Diagn Interv Radiol 2007; 13:90-93

Celiac artery compression syndrome: diagnosis with multislice CT

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ABSTRACT
Celiac artery compression syndrome is a rare disorder characterized by postprandial intestinal angina caused by insufficient blood supply to the gastrointestinal organs. In this syndrome, the root of the celiac artery is compressed and narrowed by the median arcuate ligament of the diaphragm during expiration. We report here 3 such cases that were diagnosed by the use of multislice computed tomography.

Key words: • *celiac artery* • *computed tomography*

Celiac artery compression syndrome (CACS), which is also known as median arcuate ligament syndrome, is a rare disorder characterized by postprandial intestinal angina caused by insufficient blood supply to the gastrointestinal organs (1). In this syndrome, the root of the celiac artery is compressed and narrowed by the median arcuate ligament of the diaphragm during expiration, causing abdominal angina (2, 3). We report here 3 such cases that were diagnosed by the use of multislice computed tomography (MSCT).

Case reports

Case 1

A 17-year-old male patient presented with chronic postprandial abdominal pain and marked weight loss. The pain was localized in the epigastric region and occurred after meals. The physical examination was unremarkable, except for a bruit in the upper mid-epigastrium. Investigations for gastrointestinal tract pathology, including abdominal ultrasonography (US), routine abdominal spiral CT, upper gastrointestinal tract endoscopy, and rectosigmoidoscopy, as well as genetic tests for familial Mediterranean fever (FMF) were all negative. Laboratory blood tests were unremarkable. Selective catheter angiography of the celiac axis, which was done in an outside hospital, showed multiple thrombi in the hepatic artery, which was filling by collateral pathways via the gastroduodenal artery. Both the aortic and celiac truncus images were obtained in the anteroposterior projection; lateral views were not obtained. There were no signs of extravisceral thrombosis. The etiology of the thrombosis remained unclear, despite comprehensive hematological and clinical examinations. Abdominal MSCT, including visceral angiography, was performed 6 days after the catheter angiography. The scanning was made from the diaphragmatic level to the iliac bifurcation with a 16-slice CT scanner (Light Speed 16, GE Medical Systems, Milwaukee, Wisconsin, USA) following injection of 120 ml of 300 mg/ml non-ionic contrast medium at a rate of 5 ml/sec. The examinations were performed during expiration. The CT protocol was as follows: slice thickness, 1.25 mm; pitch, 1.375:1, rotation rate, 0.5 per second. The delay time was 18–20 s. Images were sent to an online workstation. Axial and reformatted MSCT images revealed compression of the celiac axis by the median arcuate ligament, with poststenotic dilatation (Fig. 1a, b). The compression of the celiac axis created a hooked appearance in the sagittal view. The hepatic artery was normal and there were no filling defects. The patient underwent laparotomy, during which the celiac axis was observed to be severely compressed anteriorly by soft tissue formation due to the hypertrophied median arcuate ligament. An excision of the hypertrophied ligament was performed (Fig. 1c); however, the patient was not relieved of symptoms at follow-up.

