

MDCT Evaluation of Celiac Axis Compression Syndrome- A Rare Case Series

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ABSTRACT

Celiac axis compression syndrome is an uncommon entity of abdominal vascular compression syndrome which results from the compression of the proximal celiac artery by the hypertrophied median arcuate ligament. The patients present clinically with postprandial abdominal pain predominantly in the epigastric region and weight loss. The use of thin-section Multidetector Computed Tomography (MDCT) and 3-Dimensional (3D) image reconstruction has greatly improved the ability to obtain detailed images and helped to assess the abdominal vascular system non invasively. Five cases of celiac axis compression syndrome with its characteristic MDCT imaging features like hooked appearance, thickened median arcuate ligament more than 4 mm, poststenotic dilatation and posterior and inferior displacement of celiac artery with indentation on aorta have been described in the present case series and the imaging findings described by other authors in their reported studies have also been compared.

Keywords: Dunbar syndrome, Median arcuate ligament syndrome, Multidetector computed tomography, Vascular compression syndrome

INTRODUCTION

Celiac Axis Compression Syndrome (CACS) is a rare condition caused by the compression of the celiac trunk externally by the median arcuate ligament [1]. Initially, it was described by Harjola PT, in 1963 and this condition is known by different names, such as Harjola-Marable syndrome/Median Arcuate Ligament Syndrome (MALS) and Dunbar syndrome [2-4]. It's a rare condition with a reported incidence of 1.76% to 4% and to occur in 2 per 100,000 persons. It is more commonly seen in thin women in the ratio of 2:1 and 3:1 and between the age group of 20-40 years [4,5].

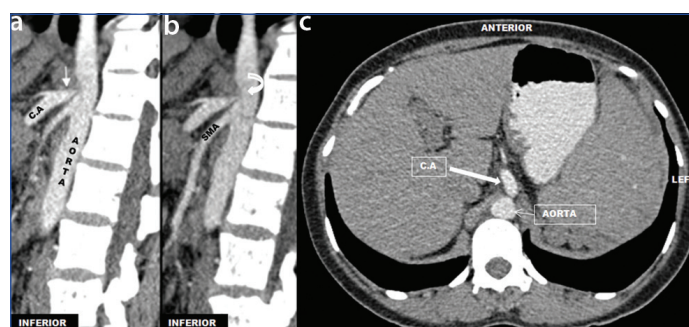
In the present case series, out of five cases of CACS, two cases showed additional findings of indentation on the aorta by hypertrophied Median Arcuate Ligament (MAL) and a brief description of its characteristic Multidetector Computed Tomography (MDCT) imaging findings is presented, which will help to diagnose this condition non invasively. The findings obtained have also been compared to imaging findings described by other reported studies.

CASE SERIES

Case 1

A 38-year-old female patient presented to the hospital with clinical symptoms of easy fatigability and weight loss since three months. The patient had abdominal pain in the epigastric region for one month which was insidious in onset and got aggravated since seven days. The provisional diagnosis was pancreatitis. The laboratory findings were within normal limits. Hence, the patient was advised contrast- enhanced MDCT abdomen for evaluating cause of pain.

The MDCT identified a focal extrinsic compression of proximal celiac artery by the hypertrophied median arcuate ligament, which measured approximately 8 mm in thickness. There was significant kinking of proximal celiac artery causing grade III stenosis with posterior displacement and post-stenotic dilatation of celiac artery. Hypertrophy of median arcuate ligament and diaphragmatic crura causing indentation on the aorta [Table/Fig-1a-c]. With theses imaging findings, diagnosis of CACS was made. The patient was managed conservatively and was advised surgery if symptoms aggravates. Follow-up of this patient could not be done.



[Table/Fig-1]: Contrast-enhanced MDCT of the abdomen of case 1: (a) and (b) sagittal section showing thickened median arcuate ligament (short white arrow) causing focal extrinsic compression of anterior wall and kinking of the proximal celiac artery with posterior displacement of celiac artery and indentation on the aorta (Curved arrow); (c) Axial section showing poststenotic dilatation of the proximal celiac artery (bold arrow).

C.A: Celiac artery; SMA: Superior mesenteric artery

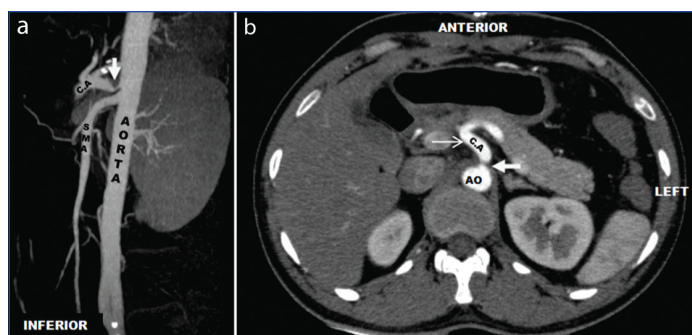
Case 2

A 42-year-old male patient presented in Outpatient Department of Medicine with complaints of pain abdomen since one month. It was gradual in onset and progressive in nature, localised to right epigastric region. It was associated with vomiting since two days which was non projectile and aggravates after taking food. The provisional diagnosis of gastritis/pancreatitis was made. An endoscopic finding was uneventful and ultrasound abdomen showed mild ascities. Contrast-enhanced MDCT abdomen was advised for detailed evaluation.

