

## The investigation of incidence and multidetector computed tomography findings of median arcuate ligament syndrome

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**Background/aim:** This study aims to evaluate the incidence and multidetector computed tomography (MDCT) findings of median arcuate ligament syndrome (MALS).

**Materials and methods:** Between January 2010 and September 2016 a total of 4028 patients who underwent angiography for MDCT based on the clinical indications for abdominal pain were retrospectively analyzed.

**Results:** The typical signs of MALS were identified in a total of 141 (3.5%) patients. Among all patients, 84 (59%) were male and 57 (41%) were female. The mean age was 42 years (range: 24 to 71 years). The incidence of celiac artery stenosis was 30% to 50% in 45 patients (32%) and 51% to 90% in 96 patients (68%). Poststenotic dilatation was reported in 74 patients (52%). Collateral vascular structures from the superior mesenteric artery were observed in 5 patients (3.5%). Twenty-one patients (14.9%) underwent laparoscopic surgery with significant stenosis.

**Conclusion:** MALS is a rare vascular pathology. It is especially important to recognize the presence of MALS with an epigastric pain that increases with expiration in adults and also its etiology before interventional procedures. MDCT visualization made the diagnosis of MALS easy.

**Key words:** Median arcuate ligament, celiac artery, multidetector computed tomography

### 1. Introduction

Median arcuate ligament syndrome (MALS) is a clinical condition that develops secondary to compression of the celiac trunk, particularly during expiration, where the celiac artery separates from the aorta, and it is characterized by gastrointestinal ischemia. It is also known as celiac artery compression syndrome or Dunbar syndrome (1,2). Clinical signs that occur following mesenteric ischemia according to the degree of compression include postprandial abdominal pain, nausea, vomiting, and weight loss.

Color Doppler ultrasonography (CDUS), multidetector computed tomography (MDCT), magnetic resonance imaging (MRI), and conventional angiography may be used in the diagnosis of MALS (3,4). Radiological diagnosis of MALS is made when the median arcuate ligament (MAL) compresses the proximal portion of the celiac artery, the celiac artery is displaced inferiorly, and it has a characteristic hook appearance (5). Poststenotic dilatation and collateral vascularization from the superior mesenteric artery may occur. False positive diagnosis of

the condition is reduced as imaging is made during deep expiration, when compression is greater, since the degree of compression of the celiac artery changes with the position of the diaphragm (6). The classical treatment of MALS is done by surgery, which involves cutting the MAL. It is performed by open surgery or through laparoscopy (7).

In this study, our aim was to evaluate the incidence and MDCT findings of MALS secondary to compression of the proximal portion of the celiac trunk by the median arcuate ligament, characterized by gastrointestinal ischemia.

### 2. Materials and methods

This study was conducted following approval from the noninterventional clinical research ethics committee from the institutional ethics committee. Written informed consent was obtained from each patient. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Between January 2010 and September 2016 a total of 4028 patients who underwent angiography for MDCT

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based on the clinical indications for abdominal pain were retrospectively analyzed. A total of 141 patients who presented to our department with complaints of abdominal pain and who were radiologically diagnosed with MALS following MDCT examination with angiography were included in the study. All the patients were examined by MDCT using a Toshiba Aquilion 64 slice CT scanner (Toshiba Medical Systems, Tokyo, Japan). The scanning area was identified between the diaphragm and the iliac crest. Images were of kVp 120, mAs 150–200 value, 0.5 mm collimated cross-section thickness, 0.3 mm reconstruction interval, diameter FOV (30 cm), and a pitch value between 1 and 1.5. Investigations were initiated 1 h before the examination every 15 min, following oral consumption of 1000–1500 mL of water. All examinations were performed with the patients in the supine position and automatic injection of 100 mL of iopromide or 100 mL of iohexol at a rate of 3 mL/s through the right antecubital vein, through single breath-holding at 65 s.

The MDCT images were transferred to the study center (VITAL, Vitrea 2, HP XW6400 Workstation, USA). The images were evaluated as multiplane and three-dimensional (3D) angiographic images. Every patient's images were evaluated with respect to rates of celiac artery stenosis, poststenotic dilatation, possible collateral vascular formation, and pathological findings of gastrointestinal structure present within the investigation area, and the results were registered.