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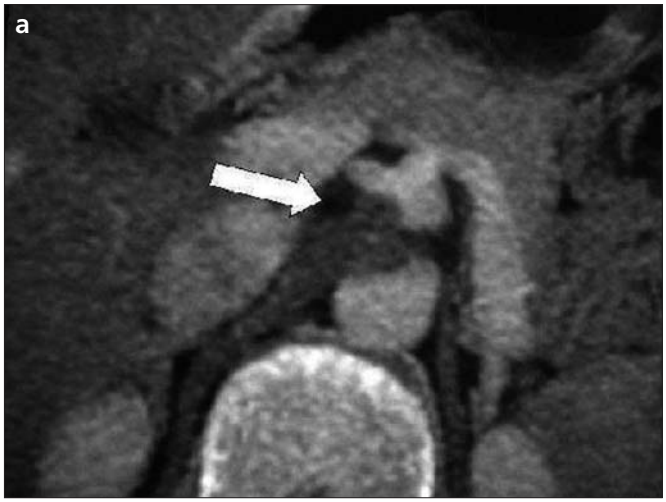
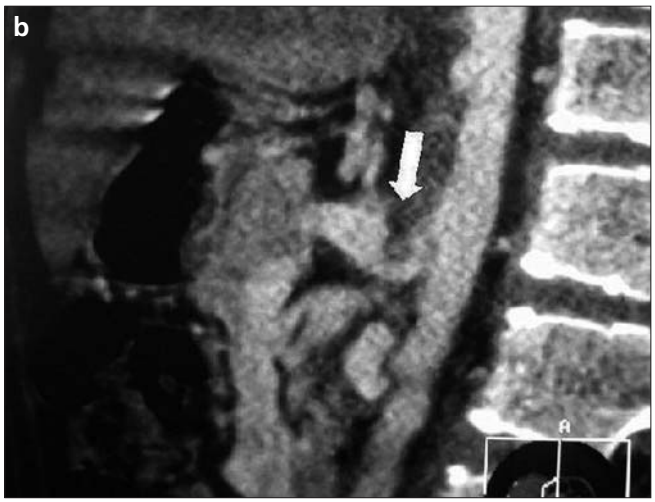
Received 5 December 2005; revision requested 27 March 2006; revision received 11 April 2006; accepted 17 April 2006.





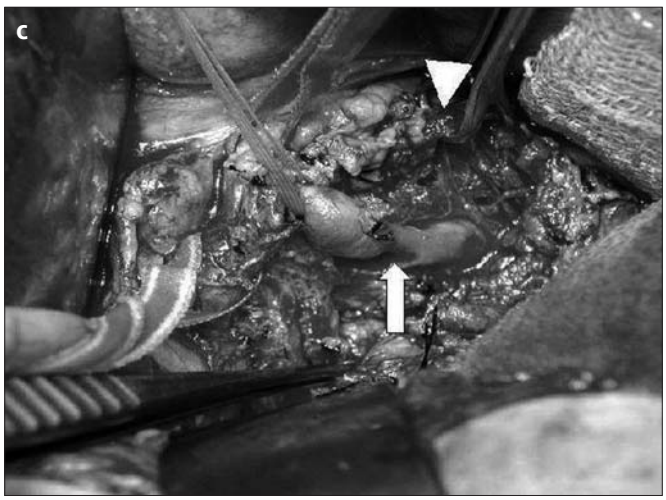


Figure 1. a–c. A 17-year-old male patient with celiac artery compression syndrome (case 1). Axial CT image **(a)** and sagittal reformatted CT image **(b)** in the arterial phase show compression of the celiac axis by the hypertrophied median arcuate ligament with poststenotic dilatation *(***a**, *arrow)*. There is kinking in the proximal celiac axis which creates a characteristic hooked appearance on the sagittal view *(***b**, *arrow)*. The excised hypertrophied median arcuate ligament *(***c**, *arrowhead)* and stenotic segment of celiac axis *(***c**, *arrow)* were seen during laparotomy.

Case 2

A 47-year-old male patient presented with chronic abdominal pain that had been ongoing for 15 years. The pain was localized in the upper abdomen and it was more prominent after meals, along with some nausea. The physical examination was unremarkable. Previous workup, including blood examinations, excretory urography, spiral pancreatic CT, abdominal US, colonoscopy, endoscopic retrograde cholangiography, upper gastrointestinal tract endoscopy, coronary angiography, enteroclysis, magnetic resonance imaging (MRI) of the lumbosacral region, and genetic tests for FMF were all normal. Selective catheter angiography of the celiac axis was normal on anteroposterior view; a lateral angiogram was not obtained. Laboratory blood tests were unremarkable, except for anemia (Hb, 9.9 g/dl). The patient underwent an exploratory laparotomy in our hospital, without a specific preoperative diagnosis. Perioperative examination of the abdomen did not reveal any pathological finding, except for interloop adhesions due to previous surgery. Following the laparotomy, the patient was examined with MSCT and compression of the celiac axis by the hypertrophied median arcuate ligament with poststenotic dilatation was observed, and CACS was diagnosed. Because the symptoms of the patient continued, a second laparotomy was considered for CACS, but the patient declined the procedure.