The MDCT showed a focal narrowing of proximal celiac artery by the thickened median arcuate ligament, which measured approximately 5 mm in thickness. There was classical hooked appearance due to kinking of the proximal celiac artery along with posterior and inferior displacement of the celiac artery. Grade III type of stenosis was present with poststenotic dilatation of celiac artery [Table/Fig-2a,b]. With these imaging findings, diagnosis of CACS was made. The patient was managed conservatively.

Case 3

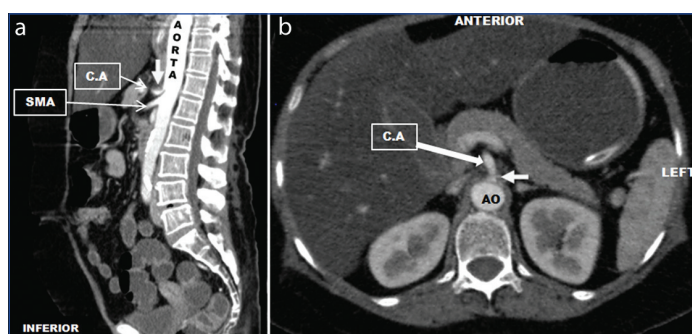
A 65-year-old female patient presented to the hospital with complaints of pain abdomen since two months. It was insidious in



[Table/Fig-2]: Contrast-enhanced MDCT of the abdomen of case 2: (a) Sagittal section surface shaded display showing thickened median arcuate ligament (bold white arrow) causing focal narrowing of proximal celiac artery. Classical hooked appearance due to kinking of the proximal celiac artery noted along with posterior and inferior displacement of the proximal celiac artery; (b) Axial section showing stenosis near the proximal portion of the celiac artery (bold arrow) with resultant post stenotic dilatation (white arrow).
C.A: Celiac artery; SMA: Superior mesenteric artery; AO: Aorta

onset, persistent type, localised to epigastric region and non radiating which aggravates on taking food. The patient also complained of non projectile type of vomiting since two month, multiple episodes per day and aggravates after taking food. There was past history of analgesic drug abuse for knee pain. The provisional diagnosis of acute gastritis was made. Patient underwent upper gastrointestinal endoscopy, showed features of Gastroesophageal Reflux Disease (GERD) with atrophic gastritis. Contrast- enhanced MDCT abdomen was advised to rule out intestinal obstruction and mesenteric ischaemia.

The MDCT showed, a focal narrowing of proximal celiac artery by the thickened median arcuate ligament, which measured approximately 4.5 mm in thickness. There was classical hooked appearance due to kinking of the proximal celiac artery along with posterior and inferior displacement of the celiac artery. Grade II type of stenosis was present with minimal poststenotic dilatation of celiac artery [Table/Fig-3a,b]. There was no abnormal dilatation of bowel loops and mesenteric artery thrombus. With theses imaging findings, diagnosis of CACS was made. The patient was managed conservatively.



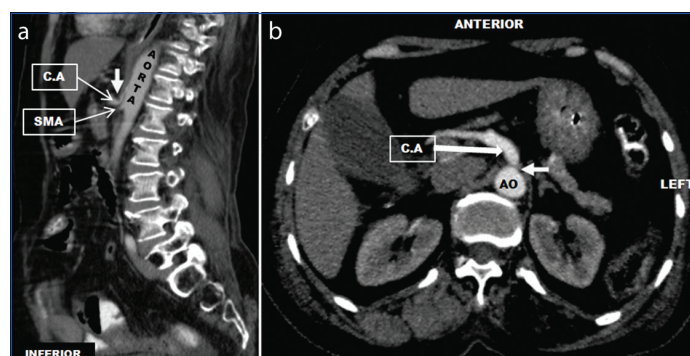
[Table/Fig-3]: Contrast-enhanced MDCT of the abdomen of case 3: (a) Sagittal section showing thickened median arcuate ligament causing focal narrowing of proximal celiac artery with hooked appearance due to kinking of the artery noted along with posterior and inferior displacement (bold white arrow). (b) Axial section showing stenosis near the proximal portion of the celiac artery (bold arrow).
C.A: Celiac artery; SMA: Superior mesenteric artery; AO: Aorta

Case 4

A 53-year-old female patient presented to the hospital with complaints of sudden onset of abdominal pain since one day and it was progressive in nature. Pain was present in all the quadrant of abdomen. The patient had multiple episode of vomiting which was watery in consistency. Past history of surgery for intestinal obstruction was present. With these clinical findings, the provisional diagnosis of intestinal obstruction secondary to adhesion was made and contrast- enhanced MDCT abdomen was advised.

