

## Median Arcuate Ligament Syndrome in A Patient with Possible Achalasia Presenting as Chest Pain: A Case Report

Louzon NE\* and Arnold WA

Universidad Autónoma de Guadalajara Medical School Avenida Patria 1201, Lomas del Valle, Zapopan, Jalisco, Mexico 45129, 110 Gallery Cir, San Antonio, TX 78258

### \*Corresponding author:

Nour E Louzon,  
Division of General Surgery, Abrazo Arrowhead  
Hospital, 18701 N 67th Ave, Glendale, AZ 85308,  
E-mail: nourlouzon@hotmail.com

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### 1. Abstract

Median arcuate ligament syndrome (MALS) is a rare syndrome presenting with nonspecific abdominal symptoms. We report a case of a 57-year-old woman with a stated history of achalasia who presented to the emergency department with chest pain. Due to her persistent chest pain despite negative cardiac workup, CT angiography (CTA) revealed proximal celiac artery narrowing with distal celiac artery dilation secondary to the median arcuate ligament (MAL) incidentally. After further workup, the patient was found to have ineffective esophageal motility as well. We recommend consideration of MALS as a differential diagnosis in those with chronic abdominal pain. Patient underwent median arcuate ligament release.

### 2. Introduction

Median arcuate ligament syndrome (MALS) is a rare condition with an incidence of 2 per 100,000 patients [1]. Patients typically present with postprandial epigastric abdominal pain, unintentional weight loss, nausea, and vomiting [2]. These symptoms are non-specific, occurring with other pathologies such as cholelithiasis. MALS is caused by compression of the celiac artery and celiac plexus by the MAL. The MAL is a ligament forming a fibrous arch that connects the right and left crura at the level of the aortic hiatus in the diaphragm. The MAL is found anterior and superior to the celiac artery arising at L1-4, while the celiac artery branches the aorta at T11-L1.3,4 The compression caused by the MAL can be exacerbated with expiration as the diaphragm moves cranially causing the MAL to tighten. On the other hand, the compression is relieved with inspiration as the diaphragm moves caudally, causing the ligament to relax. Individuals with abnormally positioned celiac origins, whether it be higher or lower are more susceptible

to compression. In about 10% of patients, the MAL is mispositioned despite having a normal celiac artery origin [3,4].

There is adequate collateral blood flow from the superior mesenteric artery (SMA) and inferior mesenteric artery (IMA) to the bowel which explains the high prevalence of asymptomatic patients with celiac artery compression [5].

The pathophysiology of MALS is still not completely understood. The first hypothesis states that there is some degree of mesenteric ischemia present despite having normal SMA and IMA collateral blood flow. The second hypothesis states that the compression causes injury to the celiac plexus which sets off a chronic inflammatory state in the splanchnic vasculature leading to vasoconstriction and eventually ischemic changes [1].