Case 3

A 54-year-old female patient presented with chronic abdominal pain, which had been ongoing for 3 years. The pain was localized in the epigastric region, and it was not related to meals. The physical examination was unremarkable, with nonspecific epigastric tenderness. The radiological examinations, including abdominal US and routine abdominal spiral CT, were normal. Genetic tests for FMF and laboratory blood tests did not reveal any pathology. The patient previously underwent a diagnostic laparotomy at a different surgical clinic and no cause for the abdominal pain was identified. After he was discharged, the same symptoms continued without any improvement. The patient was then referred to our hospital for advanced evaluation. MSCT examination was performed and CACS findings, which included severe stenosis of the celiac axis by the median arcuate ligament with poststenotic dilatation were observed (Fig. 2).

Discussion

Harjola (4) and Dunbar et al. (5), in 1963 and 1965, respectively, initially described CACS. The clinical triad, which includes epigastric pain, weight loss, and postprandial pain with emesis, characterizes this syndrome. Postprandial epigastric pain, in particular, occurs with expiration. This syndrome is also known as median arcuate ligament syndrome. The median arcuate ligament is found at the T12–L1 level and bridges the crura of the diaphragm, just anterior to the aorta (6). In patients with CACS, the median arcuate ligament compresses the celiac artery during expiration. While patients are in an erect position, with inspiration, the celiac ar-

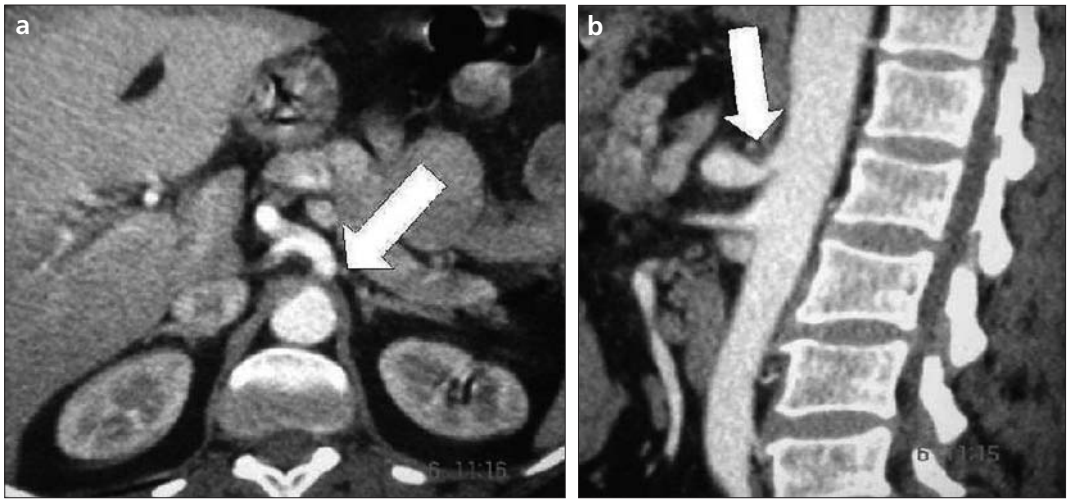


Figure 2. a, b. A 54-year-old female patient with celiac artery compression syndrome (case 3). Axial CT image **(a)** and sagittal reformatted CT image **(b)** show the hypertrophied median arcuate ligament causing severe stenosis (hooked appearance) of the celiac axis at the anterior margin of abdominal aorta, with poststenotic dilatation *(arrows)*.

tery descends to the abdominal cavity, and compression is often relieved with a more vertical orientation of the celiac artery (3).

