

# Median Arcuate Ligament Syndrome as A Rare Cause of Recurrent Chronic Abdominal Pain: Radiological Findings

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**Background:** Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome, is characterized by gastrointestinal ischemia secondary to the compression of the median arcuate ligament of the diaphragm to the proximal celiac trunk.

**Case Presentation:** Herein, we aimed to report the Color Doppler Ultrasonography (CDUS) and Computerized Tomography (CT) findings of MALS in a 27-year-old man presenting with complaints of abdominal pain and slimming.

**Conclusion:** We think that MALS should be considered in young patients with persistent abdominal pain who are admitted with chronic epigastric pain. Non-invasive methods such as CDUS and CT should be used for diagnosis.

**Keywords:** Abdominal pain, doppler ultrasonography, computed tomography, median arcuate ligament syndrome

## Introduction

Median arcuate ligament syndrome (MALS) is also called celiac artery compression syndrome or Dunbar syndrome. The most common clinical finding, upper abdominal angina, is due to the compression of the celiac artery trunk with diaphragmatic crura. The typical age is 20-40 years. Lipshutz first reported it in 1917. MALS was described by Dunbar et al. in 1965 as Dunbar syndrome (1-3). Herein, we aimed to present Multi-Slice Computed Tomography (MSCT) and Colour Doppler Ultrasonography (CDUS) findings in a case with MALS.

## Case Presentation

A 27-year-old man presented with worsening postprandial abdominal pain progressively and weight loss for six months. He also had nausea and vomiting. Physical examination was normal except epigastric pain. His history was unremarkable for any former disorder. His electrocardiographic, abdominal, biochemical, and thoracic plain radiographic examinations were all in normal limits. He was examined with CDUS examination of the abdominal aorta and its branches, which revealed stenosis of the celiac artery with left-sided preponderance. In

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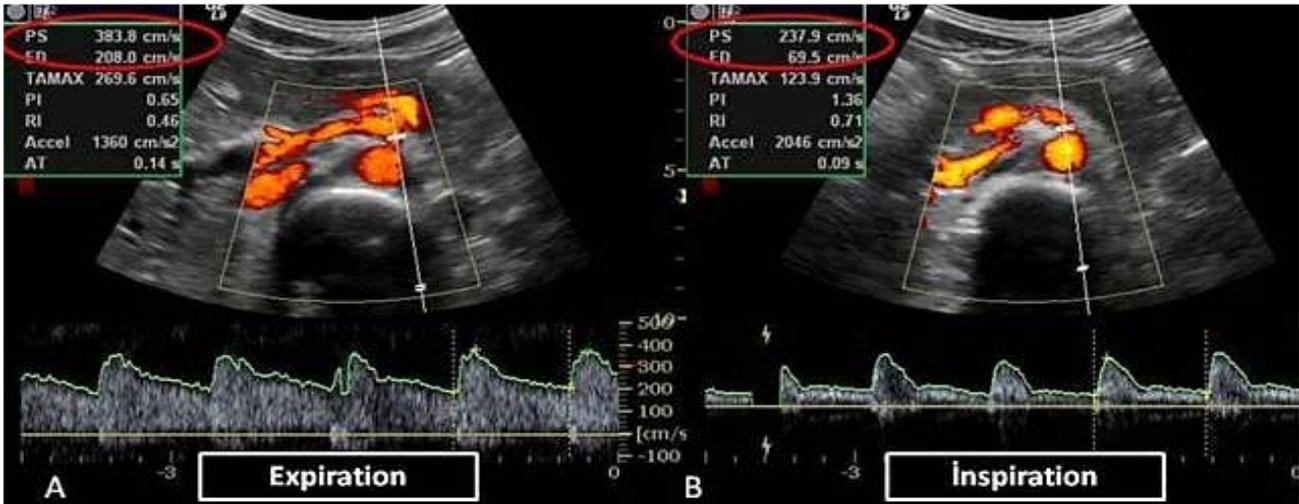


Figure 1. Doppler US findings at proximal celiac trunk level. (A) During expiration, peak systolic and end-diastolic flow rates are increased (red circle), and (B) during inspiration, return to normal at peak systolic, and end-diastolic flow rates are observed (red circle)

CDUS analysis, the peak systolic and end-diastolic velocities of the celiac artery at end-expiration were 383 cm/sec and 208 cm/sec. In maximum inspiration, the corresponding velocities were 237 cm/sec and 69 cm/sec (Figure’s 1A, B). CDUS findings suggest the diagnosis of MALS. However, to confirm the diagnosis, an angiographic examination with MSCT was performed. 128-slice MSCT (Aquilion, Toshiba Medical Systems Co., Tokyo,

Japan) imaging was performed after the administration of intravenous contrast medium. Multi-planar reconstruction and three-dimensional (3D) volume-rendered (VR) images were obtained from CT images. On CT images, the latter revealed a focal narrowing of a typical shape of a hook and post-stenotic dilation (Figure’s 2A, B), which secured the diagnosis of MALS. Conservative treatment with radiological follow-up was commenced with watchful waiting for symptom progression.

