Median Arcuate Ligament Syndrome: Evaluation with CT Angiography[1]

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The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes superior to the origin of the celiac axis. However, in some people, the ligament inserts low and thus crosses the proximal portion of the celiac axis, causing compression and sometimes resulting in abdominal pain. The diagnosis of clinically significant celiac axis compression, referred to as median arcuate ligament syndrome, is traditionally made with conventional angiography; however, the condition can now be diagnosed with three-dimensional computed tomographic (CT) angiography. In patients with median arcuate ligament syndrome, CT angiograms demonstrate a characteristic focal narrowing in the proximal celiac axis. The focal narrowing has a characteristic hooked appearance, which can help distinguish this condition from other causes of celiac artery narrowing, such as atherosclerotic disease. Once the disorder has been diagnosed, surgery can be performed to relieve the compression. In some patients, the ligamentous constriction of the celiac axis causes vascular damage, which may require vascular reconstruction. CT angiography can play a role in the diagnosis of median arcuate ligament syndrome by demonstrating the characteristic focal narrowing of the celiac artery in patients presenting with the appropriate clinical symptoms.

Abbreviation: 3D = three dimensional

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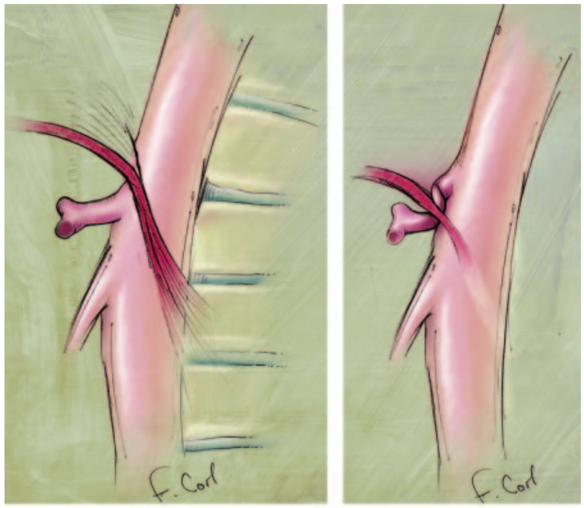
Introduction

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes superior to the origin of the celiac axis. In 10%–24% of people, however, the ligament may cross anterior to the artery; in some of these individuals, the ligament may actually compress the celiac axis, compromising blood flow and causing symptoms (1). Although the diagnosis of celiac artery compression is traditionally made by using conventional angiography, the use of thin-section multidetector computed tomography (CT) and of three-dimensional (3D) imaging techniques has greatly improved the ability to obtain detailed images of the mesenteric vessels noninvasively. Compression of the celiac axis by the median arcuate ligament produces characteristic findings visible at CT angiography. This article reviews the clinical and radiologic findings in median arcuate ligament syndrome (also referred to as celiac artery compression syndrome). The potential pitfalls in the diagnosis are also discussed.



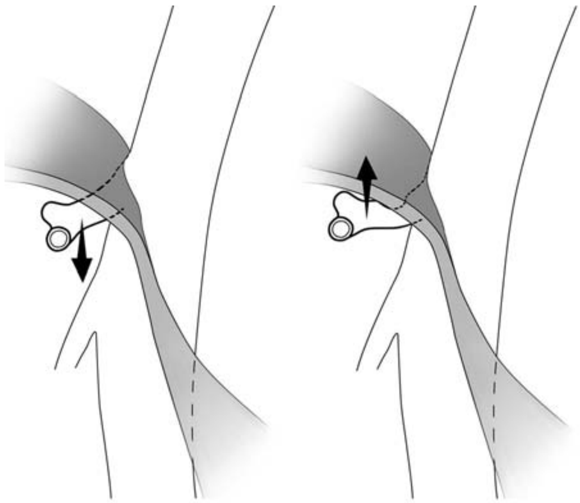
1. 2.