### 3. Case Presentation

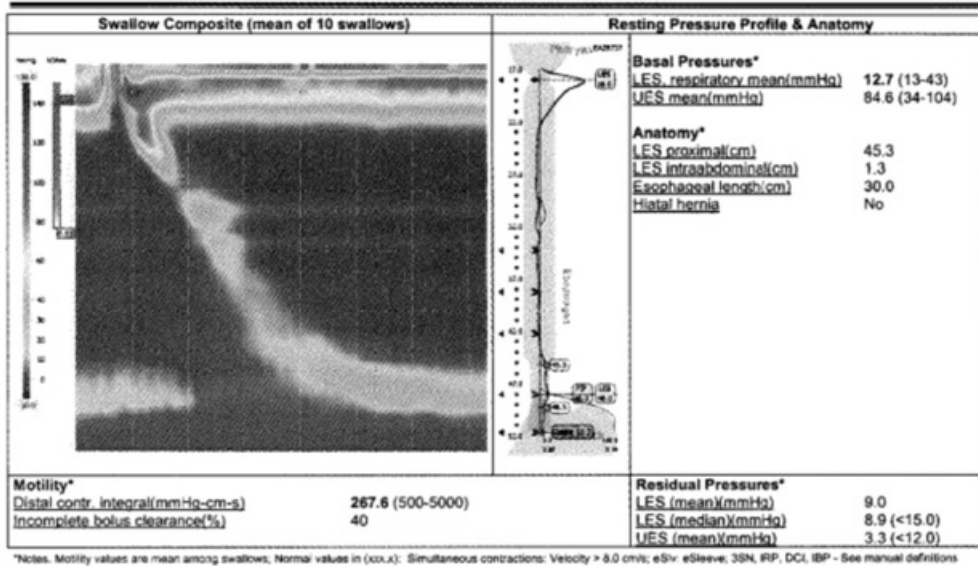
A 57-year-old female presented to the emergency department for chest pain and shortness of breath with occasional postprandial abdominal pain and dysphagia for the past three weeks worsening 4.5 hours prior to arrival. Patient has a past medical history of gastroesophageal reflux disease, achalasia, and Hashimoto's hypothyroidism. Patient stated that her gastric reflux was so severe that she had recurrent aspiration pneumonia in the past. Patient admits to weight loss, constipation, and food fear. Patient denies history of *H. Pylori*, frequent NSAID use, peptic ulcer disease, or Barrett's esophagus. An EKG was done and showed no ST segment changes and a normal sinus rhythm. Complete blood count, comprehensive metabolic panel, chest X-Ray, and troponin were all unremarkable. CTA of the heart showed that the cardiac chambers filled with contrast, with a left ventricular ejection fraction of 60%. Aorta and pulmonary vasculature appeared normal. No tortuous or dilated esophagus was seen. The celiac artery is partially occluded with

marked post-stenotic dilatation secondary to the MAL. Patient's chest pain resolved by the second day of admission but continued to have abdominal pain and dysphagia. CTA abdomen and pelvis showed narrowing of the proximal celiac artery 3x7 mm with dilation distally 11x11 mm with a "hook" appearance of the celiac artery secondary to the MAL (Figure 1). The celiac artery Doppler US with inspiration and expiration study demonstrated a systolic velocity with inspiration of 98 cm/s and with expiration of 197 cm/s. The systolic velocity on expiration was increased, consistent with MALS. An EGD was done which showed no evidence of dilation of the esophagus and no bird's beak deformity. Patient was then placed on a low dose aspirin 81 mg, and rosuvastatin 20 mg and was referred to general surgery. When the patient presented to us for surgery consultation, an esophagram with barium swallow and an esophageal manometry study was ordered to determine if the patient truly has achalasia. A right upper quadrant abdominal ultrasound (US) was ordered which showed no cholelithiasis as the cause of the postprandial abdominal pain. The barium esophagram showed no evidence of achalasia but a small sliding hiatal hernia was seen. The esophageal manometry study did not show achalasia but she had 90% ineffective swallows which confirms the diagnosis of ineffective esophageal motility according to the Chicago Classification v3.0 (Figure 2)[6,7].

The patient underwent laparoscopic median arcuate ligament release. The aorta was visualized through the diaphragmatic hiatus. The right and left crura were merged above the aorta. Once the crura were identified, the crural fibers above the aorta were divided using a Ligasure device. The dissection continued inferiorly to divide all the fibers off of the aorta until the median arcuate ligament was identified overlying the celiac trunk. The division continued until all the fibers over the aorta were dissected until the celiac trunk was completely released and free of obstructing fibers (Figure 3).



**Figure 1:** CT scan of the Abdomen and Pelvis showing narrowing of the proximal celiac artery with distal dilation.



Lower Esophageal Sphincter Region		Normal	Esophageal Motility	Normal
<b>Landmarks</b>				
Proximal LES (from nares)(cm)	45.3		Number of swallows evaluated	10
LES length(cm)	4.1	2.7-4.8	Chicago Classification	
Esophageal length (LES-UES centers)(cm)	30.0		% failed	50
Intraabdominal LES length(cm)	1.3		% weak	40
Hiatal hernia?	No		% ineffective	90
<b>LES Pressures</b>				
Pressure meas. method	eSleeve		% paraesophageal pressurization	0
	eIRP		% premature contraction	0
Basal (respiratory min.)(mmHg)	5.3	4.8-32.0	% rapid contraction	0
Basal (respiratory mean)(mmHg)	12.7	13-43	% fragmented	0
Residual (mean)(mmHg)	9.0		% intact	10
Residual (median)(mmHg)	8.9	<15.0	Number of hypercontractile swallows	0
<b>Upper Esophageal Sphincter</b>				
Mean basal pressure(mmHg)	84.6	34-104	Additional: High Resolution Parameters	
Mean residual pressure(mmHg)	3.3	<12.0	Distal latency	7.5
			Distal contractile integral(mean)(mmHg-cm-s)	267.6 500-5000
			Contractile front velocity(cm/s)	3.5 <9.0
			Impedance analysis	
			Incomplete bolus clearance(%)	40
			Bolus transit time (sec)	4.4
<b>Pharyngeal / UES Motility</b>				
			No. swallows evaluated	10
			Evaluated @ 2.0 & 3.0 above UES	
			Mean peak pressure(mmHg)	39.6

