



## Case report

## Median arcuate ligament syndrome: A case report of a rare disease

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## ABSTRACT

**Introduction:** Median arcuate ligament syndrome (MALS) is a rare condition caused by the compression of the celiac trunk by the median arcuate ligament, which may trigger a typical symptom triad: postprandial abdominal pain, weight loss, nausea and vomiting.

**Clinical case:** A 58-year-old male was admitted to the emergency department due to abdominal pain, vomiting, fever and diarrhea. On admission, the patient was tachycardic, had abdominal distension, no peritoneal irritation signs, and a serum lactate level of 5 mmol/L. The computed tomography (CT) scan showed gastric and intestinal pneumatosis associated with pneumoportia. Conservative treatment with intravenous fluids, antibiotics and low molecular weight heparin, was initiated. The reassessment CT scan showed resolution of the gastric and intestinal pneumatosis and a severe stenosis of the initial portion of the celiac trunk with associated angulation. After recovery from the acute episode, an abdominal duplex ultrasound and an aortoiliac arteriography in forced inspiration and expiration was performed, corroborating the diagnosis of MALS. A laparoscopic decompression of the celiac trunk was performed.

**Discussion:** Symptoms of MALS closely mimic other abdominal disorders, and it should be considered in the differential diagnosis. Currently, there are no international guidelines on MALS diagnostic criteria. Treatment is focused on decompression of the median arcuate ligament constriction of the celiac artery, with or without celiac lymph node removal.

**Conclusion:** MALS diagnostic and therapeutic approach must be patient focused. Laparoscopic decompression is an effective treatment and can provide immediate symptomatic relief, associated with the benefits of the less invasive nature of the procedure.

## 1. Introduction

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome or Dunbar syndrome, is a rare phenomenon caused by compression of the celiac trunk by the median arcuate ligament (MAL) [1]. The MAL is an arch-like tendinous fascial band that joins the right and left diaphragmatic crura. This structure surrounds the aortic hiatus. The MAL is normally located cephalad to the origin of the celiac artery from the descending aorta. However, in an estimated 10 to 24% of individuals, the MAL extends more caudally, thereby enclosing the celiac artery and adjacent sympathetic nerve and ganglia, a phenomenon first described anatomically in cadavers by Lipschutz in 1917 [2]. The first clinical cases of MALS with angiographically visible

stenosis of the celiac trunk were described by Harjola and Dunbar and collaborators in 1963 and 1965, respectively.

Celiac artery lumen compression and narrowing, especially during expiration, can cause ischemia of the involved organs and may trigger a typical symptom triad: postprandial abdominal pain, weight loss, nausea and vomiting [3].

Controversy about MALS existence arises from its poorly understood pathophysiologic mechanism, variable presentation and therefore variable and unpredictable response to treatment. Accepted universal diagnostic criteria are lacking. Diagnosis remains one of exclusion and is typically the result of extensive investigations to exclude more common, alternative causes of abdominal pain [4].

Traditional surgical therapy consists on the division, through a

**Abbreviations:** MALS, median arcuate ligament syndrome; MAL, median arcuate ligament; CT, computed tomography; DUS, duplex ultrasound; CA, celiac artery; DSA, digital subtraction angiography; CTA, computed tomography angiography; MRA, magnetic resonance angiography.

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midline laparotomy, of the anomalous fibrous diaphragmatic bands overlying the celiac artery (CA), along with the removal of the celiac plexus and lymphatic tissue [5].

The aim of this report is to document a case of MALS treated with laparoscopic decompression. This case report was elaborated in accordance to SCARE criteria [6].