The MDCT showed, a focal narrowing of proximal celiac artery by the thickened median arcuate ligament, which measured approximately 6 mm in thickness. There was classical hooked/

“J”-shaped appearance due to kinking of the proximal celiac artery along with posterior and inferior displacement of the celiac artery. Grade II type of stenosis was present with poststenotic dilatation of celiac artery [Table/Fig-4a,b]. There was no abnormal dilatation of bowel loops. With theses imaging findings, diagnosis of CACS was made. The patient was managed conservatively.

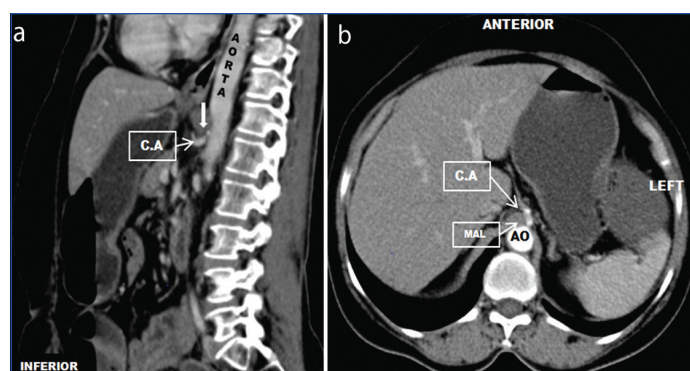


[Table/Fig-4]: Contrast-enhanced MDCT of the abdomen of case 3: (a) Sagittal section showing thickened median arcuate ligament causing classical hooked / “J”-shaped appearance due to kinking of the proximal celiac artery along with posterior and inferior displacement of the celiac artery (bold white arrow). (b) Axial section showing stenosis near the proximal portion of the celiac artery (bold arrow) and post stenotic dilatation (long bold arrow).
C.A: Celiac artery; SMA: Superior mesenteric artery; AO: Aorta

Case 5

A 62-year-old female patient presented to the hospital with complaints of constipation since four days which was gradual in onset, progressive in nature and subsequently developed diffuse abdominal pain along with abdominal distension since two days. It was insidious in onset, gradual in progression with no aggravating or relieving factors. Patient was known case of rectosigmoid carcinoma and underwent surgical resection of growth two years back. With all these presenting clinical symptoms and past history, clinical diagnosis of recurrent growth of colon was made and contrast- enhanced MDCT abdomen was advised.

The MDCT showed, a focal extrinsic compression of proximal celiac artery by the hypertrophied median arcuate ligament, which measured approximately 10 mm in thickness. There was significant kinking of proximal celiac artery causing grade III stenosis with posterior displacement and poststenotic dilatation of celiac artery. Hypertrophy of MAL and diaphragmatic crura caused indentation on the aorta [Table/Fig-5a,b]. There was abnormal dilatation of large bowel loop with a large intraluminal polypoidal mass near distal colon. With theses imaging findings, diagnosis of large bowel obstruction secondary to recurrent distal colonic polypoidal mass with a additional finding of CACS was made. The patient was advised surgical resection of colonic mass. Follow-up of this patient could not be done.



[Table/Fig-5]: Contrast-enhanced MDCT of the abdomen of case 5: (a) Sagittal section showing thickened median arcuate ligament (short white arrow) causing focal extrinsic compression of anterior wall and kinking of the proximal celiac artery with posterior displacement of celiac artery and indentation on the aorta. (b) Axial section showing thickened median arcuate ligament showing stenosis of the proximal celiac artery (arrow).
C.A: Celiac artery; AO: Aorta; MAL: Median arcuate ligament

DISCUSSION

The median arcuate ligament is a fibrous bridge that connects the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes cranial to the origin of the celiac axis but in 10%-24% of people, the ligament may cross anterior to the artery. In a few of these individuals, the ligament may compress the celiac axis and compromise blood flow which results in causing symptoms [6]. The common complaint of patients with MAL syndrome is vague intermittent abdominal pain, typically epigastric and usually postprandial with associated symptoms including nausea and diarrhea [7]. The main pathophysiology of CACS is foregut ischemia and vascular steal phenomenon with splanchnic vasoconstriction resulting from compression of the celiac artery. Few authors believe pain may be due to the compression of the celiac plexus and ganglion [5].