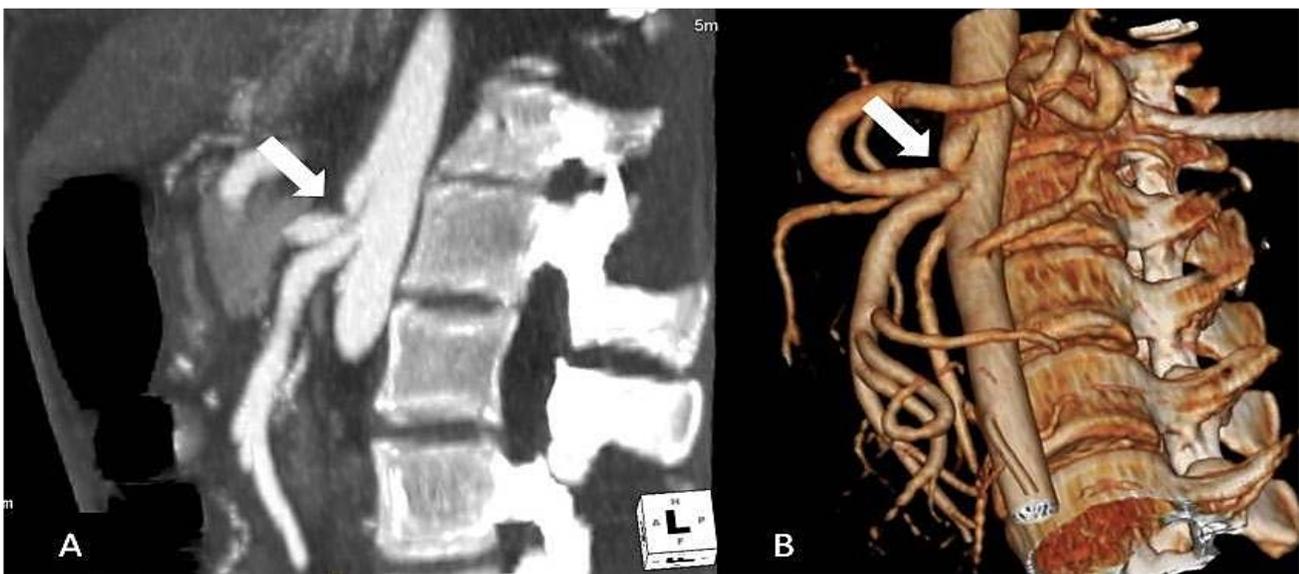


Figure 2. (A) In the sagittal maximum intensity projection image of CT angiography, and (B) 3D-CT angiography; the proximal part of the celiac artery shows a marked narrowing leading to a hook-like appearance (white arrows)

## Discussion

The median arcuate ligament is a fibrous structure that connects the diaphragmatic crura on both sides of the aortic hiatus and generally passes over the celiac trunk. The median arcuate ligament is localized at the T<sub>12</sub>-L<sub>1</sub> level. In some persons who suffer the condition named the median arcuate ligament syndrome, this ligament inserts at a lower level that is from the proximal part of the celiac trunk, compressing the latter. This condition results in ischemia of the gastrointestinal system. One of the main symptoms of the latter is postprandial abdominal pain. MALS is characterized by nausea, epigastric pain, weight loss, and postprandial pain. Postprandial pain primarily occurs during expiration. MALS is much more common in patients with postural orthostatic tachycardia syndrome. The reason for this is not yet understood (1,2). In a study that retrospectively examined the aortograms of 1500 patients, symptomatic celiac artery compression was identified in 1% of patients (3). However, the incidence of MALS in general population is not fully known. MALS is more common in women than in men, and most commonly occurs between 20 and 40 years old.

Two theories have been advanced for the occurrence of clinical signs and symptoms. The first theory is characterized by mesenteric ischemia due to the compression of the celiac artery. The second theory hypothesizes the irritation of somatic pain fibers secondary to neurological stimulation due to condensation on the celiac ganglion and plexus. (4,5). The diagnosis of MALS can be examined by CDUS, CT-angiography, catheter angiography and magnetic resonance angiography. CDUS has been reported to confer a high sensitivity for MALS and has been recommended as the

diagnostic modality of choice (6). The diagnosis of MALS can be made by Doppler flow measurements of the celiac artery and during inspiration and expiration. In Doppler flow velocity measurements in the compressed segment of the celiac artery, a change in peak systolic velocity of more than 200 cm/s during expiration is considered to be diagnostic (7). Thanks to advances in MSCT technology, MALS can be diagnosed even using routine contrast-enhanced abdominal CT. CT-angiography using MSCT characteristically demonstrates focal narrowing of the proximal celiac artery. The focal narrowing is characteristically seen as a hook. This sign may be beneficial for differentiating the condition from other conditions like atherosclerosis that may cause celiac artery stenosis. Post-stenotic dilation and collateral vessels are the other MSCT findings. The characteristic hook sign and post-stenotic dilation were present in our case. MSCT is a minimally invasive technique which helps in the diagnosis of MALS and may be preferred as an alternative to catheter angiographic procedures (4-6).

In the differential diagnosis of MALS, superior mesenteric artery compression syndrome (SMA syndrome, or Wilkie syndrome) and Nutcracker Syndrome should be considered as a priority. In particular, SMA syndrome can be easily confused with MALS due to the similarity of clinical findings. The pain in the SMA syndrome decreases in the position of the prone or left lateral decubitus. As in the diagnosis of MALS, CT is the most appropriate diagnostic method in the diagnosis of SMA syndrome (4,5,8,9). The treatment options for MALS include operative resection of the median arcuate ligament, celiac ganglion destruction, and vascular bypass surgery (10).

In conclusion, MALS is a rare and challenging diagnosis that should be considered in patients presenting with non-specific chronic abdominal pain. Doppler US with a dynamic evaluation during inspiration-expiration and in the supine as well as upright position is very useful in the diagnostic workup of suspected MALS. Confirmatory CT and CT-angiography should usually be obtained before treatment.

### Conflict of Interests

The authors declare that they have no conflict of interest in the current study.

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