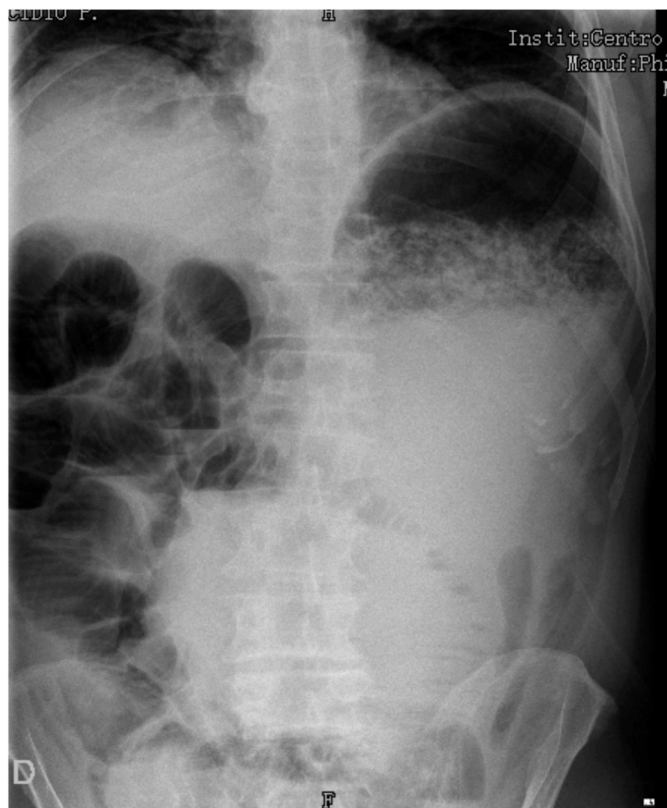
## 2. Clinical case

A 58-year-old male was admitted to the emergency department due to abdominal pain, nausea, vomiting, fever and diarrhea. He had a 2-year history of chronic and recurrent postprandial abdominal pain and weight loss. On admission, the patient was tachycardic, had abdominal distension, no peritoneal irritation signs and serum lactate level of 5 mmol/L. No other relevant laboratory results were obtained. The abdominal X-ray showed gastric distension and small bowel air-fluid levels (Fig. 1). Fluid resuscitation was started and a nasogastric tube placed. Drainage of an enteric content and an immediate relief of abdominal discomfort was observed. The computed tomography (CT) scan showed gastric and small bowel distension, gastric and intestinal pneumatosis, and pneumoportia (Fig. 2).

Given significant clinical improvement and regression of serum lactate levels, conservative management was implemented, with intravenous fluid therapy, broad spectrum antibiotics and low molecular weight heparin.

The patient remained hemodynamically stable, afebrile and completely asymptomatic. Repeated laboratory blood work showed a slight increase in inflammatory markers [leukocytes and C-reactive protein (CRP)], but without an increase in cellular ischemia/necrosis markers.

The reassessment Angio-CT scan showed a moderately distended stomach with slightly thickened walls, resolution of the gastric and intestinal pneumatosis, less distended small bowel loops, a severe stenosis



**Fig. 1.** Abdominal X-ray showed gastric distension and small bowel air-fluid levels.

of the initial portion of the coeliac trunk with associated angulation (median arcuate ligament syndrome), a permeable mesenteric artery, a mesenteric vein of normal caliber and a small quantity of free intraperitoneal fluid in the pelvis (Fig. 3).

After recovery from the acute episode, the patient was discharged, and an abdominal duplex ultrasound (DUS) was performed, which revealed an “extrinsic compression with hemodynamic significance (luminal reduction superior to 70%) of the celiac trunk”. An aortoiliac arteriography in forced inspiration and expiration revealed a “stenosis of the origin of the celiac trunk with forced expiration”.

After confirming MALS diagnosis and as the patient remained symptomatic (postprandial abdominal pain and weight loss), laparoscopic decompression was proposed.

The patient was submitted to a laparoscopic celiac trunk lymphadenectomy and arcuate ligament release (Fig. 4). No complications were recorded in the postoperative period and the patient was discharged on the second postoperative day.

The patient remained asymptomatic during a follow-up period of 3 years and had a weight gain of 8 kg.

Follow-up abdominal DUS showed “absence of hemodynamic changes of the celiac trunk, with inspiration and expiration, suggestive of stenosis”.

## 3. Discussion

Controversies still exist regarding the clinical features, pathophysiology, diagnosis and treatment of MALS. The observation of CA compression in asymptomatic patients makes us question the real incidence of the syndrome [5].

Skepticism is rooted by an unclear pathophysiologic mechanism and currently there are two theories that try to elucidate the symptoms associated with MALS. One commonly accepted theory suggests that the increased demand for blood flow through a compressed celiac artery leads to foregut ischemia resulting in epigastric pain. The development of collateral vessels usually prevents intestinal ischemia. Another hypothesis is that the pain associated with MALS has a neuropathic component, resulting from a combination of chronic compression and overstimulation of the celiac ganglion. This neuropathic compression may lead to a direct irritation and stimulation of sympathetic pain fibers and/or splanchnic vasoconstriction and ischemia. Additionally, vascular steal of blood flow, by larger collateral vessels, may lead to symptoms of celiac artery compression in patients with an occluded or compressed celiac trunk [3,7]. Both mechanisms may be involved [5].

Although the incidence of MALS is not well known, it is more prevalent in women (4:1 ratio) ages 30 to 50, and in patients with a thin body habitus [7].

