

**A RARE CASE OF MEDIAN ARCUATE LIGAMENT SYNDROME - A CASE REPORT**Dr. Akansha Singh<sup>1</sup>, Dr. Abhinav Kumar<sup>2</sup>, Dr. Cliffin Mathai Kattoor\*<sup>2</sup> and Dr. Utkarsh Bhardwaj<sup>2</sup><sup>1</sup>Junior Resident, Department of Anaesthesia and Critical Care, IGMC Shimla.<sup>2</sup>Junior Resident, Department of General Surgery, IGMC Shimla.**\*Corresponding Author: Dr. Cliffin Mathai Kattoor**

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**ABSTRACT**

Median Arcuate Ligament Syndrome (MALS) is a disease entity with unclear pathogenesis. This syndrome is also known by other names – Celiac axis syndrome, Celiac artery compression, Dunbar syndrome. It 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. However, the clinical diagnosis of the disease is very difficult because patients complain of digestive discomfort including pain which is not specific to this syndrome. Coeliac axis syndrome is considered a diagnosis of exclusion. We present the case of a 23-year-old male with median arcuate ligament syndrome that caused abdominal pain associated with nausea, emesis, and bloating.

**KEYWORDS:** Median Arcuate Ligament Syndrome (MALS), Celiac artery compression, postprandial abdominal pain.

**Abbreviations**

MALS – Median Arcuate Ligament Syndrome

CA – Coeliac Axis

DUS – Duplex Ultra Sonogram

CT – Computed Tomography

DSA- Digital subtraction angiography

CTA- Computed tomography angiography

MRA-Magnetic resonance angiography

**INTRODUCTION**

Median arcuate ligamentum syndrome (MALS) is a disease entity with unclear pathogenesis.<sup>[1,2]</sup> The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The MAL is normally located cephalad to the origin of the celiac artery from the descending aorta near the first lumbar vertebra. 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 Lipshutz in 1917.<sup>[3]</sup> This syndrome has an incidence of 2 cases per one lakh with diffuse and non specific abdominal pain. Median arcuate ligament syndrome (MALS) occurs when the arc-shaped band of tissue in the chest area (median arcuate ligament) presses on, or traps, the artery that supplies blood to the organs in your upper abdomen (celiac artery). 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

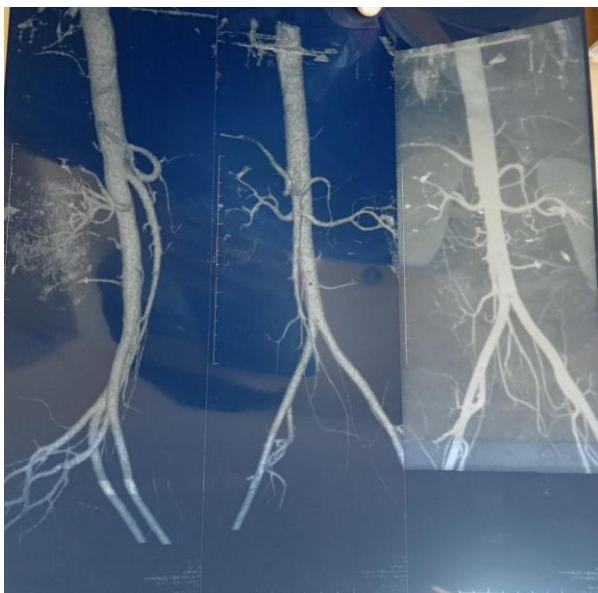
vomiting.<sup>[4]</sup> Controversy about the existence of MALS arises from its poorly understood pathophysiologic mechanism, variable presentation and therefore variable and unpredictable response to treatment. It is considered a diagnosis of exclusion and is typically the result of extensive investigations to exclude more common, alternative causes of abdominal pain.<sup>[1]</sup> Angiography with breathing maneuvers is the diagnostic gold standard exam. Laparoscopic decompression through division of the anomalous fibrous diaphragmatic bands overlying the celiac artery (CA), along with the removal of the celiac plexus and lymphatic tissue is an effective treatment with immediate symptomatic relief.<sup>5</sup> The aim of this report is to document a case of MALS managed conservatively.

**CASE REPORT**

A 23-year-old male with no significant past medical history presented to the emergency department with a history of intermittent epigastric abdominal pain since 2 days. The pain was associated with nausea, multiple episodes of nonbilious emesis, and bloating. The pain became worse on eating fatty foods; the nausea worsened with any oral intake and relieved with bowel rest. There was no history of associated diarrhea and he also denied any radiation of the pain to any other region. On admission, the patient was tachycardic and had abdominal distension. His physical examination revealed epigastric tenderness to palpation but no other abnormalities. Bowel sounds were heard normally and a per rectal examination revealed normal findings. On

blood investigations, serum sodium was 130 mmol/L, potassium was 3.2 mmol/L and chloride was 96 mmol/L. Serum Lactate was elevated. Rest of the laboratory investigations were normal. Fluid resuscitation was started and a nasogastric tube placed. Drainage of an enteric content and an immediate relief of abdominal discomfort was observed. Abdominal X-ray showed no evidence of dilated bowel loops or air fluid levels. CT-Angiography was performed in view of diagnostic

uncertainty, which showed splenic and common hepatic artery arising directly from abdominal aorta with mild focal narrowing at the distance of 1cm from the origin with a normal caliber distally. As a result of the radiological findings along with clinical symptoms, a diagnosis of MALS was made. The patient was managed conservatively. He symptomatically improved within a few days and was discharged.