The different phases of respiration are important factors to be considered in these patients. During inspiration, coeliac artery migrates inferiorly in the abdominal cavity attaining a vertical orientation and thereby compression gets relieved. But during expiration, the celiac artery gets compressed and narrowed [2,8]. On clinical examination, there may be mild epigastric tenderness, with an occasional finding of a midabdominal systolic bruit on auscultation [7]. The compromised blood flow by compression of the celiac artery by the median arcuate ligament causes the symptom. However, some patients are asymptomatic due to sufficient collateral supply from superior mesenteric circulation [1].

The diaphragmatic crura normally arise from the anterior surface of the L1 to L4 vertebral bodies on the right and the first 2 or 3 lumbar vertebral bodies on the left. At the level of the first lumbar vertebra, the median arcuate ligament passes anterior to the aortic hiatus and unites both the diaphragmatic crura [9]. The exact aetiology of compression is not known; however, two anatomic variants like lower insertion of MAL or higher origin of a celiac artery can predispose to CACS [4,9]. Initially, this condition was diagnosed by using conventional angiography but the use of thin-section MDCT and of 3-Dimensional (3D) image reconstruction has greatly improved the ability to obtain detailed images and helped us to assess the abdominal vascular system non invasively [6].

In addition to routine axial MDCT angiography, sagittal plane reconstruction plays an important role in optimal visualisation of the proximal portion of the celiac axis. The characteristic imaging findings seen in patients with celiac artery compression include abnormal thickening of median arcuate ligament more than 4 mm, hypertrophy of diaphragmatic crura, focal narrowing in the proximal celiac axis with a hooked appearance, post-stenotic dilatation of celiac artery, kinking or posterior/inferior displacement of celiac and superior mesenteric arteries and presence of collateral vessels [2,4,7-10].

In study done by Ilica A et al., all patients showed proximal smooth narrowing of the artery, hooked appearance, kinking of the celiac artery and also they classified the arterial stenosis into four grades [9]:

- Grade I- 0% to 49% diameter reduction as mild stenosis;
- Grade II- 50% to 74% diameter reduction as moderate stenosis;
- Grade III -75% to 99% diameter reduction as severe stenosis; and
- Grade IV-total occlusion

The present study also showed smooth narrowing of the proximal artery and hooked appearance due to kinking of the celiac artery, posterior and inferior displacement of the celiac artery in all patients and two patients showed indentation on the aorta. The stenosis of the celiac artery was grade III in three patients and grade II in two patients.

Sharma PK et al., in their study, described thickness of MAL >4 mm as abnormal and all of their patients showed thickening of median arcuate ligament measuring 4 mm, focal extrinsic compression of the anterior wall of the proximal celiac artery, poststenotic segment dilatation and hooked appearance [4]. In present case series, all patients showed extrinsic compression of the proximal celiac artery, poststenotic dilatation of artery, hooked appearance and thickening of MAL. The thickness of the median arcuate ligament was more than 4mm in all patients of present study. Göyaa C et al., in their study, described hypertrophy of MAL, narrowing of the celiac trunk, and hooked appearance [8]. In present case series, all the patients showed similar MDCT finding. In Başkan et al case report study, acute angulation and narrowing of the proximal celiac artery with a classical hooked appearance was found [11] and present study also showed similar MDCT findings.

In the present case series few limitations were noted- MDCT was done under full inspiration; hence changes in different phases of respiration could not be demonstrated, Doppler ultrasound was not done; hence a dynamic evaluation of the compressed segment of the celiac artery could not be assessed, Few of the patients had come on out patient basis; hence follow-up could not be done. [Table/Fig-6] shows comparison of characteristics MDCT findings of CACS described by other reported studies [2,4,8,9].

MDCT findings of CACS	Ilica et al., 2007 Turkey [9] (n=8)	Sharma et al., 2022 Tamil Nadu, India [4] (n=5)	Göyaa et al., 2013 Turkey [8] (n=3)	Kamal V et al., 2017 Pune, India [2] (n=1)	Present study, 2022 (n=5)
Hypertrophied/ Thickened MAL	Present	Present	Present	Present	Present
Focal narrowing at proximal celiac artery	Present	Present	Present	Present	Present
Post-stenotic dilatation	-	Present	Present	Present	Present
Hooked appearance	Present	Present	Present	Present	Present
Posterior and inferior displacement of celiac artery	Present	-	-	-	Present
Indentation of aorta	Present	-	-	-	Present

[Table/Fig-6]: Comparison of characteristics MDCT findings of CACS described by other reported studies.

CONCLUSION(S)

CACS is a rare condition caused by the compression of the celiac trunk externally by the median arcuate ligament. In present case series, smooth narrowing of the proximal artery, hooked appearance due to kinking of the celiac artery and posteroinferior displacement of the celiac artery had been showed in all five patients . However two patients showed indentation on the aorta. The MDCT is an ideal modality for early diagnosis of median arcuate ligament syndrome non invasively.

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