It is most often characterized by chronic postprandial abdominal pain, nausea, vomiting, diarrhea, and unintentional weight loss. Pain location is variable but is most often in the epigastrium. Physical examination may reveal epigastric tenderness or a bruit that is amplified with expiration [4]. Because symptoms of MALS closely mimic those of other abdominal disorders, it is commonly considered an exclusion diagnosis.

Abdominal DUS imaging should be the first diagnostic approach when CA stenosis is suspected. DUS imaging during maximum inspiration and expiration can easily demonstrate the existence and the exact configuration of a dynamic CA stenosis [5]. DUS has advantages as an initial investigation, compared to angiography, as it is cheaper and noninvasive, and it does not expose patients to high doses of radiation [4].

If MALS is suspected, invasive digital subtraction angiography (DSA), non-invasive computed tomography angiography (CTA) or magnetic resonance angiography (MRA) can be used to verify the location of the celiac trunk [9].

CTA offers the advantage of 3-dimensional reconstruction and allows visualization of the compressed artery from different angles. Both CTA

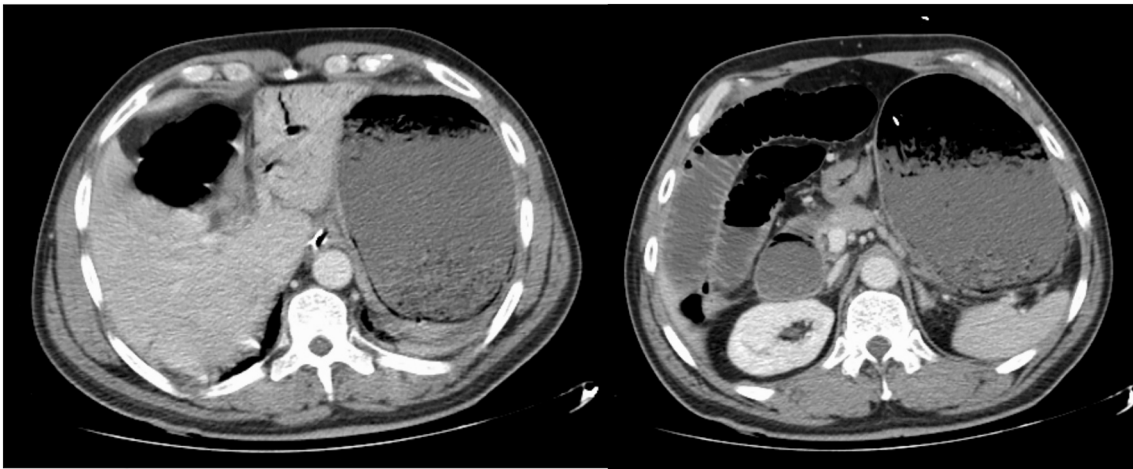


Fig. 2. Computed tomography showed gastric and small bowel distension, with signs of gastric and intestinal pneumatosis and portal vein gas.

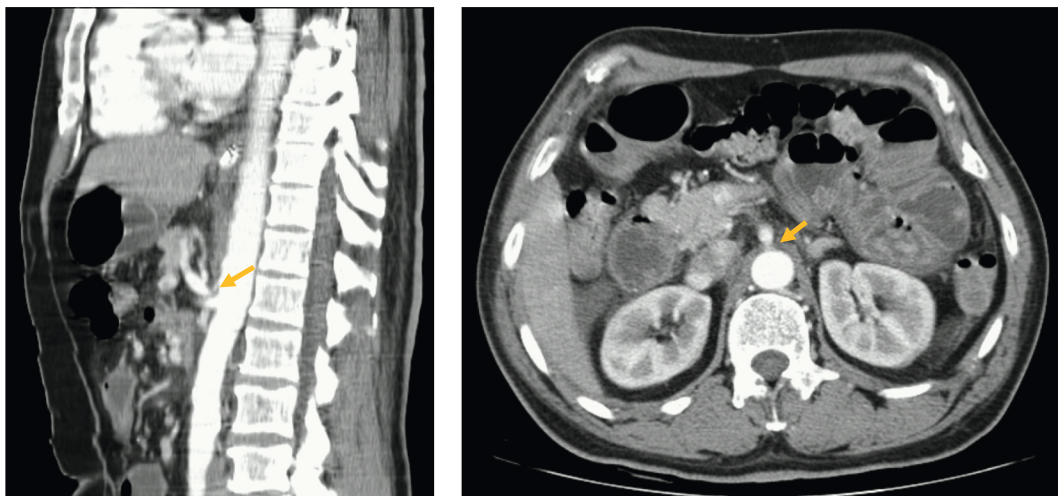


Fig. 3. Reassessment CT severe stenosis in the initial portion of the coeliac trunk with angulation (median arcuate ligament syndrome).