Figures 1, 2. **(1)** Drawing from the sagittal perspective demonstrates the typical anatomy of the median arcuate ligament as it crosses anterior to the aorta and superior to the origin of the celiac axis. **(2)** Another sagittal view shows a normal variant anatomy in which the median arcuate ligament crosses the proximal portion of the celiac axis, causing an indentation.

Anatomy

The diaphragmatic crura arise from the anterior surface of the L1-L4 vertebral bodies on the right and the first two or three lumbar vertebral bodies on the left. In addition, the crura arise from the intervertebral disks and anterior longitudinal ligament (2). The crura pass superior and anterior to surround the aortic opening and to join the central tendon of the diaphragm.

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes over the aorta at the level of the first lumbar vertebral body, superior to the origin of the celiac axis (Fig 1). However, in 10%–24% of people, the ligament may be low and therefore cross over the proximal portion of the celiac axis, causing a characteristic indentation (Fig 2). In a small subset of these patients, the median arcuate ligament can compress the proximal celiac axis enough to be hemodynamically significant and cause symptoms.

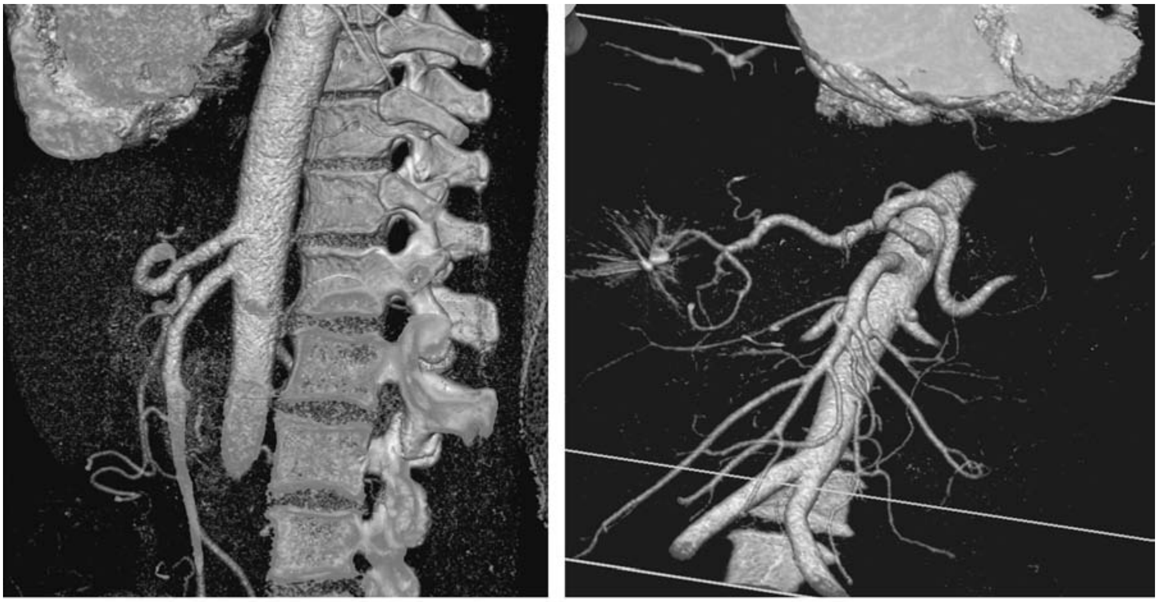


3. 4.

Figures 3, 4. Diagrams show the orientation of the celiac axis during inspiration **(3)** and expiration **(4)**.

Median Arcuate Ligament Syndrome

The median arcuate ligament syndrome (or celiac artery compression syndrome) was first described in 1963 by Harjola (3). The definition of the syndrome relies on a combination of both clinical and radiographic features. It typically occurs in young patients (20–40 years of age) and is more common in thin women, who may present with epigastric pain and weight loss (4). The abdominal pain may be associated with eating, but not always (5). At physical examination, an abdomi-



a. b.

Figure 5. **(a)** Sagittal 3D volume-rendered image of a normal aorta demonstrates the normal appearance of the celiac axis and superior mesenteric artery. **(b)** Coronal oblique 3D volume-rendered image shows the normal anatomy of the celiac axis and superior mesenteric artery.

nal bruit that varies with respiration may be audible in the midepigastric region. Symptoms are thought to arise from the compression of the celiac axis, resulting in a compromise in blood flow.