There are 2 main theories used to explain the pathogenesis of the symptoms (7–9). The first theory is mesenteric ischemia due to celiac artery compression. Mesenteric ischemia arises either from direct foregut ischemia or, alternatively, through postprandial steal via collaterals from the superior mesenteric to the celiac bed, leading to midgut ischemia. The second theory has it that neurogenic stimulation is caused by compression of the celiac ganglion and plexus. Neurogenic stimulation-related pain, can be caused either from celiac plexus stimulation leading to splanchnic vasoconstriction or via direct sympathetic pain fiber irritation (10). Although the syndrome has been described in the 1960s, controversy continues as to whether celiac compression leads to the clinical picture or not. The controversy stems from an undefined pathophysiological mechanism and the existence of celiac compression in asymptomatic patients. It has been reported that 13%–50% of healthy individuals may exhibit, to a variable degree, angiographic features of compression especially during expiration (11). Furthermore, treatment approaches, such as ligament excision or ganglion blockage, can be ineffective in relieving the symptoms. Thus, it is difficult to identify patients who will benefit from surgical intervention which consequently requires diligent and careful elimination of other causes of abdominal pain.

Celiac artery compression may be investigated with Doppler US, spiral CT angiography, selective catheter angiography, and magnetic resonance angiography (3, 7, 12). Doppler US has been reported to have a high sensitivity for the diagnosis of CACS and was proposed to be the modality of choice for diagnosing CACS (13). The gold standard diagnostic method is selective angiography, which should be performed during both inspiration and expiration, in the lateral position (2, 12). However, recent implementation of MSCT with 4- and 16-row detectors has permitted the acquisition of images with 1.25-mm slice thickness during a single breath-hold of 10–12 sec. These thinner images not only provide increased resolution and improved lesion detection, but also have permitted the production of excellent multiplanar reconstruction as well as angiographic images without the stair-step artifact of thicker slices (14). In our study with 16-slice CT, angiographic images excellently demonstrated the vascular anatomy and compression without any need for catheter angiography, which is a more invasive technique associated with considerable side effects.

Although many cases with CACS diagnosed with catheter angiography have been reported, reports based on CT diagnosis are rare. CT was first found to demonstrate CACS on axial images by Patten et al. (15) in 5 patients with CACS. With the advent of CT equipment, several cases of CACS diagnosed with CT have been reported (1, 7, 12, 16)

CT angiography demonstrates a characteristic focal narrowing in the proximal celiac axis in patients with CACS. The focal narrowing has a characteristic hooked appearance, which can be useful in distinguishing this condition from other causes of celiac artery stenosis such as atherosclerosis. Poststenotic dilatation and collateral vascular vessels are among the features of CACS (17). The characteristic hooked appearance and poststenotic dilatation were seen in all 3 of our patients; however, we did not observe collateral vessels on CT angiography.

In the 3 presented cases, the diagnosis of CACS was made with MSCT, with multiplanar reformatted and 3D angiographic images. The lateral view of the celiac truncus was the most useful. In cases 1 and 2, although catheter angiography examinations were performed, the diagnosis of CACS could not be made because we obtained only anteroposterior views of aortograms. Kopecky et al. (12) reported a case with CACS in whom the diagnosis was made with spiral CT angiography, although 2 previous catheter angiography examinations were performed. They emphasized the importance of lateral aortograms, and even oblique projections, for the diagnosis of celiac stenosis. They suggested that spiral CT angiography might be superior to conventional catheter arteriography, which has been the gold standard for

the diagnosis of CACS. In cases 1 and 2, diagnosis could not be achieved with catheter angiography, or even by laparotomy (case 2). Appropriate projections, including lateral aortograms were not available for these patients. In fact, the negative laparotomy was due to the surgeon's lack of knowledge about CACS and the need for considering this syndrome in patients with chronic abdominal pain.

The options of treatment for CACS include surgical or laparoscopic division of the median arcuate ligament, celiac ganglion destruction, and bypass surgery (7, 10, 16). Surgical treatment of CACS is the first choice. Surgical treatment with division of the median arcuate ligament and excision of celiac plexus is adequate for patients without persistent vessel deformity (16).

Catheter angiography has been traditionally used to confirm the diagnosis of celiac compression; however, the diagnosis may be easily missed if optimal views are not obtained. Because the ostium of the celiac truncus is located anteriorly, only lateral aortograms can show the stenosis.

In conclusion, MSCT is a minimally invasive and useful diagnostic tool for the diagnosis of CACS, which can be used alternatively to invasive angiograms.

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