### 3. Results

Among the patients enrolled in the study, 84 (59%) were male and 57 (41%) female. The mean age was 42 years (range: 24 to 71 years). Using the reformatted images obtained particularly in the sagittal plane and MDCT arterial angiographic examination, 141 patients (3.5%) with typical MALS findings were identified following proximal compression of the celiac trunk by the MAL and visualization of inferior displacement of the celiac artery and the characteristic hook appearance (Figures 1a–1c). The incidence of celiac artery stenosis was reported as 30% to 50% in 45 patients (32%) and as 51% to 90% in 96 patients (68%). Poststenotic dilatation was found in 74 patients (52%) (Figure 2). Nonsignificant stenotic atherosclerotic calcified plaques were observed in 58 patients (41%) where the celiac artery separates from the aorta. Collateral vascular structures from the superior mesenteric artery were observed in 5 patients (3.5%) (Figure 3). Radiological findings of the patients are shown in Table 1.

Abdominal ultrasonography (US) results of 43 patients were recorded; however, no significant clinical findings were reported. Although Doppler US was reported as

highly sensitive in the diagnosis and is accepted as a modality for diagnosis of vascular pathology, three of our patients were reported to have no celiac artery pathology by Doppler US examination.

A total of 47 patients (33%) presented with complaints of epigastric pain that particularly developed after meals. Nausea, vomiting, and diarrhea were reported in 23 patients (16%), whereas there was weight loss in 8 patients (5.6%). Clinical complaints of the patients are shown in Table 2.

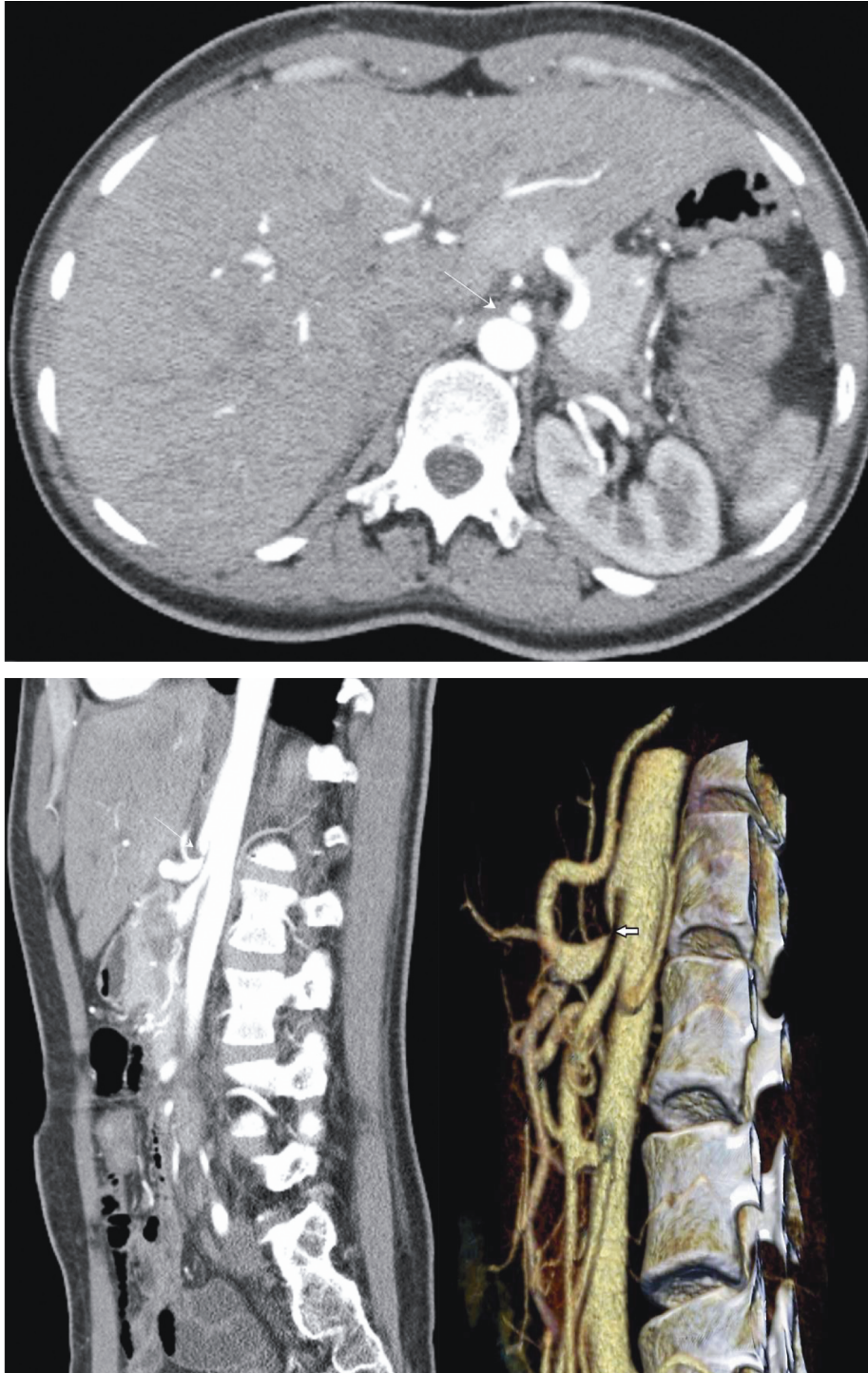
Twenty-one patients (14.9%) underwent laparoscopic surgery with significant stenosis identified in their follow-up MDCT angiography. No recurrence was reported during the 1-year follow-up visits of the 21 patients who underwent surgical operation.

