament causes relief in symptoms.⁶ In other cases, the constriction of the celiac axis by the ligament can cause vascular damage, requiring vascular reconstruction.⁷

CONCLUSION

Median arcuate ligament syndrome is a rare clinical entity which can be found in normal asymptomatic people. In few cases, however, celiac axis compression can cause symptoms which can be relieved with laparoscopic surgical decompression.

CLINICAL SIGNIFICANCE

The median arcuate ligament syndrome as an entity must be kept in mind when no other cause of abdominal pain is zeroed on even after all the clinical examinations and investigations.

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