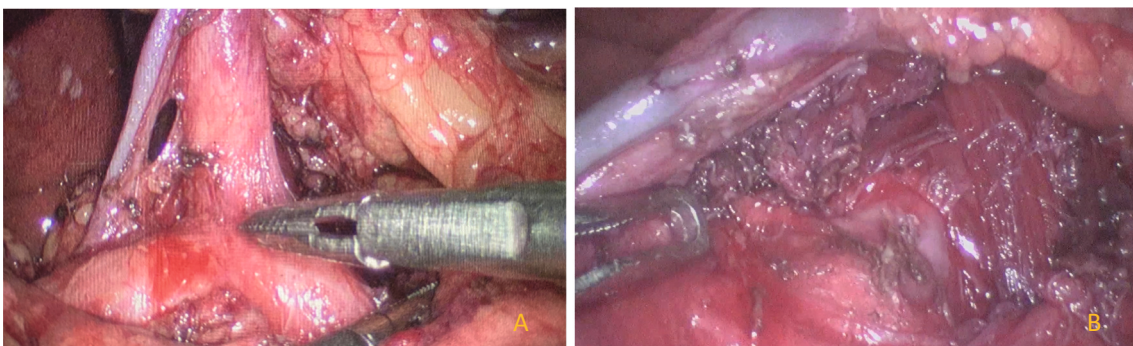


Fig. 4. The patient was submitted to lymphadenectomy of the celiac trunk (A) and release of arcuate ligament (B) by laparoscopy.

and MRA allow identification of concomitant abdominal pathology in addition to findings of MALS. MRA can be used in some patients with intravenous contrast allergy and provides results similar to those of CTA. Angiography with breathing manoeuvres is the diagnostic gold standard exam. It can be used to demonstrate celiac artery compression in MALS and the cephalad movement of the celiac axis during inspiration can reveal celiac artery compression and post stenotic dilatation on expiration [7].

Celiac artery decompression is indicated only for symptomatic patients with confirmed celiac artery compression on inspiratory and expiratory vascular imaging studies. Asymptomatic patients should be recommended surveillance of postprandial abdominal symptoms.

Interventions for MALS aim to address the hypothesized pathophysiologic mechanisms: decompression of the MAL's constriction of the celiac artery, with or without celiac lymph node removal, or to target the neuropathic component to the pain. Decompression of the celiac artery

was traditionally achieved through an open approach. Recent years, however, have seen a trend toward laparoscopic intervention [4]. The laparoscopic approach is becoming more widely adopted owing to benefits such as shorter hospital stay, decreased time to feeding, smaller risk of postoperative complications, decreased blood loss, greater postoperative pain relief and better cosmetic outcomes [1]. Persistent stenosis of the celiac artery after decompression may be seen with intraoperative handheld doppler assessment or angiography. If it persists, it can be addressed with arterial reconstruction, which include an aortoceliac bypass or a celiac artery patch angioplasty. Percutaneous transluminal angioplasty with or without stenting provides an adjunctive endovascular approach to revascularization of a persistent stenosis after MAL release. Endovascular intervention alone does not address the extrinsic compression of the celiac artery and has thus proven ineffective as an isolated treatment of MALS [4,5,7].

MALS prognosis is generally good, considering the high response rate to surgical decompression. The largest contemporary series report a symptom free index of 75% with a mean follow-up of 9 years [10].

#### 4. Conclusion

MALS is a rare, incompletely understood syndrome. Despite no general consensus on MALS diagnostic or management criteria, patient presentation and imagiologic signs appear consistent across the literature. MALS diagnostic and therapeutic approach must be patient focused. Laparoscopic decompression is an effective treatment for MALS and can provide immediate symptomatic relief, with the benefits of its less invasive nature.

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#### Ethical approval

Study exempt from ethical approval.

#### Consent

Studies on patients or volunteers require ethics committee approval and fully informed written consent which should be documented in the paper.

#### Author contribution

Joana Isabel Almeida did the conceptualization, data collection,

surgical therapy, and writing the original draft.

Margarida Nunes Coelho wrote the manuscript – review and editing

Isabel Armas wrote the manuscript – review and editing

Carlos Soares wrote the manuscript – review and editing

Tatiana Santos did surgical therapy for this patient and wrote the manuscript – review and editing

Carla Freitas did surgical therapy for this patient and wrote the manuscript – review and editing

#### Registration of research studies

Not applicable.

#### Guarantor

Joana Isabel Almeida.

#### Declaration of competing interest

The authors report no declarations of interest.

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