### 4. Discussion

MALS is a rare vascular pathology that develops secondary to proximal compression of the celiac artery by the MAL, and it is characterized by clinical findings such as postprandial abdominal pain, nausea, vomiting, and weight loss. The MAL is formed by the union of the right and left diaphragmatic crura anterior to the descending aorta. In patients, the MAL compresses the celiac artery while passing in front of the aorta. The increased complaints during expiration may be explained by significant compression of the celiac artery following cranial movement of the diaphragm. Exercise-associated abdominal pain may be reported in severe cases (8). However, some patients are reported to be asymptomatic, despite severe stenosis of the celiac artery. This condition may be attributed to the development of collateral structures of the celiac artery or its branches from the superior mesenteric artery. The most common collateral development is from the gastroduodenal artery (9).

Marked epigastric murmur observed on expiration during physical examination in patients who present with complaints of postprandial epigastric pain and weight loss is a significant sign in the diagnosis of MALS (10). The clinical symptoms are nonspecific and may easily be confused with cases of peptic ulcer and functional dyspepsia. Furthermore, the differential diagnosis should be considered with superior mesenteric artery syndrome, which presents with similar symptoms. It develops secondary to compression of the third portion of the duodenum by the superior mesenteric artery. Literature studies have reported concomitant cases (11).

It is especially important to recognize the presence of MALS and its etiology before interventional procedures (12). In the diagnosis of MALS, Doppler US, MDCT, MRI angiography, and conventional angiography may be helpful (3,4,13). Doppler US has been reported as sensitive



**Figure 1.** Enhanced abdominopelvic MDCT revealed a focal narrowing in the proximal coeliac artery with excessive fibers arising from the diaphragm in axial (a) and reconstructed sagittal views (arrow) (b). Three-dimensional reconstructed MDCT confirmed the diagnosis of coeliac artery compression (arrow) (c).