Radiologic Diagnosis

Conventional Angiography

In the past, median arcuate ligament syndrome was detected during conventional angiography. When the median arcuate ligament passes anterior to the celiac axis, a characteristic superior indentation is noted along the proximal celiac axis, usually about 5 mm from its origin at the abdominal aorta. Any compression caused by this indentation typically is less apparent during inspiration, when the celiac axis assumes a more caudal orientation as the lungs expand. During expiration, compression of the celiac axis typically increases (Figs 3, 4).

Isolated compression of the celiac axis during expiration may not be clinically significant. It is important to realize that up to 13%–50% of healthy patients may exhibit the angiographic feature of compression to a variable degree, especially during expiration (6,7). The vast majority of these patients have no symptoms. Therefore, the radiographic finding of celiac axis compression alone may not be significant, unless it is correlated with clinical symptoms.

Severe compression occurs in approximately 1% of patients and persists during inspiration (7,8). Severe stenosis will result in poststenotic dilatation, and in some cases, the celiac axis will be fed by the superior mesenteric artery via the pancreaticoduodenal arcade.

CT Angiography

Although the diagnosis of median arcuate ligament syndrome is traditionally made by using catheter angiography, the condition has been observed with Doppler ultrasound and CT (4,9). New thin-section multidetector CT scanners, along with 3D software, have greatly improved the ability to obtain high-resolution images of the aorta and its branches. The mesenteric vessels are very well visualized, and in many cases, CT angiographic examination will obviate conventional angiography (Fig 5).

For a complete CT angiographic evaluation in patients with suspected median arcuate ligament syndrome, 3D imaging is necessary. The findings characteristic of the syndrome may not be appreciated on axial images alone. Typically, the sagittal plane is optimal for visualizing the proximal portion of the celiac axis, and in many cases, 3D imaging also allows identification of the actual median arcuate ligament. In patients with median

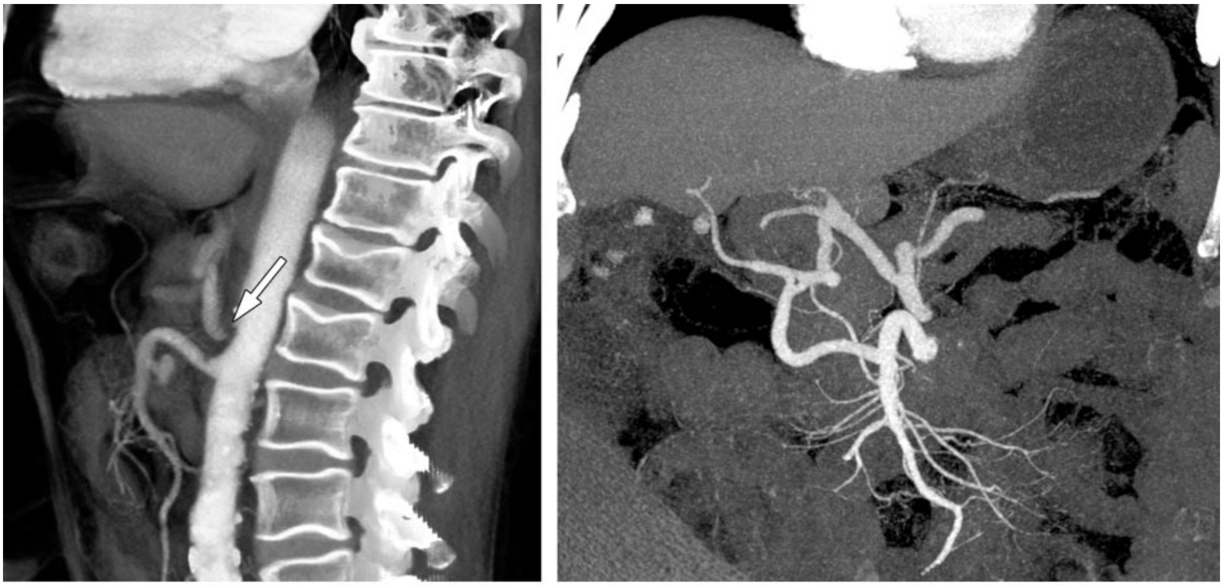
a. b.