**Fig. 1: CT Angio was done showing common origin of coeliac and splenic artery from aorta with focal narrowing.**

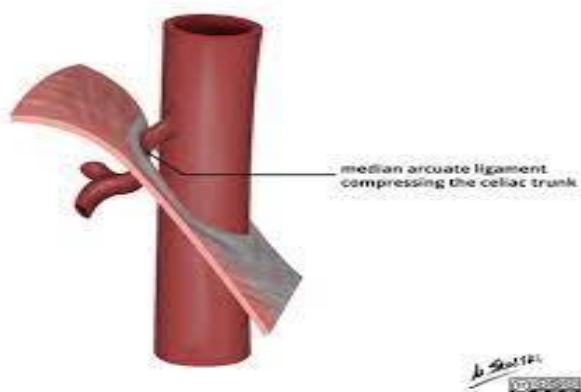
## DISCUSSION

Median arcuate ligament syndrome (also known as Dunbar syndrome or celiac artery compression syndrome) was first described by Harjola in 1963.<sup>6</sup> It occurs when the arc-shaped band of tissue in the chest area (median arcuate ligament) presses on, or traps, the artery that supplies blood to the organs in your upper abdomen (celiac artery). The location of the median arcuate ligament and celiac artery varies slightly from person to person. Typically, the ligament runs across the largest blood vessel in the body (aorta) and sits above the

celiac artery without causing problems. But sometimes the ligament or artery may be out of place, causing MALS.

The ligament may also put pressure on the network of nerves surrounding the celiac artery (celiac plexus).

Although the incidence of MALS is not well known, it is more prevalent in women aged 30 to 50 years, and in patients with a thin body habitus.<sup>[2]</sup>



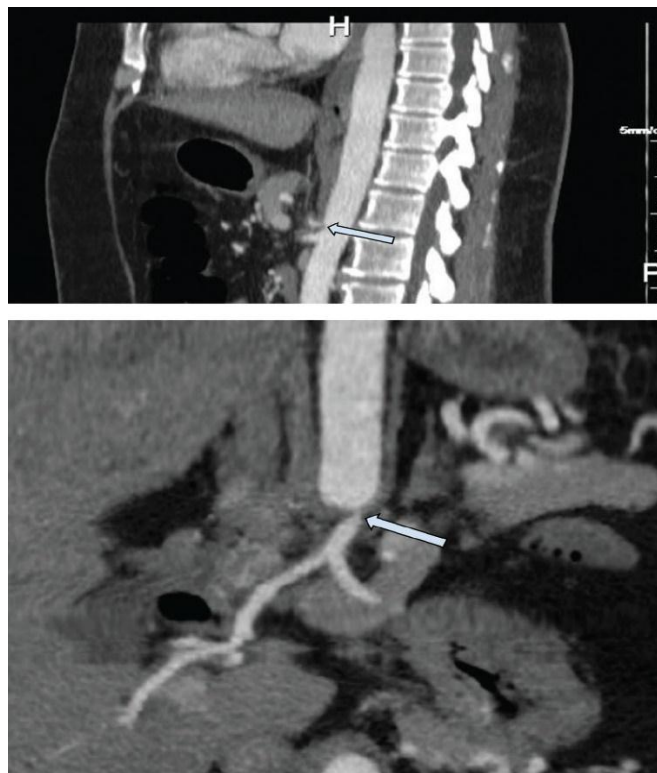
**Fig. 2: Diagrammatic representation of MALS.**

Theories regarding the pathophysiology of epigastric pain associated with MALS include foregut ischemia due to compressed celiac artery, midgut ischemia due to vascular steal syndrome, and overstimulation of the celiac plexus with subsequent splanchnic vasoconstriction and ischemia.

Recently, ideas about the etiology of MALS have shifted from its being a vascular disease to a neurogenic disorder with compression of the surrounding celiac plexus and ganglion.

Abdominal DUS (Duplex ultra sonogram) 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.<sup>5</sup> 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.<sup>[1]</sup>

If MALS is suspected, invasive DSA, non-invasive CTA or MRA can be used to verify the location of the celiac trunk.<sup>[7]</sup>



**Fig. 3: Contrast-enhanced computed tomographic angiography, sagittal (top) and 3-D coronal (bottom) views, showing severe stenosis at the origin of the celiac artery (arrow) with associated soft-tissue attenuation, suggestive of median arcuate ligament syndrome.**

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.

Laparoscopic release of the arcuate ligament has become a widely accepted treatment. Endovascular therapy may be necessary as well, given the possible recurrence of stenosis. Multidisciplinary assessment by a general surgeon, vascular surgeon, radiologist, and gastroenterologist is helpful.

Cienfuegos et al offered the following selection criteria for laparoscopic treatment: young woman, intense

postprandial pain, greater than 70% stenosis of the trunk, and development of collateral circulation.<sup>[8]</sup>

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.

MALS prognosis is generally good, considering the high response rate to surgical decompression.<sup>[9]</sup> However, it is a difficult diagnosis to obtain in a majority of patients which requires extensive workups. MALS diagnostic and therapeutic approach must be patient focused.

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