Figure 2: Esophageal Manometry findings of ineffective esophageal motility.

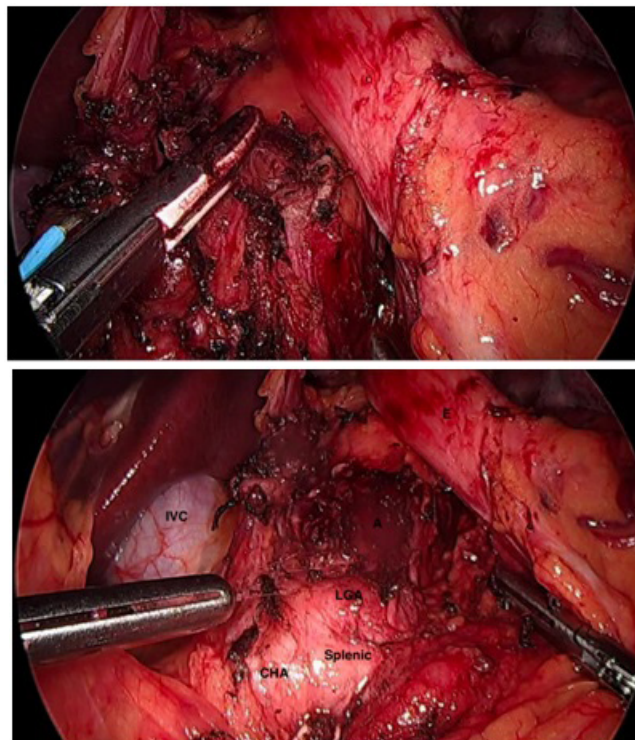


Figure 3: Top: Shows Ligasure surrounding median arcuate ligament.

Bottom: After ligation of the median arcuate ligament with view of the A. Aorta. E. Esophagus. C.H.A. Common Hepatic Artery, L.G.A. Left Gastric Artery, Splenic Artery and I.V.C. Inferior Vena Cava.

#### 4. Discussion

MALS and achalasia are both rare disorders that share symptoms and patient presentation.<sup>8</sup> After any possible cardiac etiology was excluded, further esophageal imaging revealed ineffective esophageal motility disorder and a hiatal hernia. Visceral pain originating in the esophagus is commonly referred to the chest because visceral afferents accompany somatic afferents as they travel from the body wall [9]. In treating MALS, celiac artery decompression by laparoscopic surgical release of the MAL is the goal [10,11]. Additionally, it has been recommended to revascularize the celiac artery if the celiac artery's lumen is stenosed and to perform a celiac ganglionectomy [9]. It is necessary to perform a celiac ganglionectomy because it will relieve the patient's pain and is an inherent part of the dissection to the MAL [12,13]. MALS has implications in esophagectomies due to insufficient blood supply despite the collateral blood supply. It has been shown that there is an increase in morbidity in esophagectomies in those with MALS as well as an increase in esophageal conduit necrosis after the Ivor Lewis procedure [14]. Also, MALS has implications in orthotopic liver transplant (OLT) when the vascular flow is reduced combined with ischemia-reperfusion injury can potentiate together and alter blood flow from the hepatic artery increasing the risk of hepatic artery thrombosis [15]. Is it important to evaluate for MALS during surgical planning for esophagectomies as well as OLT to release the MAL beforehand to prevent these complications. This case adds to the limited literature on esophageal disorders and MALS.

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