Figure 6. **(a)** Sagittal 3D volume-rendered image shows normal anatomy of the celiac axis and superior mesenteric artery. In this case, there is a slight impression (arrow) in the proximal celiac axis, which may be related to a nonobstructing median arcuate ligament. **(b)** As evident from this axial oblique 3D volume-rendered image of the same patient, the median arcuate ligament and the indentation it causes on the proximal celiac axis can be difficult to detect in the axial plane.

a. b.

Figure 7. **(a)** Sagittal 3D image of a patient with epigastric pain demonstrates acute angulation and narrowing of the proximal celiac axis. There is minimal poststenotic dilatation, which creates a "hooked" appearance (arrow) that is characteristic of median arcuate ligament syndrome. **(b)** Coronal oblique CT angiogram of the same patient reveals a prominent collateral vessel and dilatation of the gastroduodenal artery (arrowhead) that is supplying the common hepatic artery (short arrow) off the superior mesenteric artery (long arrow). This configuration is a common collateral pathway seen in patients with celiac axis stenosis. The patient underwent surgical decompression and experienced relief of symptoms.

arcuate ligament syndrome, CT angiograms demonstrate a characteristic focal narrowing in the proximal celiac axis (Figs 6–8). The focal narrowing has a characteristic hooked appearance, which can help distinguish this condition from other causes of celiac artery narrowing, such as atherosclerotic disease (Figs 9, 10). CT is typi-



a. b.

Figure 8. **(a)** Sagittal volume-rendered CT angiogram of a patient with abdominal pain demonstrates characteristic kinking of the proximal celiac axis, creating a hooked appearance (arrow). **(b)** Coronal maximum intensity projection CT angiogram shows the prominent gastroduodenal artery that feeds the common hepatic artery off the superior mesenteric artery.

9.

10.

Figures 9, 10. **(9)** Sagittal 3D volume-rendered image of a patient with abdominal pain demonstrates a focal narrowing of the proximal celiac axis caused by atherosclerotic disease. This appearance is distinct from the hooked appearance seen in median arcuate ligament syndrome. **(10)** Sagittal maximum intensity projection image of a patient with recurrent abdominal pain reveals focal calcified plaque that causes narrowing of the proximal celiac axis. There is minimal poststenotic dilatation.

cally performed during inspiration; therefore, if the focal narrowing is observed during inspiratory CT, it may be clinically significant, since the transient compression seen only during expiration in some patients would not be manifested at inspiratory CT. Also, associated poststenotic dilatation or collateral vessels may suggest actual pathologic conditions and warrant clinical correlation.

The multiplanar capabilities of 3D imaging are also valuable in identifying the collateral vessels, which develop as a result of celiac axis compression. This information can help determine the hemodynamic significance of the narrowing and is important for surgical planning.

In the imaging protocol for CT angiography, intravenously administered contrast material is necessary to image the mesenteric vessels. We use 120 mL of nonionic contrast agent injected at a rate of 3 mL/sec. Scans are obtained during both the arterial (25 seconds after injection) and venous (50 seconds after injection) phases to ensure opacification of the mesenteric arteries and veins. On our multidetector CT scanner (Siemens Sensation 16; Siemens Medical Solutions, Malvern, PA), we use the 16 × 0.75-mm collimator setting to obtain 0.75-mm-thick sections reconstructed every 0.5 mm for 3D imaging. For hard-copy review, 3-mm-thick sections are adequate.