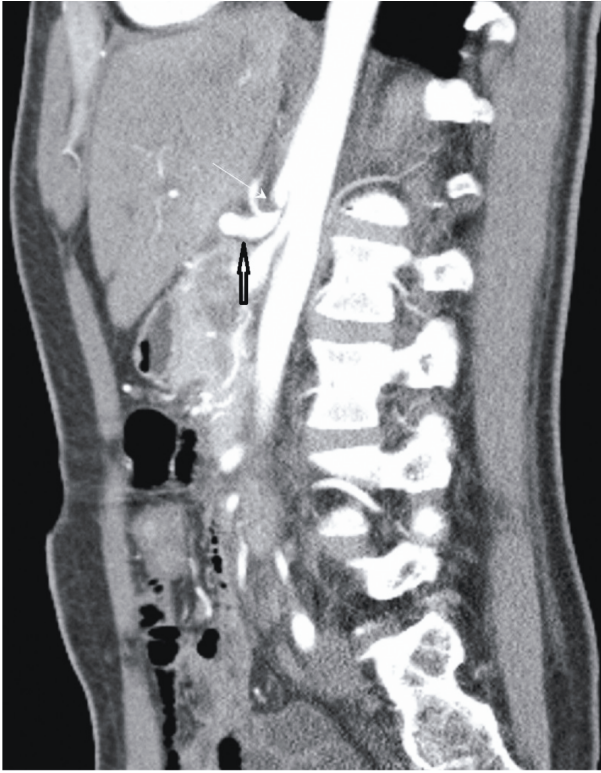


Figure 2. Enhanced abdominal reconstructed sagittal MDCT image shows poststenotic dilatation (black arrow).



Figure 3. Coronal oblique MDCT angiogram reveals a collateral vessel and dilatation of the gastroduodenal artery (arrow).

Table 1. Stenosis rates obtained as a result of radiological investigations.

Celiac artery stenosis	n: 45 (32%)	Stenosis: 30%–50%
	n: 96 (68%)	Stenosis: 51%–90%
Poststenotic dilatation	n: 74 (52%)	
Collateral vascular structures	n: 5 (3.5%)	

Table 2. Incidence of clinical symptoms.

Postprandial epigastric pain	n: 47 (33%)
Nausea, vomiting, diarrhea	n: 23 (16%)
Weight loss	n: 8 (5.6%)

in the diagnosis and is accepted as a diagnostic modality (13). In our study, 23 patients (16%) were subjected to Doppler US and hemodynamically significant stenosis was reported in the vascular structure. Selective angiography is considered as the gold standard in the diagnosis. Images can be obtained during inspiration and expiration (14). Angiography is an invasive diagnostic procedure.

Noninvasive MDCT is currently used in many centers for diagnosis. It is particularly useful for sagittal reformatted visualization of this syndrome. Sagittal reformatted MDCT demonstrates proximal celiac stenosis, poststenotic dilatation, and a characteristic hook appearance (hook-shaped contour) of the celiac artery. The appearance of pancreaticoduodenal collateral vascular structures has been reported in patients with MALS. The hook appearance can be helpful in the differential diagnosis of other pathologies of the celiac artery and atherosclerosis from the condition (5,15). Atherosclerotic stenosis occurs at the orifice of the celiac artery, while the median arcuate ligament compression occurs 5 mm from the base. Furthermore, images obtained from all imaging modalities should correlate with clinical symptoms (16,17).

The incidence of gastrointestinal symptoms such as abdominal pain, nausea, vomiting, diarrhea, and constipation is not more than in the overall population. In this study gastrointestinal symptoms were described in 53 patients (37%). The main goal of treatment in patients diagnosed with MALS is MAL decompression and maintenance of normal celiac artery blood flow. Cutting the MAL by open or laparoscopic surgery reduces the morbidity and mortality rates and these are conventional procedures with high success rates (18). On the other hand, different treatment modalities such as

patch angioplasty, aortoceliac bypass, aortoceliac artery reanastomosis, percutaneous endovascular interventions, balloon dilatation, stent placement, and endoscopic retroperitoneal approach may be instituted in the absence of conventional methods (19–22). Prolonged follow-ups after surgical treatment have provided a success incidence rate of 70% to 80% (14).

The limitation of this study is that, in patients with MALS, the celiac artery is compressed by the MAL with expiration. With inspiration, the celiac artery descends in the abdominal cavity, resulting in a more vertical

orientation of the celiac artery, which often relieves the compression.

In conclusion, MALS is a rare vascular pathology. MDCT has provided rapid, small, cross-sectional, multiplane reformatted, and 3D angiographic images, making noninvasive procedures preferable for the diagnosis. It is important to recognize the presence of MALS before interventional procedures. Since there is a possibility of postoperative recurrence, patients should be monitored after surgery.

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