

# Median arcuate ligament syndrome, a rare case of chronic abdominal pain

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## ABSTRACT

The median arcuate ligament syndrome (MALS) is a rare disease characterized by abdominal pain caused by the external compression of the celiac artery by the median arcuate ligament. Surgical treatment is indicated, but given the non-specific symptoms, these patients are often hospitalized in the Departments of Internal Medicine where the diagnosis may be unknown. We present a case of MALS admitted to our Internal Medicine Division. An abdominal ultrasound in a woman with longstanding abdominal pain showed elevated celiac artery velocities during forced expiration. Computed tomography angiography (CTA) of the abdomen showed stenosis of the origin of the celiac artery and confirmed the diagnosis of MALS. MALS is a syndrome that has to be considered, especially in young women with abdominal pain of unclear etiology; evaluated by color Doppler ultrasound, in the presence of elevated hepatic artery velocities during forced expiration, the confirmatory test is CTA.

## Introduction

The median arcuate ligament syndrome (MALS) or Dunbar Syndrome is a rare disease caused by the external compression of the celiac trunk by the MAL and characterized by abdominal pain, nausea, vomiting, weight loss.<sup>1</sup>

The MAL passes superior to the origin of the

celiac artery; in 10% to 24% of the general population this fibrous arch is positioned lower, crossing over the proximal portion of the celiac axis, compressing it and some of these patients may have hemodynamically significant stenosis.<sup>1,2</sup>

Surgical treatment is indicated for symptomatic MALS, but given the non-specific symptoms, these patients are often hospitalized in the Departments of Internal Medicine where the diagnosis may be unknown.<sup>1</sup>

We present a case of a patient with MALS admitted to our Internal Medicine Division presenting with chronic abdominal pain and weight loss.

## Case Report

A 44-year-old woman presented to the emergency room with epigastric abdominal pain.

The patient reported a 2-year history of intermittent abdominal pain associated with nausea and weight loss. The medical checks carried out at that time were normal. She had rare diarrhea and she denied alcohol or drug abuse.

The physical examination revealed mild epigastric tenderness to palpation.

Laboratory tests (complete blood counts, liver function tests, serum amylase and lipase) and esophagogastroduodenoscopy were normal.

Abdominal ultrasound ruled out cholelithiasis whereas color Doppler ultrasound (CDUS) revealed increased celiac artery velocities during forced expiration (Figure 1A and B). Suspecting MALS, a computed tomography angiography (CTA) of the

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abdomen was performed, which showed stenosis of the origin of the celiac artery caused by the median arcuate ligament with its typical hook-shaped appearance (Figures 2 and 3). These findings confirmed the diagnosis of MALS.

The patient was referred to the surgeons for a six-month follow-up.

## Discussion

The median arcuate ligament syndrome or celiac axis compression syndrome, or Dunbar syndrome is a rare disease caused by the external compression of the celiac trunk by the median arcuate ligament.<sup>2</sup>

Given the non-specific symptoms and presentation, MALS is very difficult to be diagnosed and needs to be differentiated from other causes of abdominal pain, including biliary diseases and peptic ulcer diseases. Therefore, these patients are often hospitalized in the Departments of Internal Medicine where the diagnosis may be unknown.

This syndrome was described in 1963 by Harjola<sup>3</sup> and in 1965 by Dunbar *et al.*<sup>4</sup> and the definition is based on a combination of clinical and radiographic features. The majority of affected patients are asymptomatic due to sufficient collateral supply from superior mesenteric circulation and radiographic finding of celiac axis compression may not be significant. Severe compression occurs in approximately 1% of patients.<sup>3</sup>

The MALS's pathophysiologic mechanism is unknown but there are two main theories, mainly mesenteric ischemia and nerve dysfunction.<sup>5-7</sup>

Usually, the disease occurs in young women and is characterized by postprandial abdominal pain, weight loss, nausea, vomiting, bloating, diarrhea,

reduced appetite, and the presence of extrinsic celiac compression on vascular imaging.<sup>2,8</sup>

In the past, angiography was considered the gold standard in the diagnosis of MALS. Nowadays it has been substituted by new diagnostic tools.

A color Doppler ultrasound is a good screening tool; once suspected on CDUS, the diagnosis of celiac artery compression is made by CTA and magnetic resonance angiogram (MRA).<sup>9</sup>

The mesenteric ultrasound, performed during deep expiration, shows increased blood flow velocity in celiac artery narrowing that may normalize with inspiration or with standing (erect); the main indicators for MALS are the peak systolic velocity (PSV) of >350 cm/s during deep expiration and a deflection angle greater than 50°.<sup>2,10,11</sup>

Another criterion to diagnose MALS is a PSV ratio greater than 3:1 in the celiac artery during expiration compared with the PSV in the abdominal aorta under the diaphragm.<sup>12</sup>

Sagittal view of CTA identifies a characteristic focal narrowing of the celiac artery exacerbated during the expiratory phase;<sup>13</sup> the hooked appearance with the absence of arterial calcifications differentiate this condition from atherosclerotic disease, main causes of celiac artery narrowing.<sup>7,14</sup>

MRA can also be used in patients with allergy to iodinated contrast media and with results similar to CTA.