All the data are then transferred to our 3D workstation (Leonardo workstation; Siemens Medical Solutions, Malvern, Pa) for volume rendering. Use of the 0.75-mm collimators creates isotropic data sets, which allow production of excellent quality 3D images of the aorta and its branches. All images are reviewed by using InSpace software (Siemens Medical Solutions), which enables multiplanar reconstructions as well as interactive 3D volume rendering. The brightness, opacity, window width, and window level can be adjusted in real time to optimize visualization of the mesenteric vessel. We usually use a combination of volume-rendered and maximum intensity projection images. Review and reconstruction of the 3D images can usually be completed in approximately 5 minutes.

Treatment

The surgical management of median arcuate ligament syndrome is controversial (10). Many more patients have the anatomic abnormality of low insertion of the median arcuate ligament than actually have symptoms caused by the abnormality. Surgical treatment is more likely to relieve symptoms in patients 40–60 years of age with postprandial pain, greater than 20-lb weight loss, and poststenotic dilatation and collateral vessels (11). In the rare patients who indeed have such symptoms, some will experience relief when the ligament is divided surgically, a task that can now be accomplished laparoscopically (12,13). In others, the ligamentous constriction of the celiac axis causes vascular damage, which may require vascular reconstruction (14). Because surgical ligation of the constricting ligamentous band has been performed with only variable success, many surgeons no longer accept the median arcuate ligament syndrome as a true clinical entity (6,15).

Conclusions

Median arcuate ligament syndrome is a controversial entity. It is clear that the abnormally low insertion of the median arcuate ligament can be found in normal asymptomatic people. In a small subset of patients, however, the compression of the celiac axis can cause symptoms that may be relieved after surgical decompression. CT angiography can play a role in the diagnosis of this condition by demonstrating the characteristic focal narrowing of the celiac artery in patients presenting with the appropriate clinical symptoms.

References

1. Lindner HH, Kemprud E. A clinicoanatomic study of the arcuate ligament of the diaphragm. Arch Surg 1971;103:600–605.
2. Skandalakis JE, Gray SW, Rowe JS Jr. Surgical anatomy of the diaphragm. In: Nyhus L, Baker R, eds. Mastery of surgery. Boston, Mass: Little, Brown, 1984; 307–318.
3. Harjola PT. A rare obstruction of the coeliac artery. Ann Chir Gynaecol Fenn 1963;52:547–550.
4. Sproat IA, Pozniak MA, Kennell TW. US case of the day: median arcuate ligament syndrome. RadioGraphics 1993;13:1400–1402.
5. Dunbar JD, Molnar W, Berman FF, et al. Compression of the celiac trunk and abdominal angina: preliminary report of 15 cases. Am J Roentgenol Radium Ther Nucl Med 1965;95:731–744.
6. Szilagyi DE, Rian RL, Elliott JP, Smith RF. The cardiac artery compression syndrome: does it exist? Surgery 1972;72:849–863.
7. Bron KM, Redman HC. Splanchnic artery stenosis and occlusion: incidence, arteriographic, and clinical manifestations. Radiology 1969;92:323–328.
8. Cornell SH. Severe stenosis of celiac axis: analysis of patients with and without symptoms. Radiology 1971;99:311–316.
9. Patten RM, Coldwell DM, Ben-Menachem Y. Ligamentous compression of the celiac axis: CT findings in five patients. AJR Am J Roentgenol 1991;156:1101–1103.
10. Bech FR. Celiac artery compression syndromes. Surg Clin North Am 1997;77:409–424.
11. Reilly LM, Armmar AD, Stoney RJ, Ehrenfeld WK. Late result following operative repair for celiac artery compression syndrome. J Vasc Surg 1985;2:79–81.
12. Kernohan RM, Barros D'Sa AA, Cranley B, Johnston HM. Further evidence supporting the existence of the celiac artery compression syndrome. Arch Surg 1985;120:1072–1076.
13. 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;32:814–817.
14. Takach TJ, Livesay JJ, Reul GJ Jr, Cooley DA. Celiac compression syndrome: tailored therapy based on intraoperative findings. J Am Coll Surg 1996;183:606–610.
15. Plate G, Eklof B, Vang J. The celiac compression syndrome: myth or reality? Acta Chir Scand 1981;147:201–203.