In our patient, CTA demonstrated focal narrowing of the celiac axis at the origin suggestive of MALS.

The treatment for symptomatic MALS patients is open surgery or laparoscopic division of the median arcuate ligament to relieve the compression<sup>2</sup> restoring normal blood flow in the celiac axis; percutaneous endovascular treatment is an alternative technique and may be considered in selected cases.<sup>15</sup>

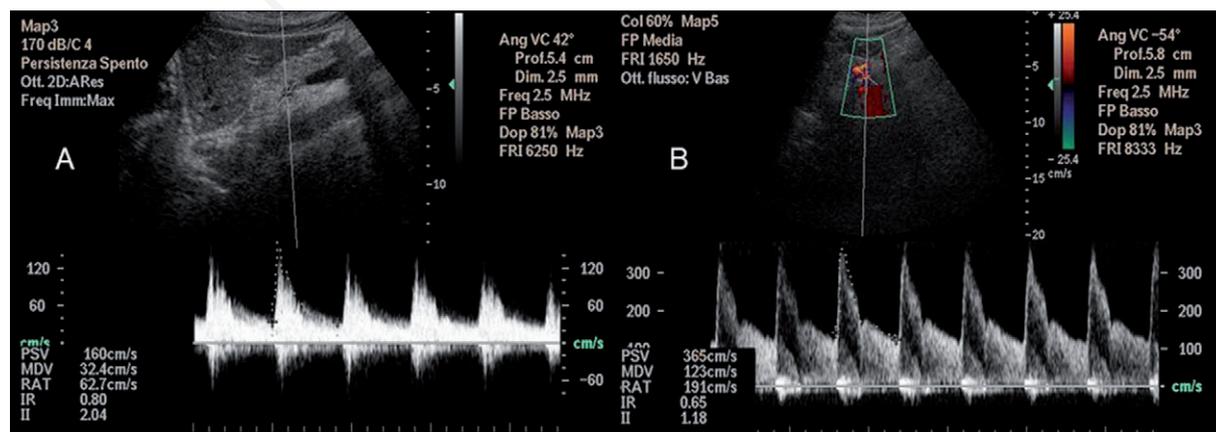


Figure 1. A) Doppler spectral analysis of the celiac artery in inspiration, flow with a peak systolic velocity of 365 cm/sec; B) color Doppler and spectral analysis of celiac artery during forced expiration, color Doppler shows flow with a peak systolic velocity of 365 cm/sec.

## Conclusions

Median arcuate ligament syndrome is a difficult diagnosis to obtain in the majority of patients due to its nonspecific symptoms. MALS is a diagnosis of exclusion and should be considered, when evaluating patients, especially young females, who present with abdominal pain of unclear etiology.

A color Doppler ultrasound to assess celiac artery velocities is a good screening tool for patients with

suspected median arcuate ligament syndrome. This diagnosis can be confirmed after performing a CTA.

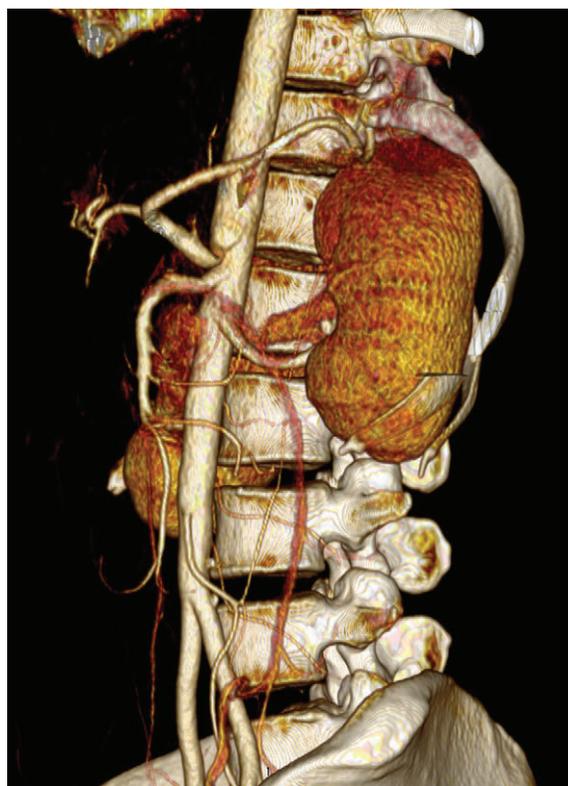
Surgical treatment is indicated for symptomatic MALS, but given the non-specific symptoms, these patients are often hospitalized in the Departments of Internal Medicine where the diagnosis may be unknown.

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**Figure 2.** Reconstruction computed tomography angiography SLAB 3D in sagittal plain shows stenosis of celiac artery caused by the median arcuate ligament with its typical hook-shaped appearance.



**Figure 3.** Reconstruction computed tomography angiography 3D volume rendering in sagittal plane: shows stenosis of celiac artery.

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