

# Minimally invasive approach & key aspects for the treatment of median arcuate ligament syndrome: case report

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**Background:** Median arcuate ligament syndrome (MALS) is a rare condition resulting from the external compression of the celiac artery by an abnormally downward located media arcuate ligament (MAL). The aim of this video is to illustrate the key steps for minimally invasive approach for the surgical treatment of MALS.

**Case Description:** We present the case of an 18-year-old female without any important past medical history referring 1 year of postprandial epigastric pain associated with weight loss. After normal complementary tests, computed tomography (CT) scan showed 60% celiac trunk stenosis with post-stenotic dilation. Surgical approach started with two 5 mm, one 11 mm and one 12 mm laparoscopy trocars. Opening the pars flaccida of the lesser omentum and separating the right crus from the esophagus gives access to the abdominal aorta. Additional care must be taken to avoid injury of the branches of the celiac trunk, especially the common hepatic and left gastric arteries. Careful dissection of adhesions using laparoscopic sealer and hook cautery gives access to the fibrous tissue of the MAL. Finally, as when the musculo-fibrous structure of MAL is seen it must be divided until the celiac trunk is freed completely and its branches are seen. Surgical time was 1 hour 33 minutes long with no blood loss. Post-operative was uneventful (Clavien-Dindo: 0) and the patient was discharged at 48 hours from the admission. During the follow-up, the patient presented adequate oral intake without epigastric postprandial pain.

**Conclusions:** Minimally invasive approach for MAL release is feasible, effective and safe following key steps and paying attention to vascular the structures near the celiac trunk.

Keywords: Median arcuate ligament syndrome (MALS); minimally invasive surgical procedures; laparoscopy; case report

Received: 28 December 2021; Accepted: 19 April 2022; Published online: 27 April 2022. doi: 10.21037/asj-21-116 View this article at: https://dx.doi.org/10.21037/asj-21-116

#### Introduction

Median arcuate ligament syndrome (MALS) is a rare condition resulting from the external compression of the celiac artery by an abnormally downward located media arcuate ligament (MAL) (1,2). The median arcuate ligament is a band of fibrous tissue that joins the left and right crura of the diaphragm to form the anterior surface of the aortic hiatus at the level of the 12th thoracic vertebra. The median arcuate ligament normally is in contact with the aorta above

The incidence of MALS is unknown, but case and series reports have increased in the last years probably due to extensive and more general use computed tomography (CT) and magnetic resonance imaging (MRI).

Various symptoms have been described for this condition, varying from abdominal epigastric pain, weight loss, nausea, vomiting and diarrhea. This vague symptomatology makes

the origin of the celiac axis, but in the case of MALS it is a fibrous tissue located abnormally downward.

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**Video 1** Video showing minimally invasive approach for arcuate ligament syndrome.

clinical diagnosis very difficult and most patients undergo several diagnostics procedures (1).

The standard treatment available is the surgical decompression of the celiac axis by division of the median arcuate ligament. This procedure can be achieved by open, laparoscopic or even robotic approach.

The aim of this video article is to illustrate the key steps for minimally invasive laparoscopic approach for the surgical treatment of MALS. We present the following case in accordance with the CARE reporting checklist (available at https://asj.amegroups.com/article/view/10.21037/asj-21-116/rc).

#### **Case presentation**

We present the case of an 18-year-old female without any important past medical history referring 1 year of postprandial epigastric pain associated with weight-loss. After normal complementary tests, CT scan showed 60% celiac trunk stenosis with post-stenotic dilation.

Surgical approach started with two 5 mm, one 11 mm and one 12 mm laparoscopic trocars. Opening the pars flaccida of the lesser omentum and separating the right crus from the esophagus gives access to the abdominal aorta (*Video 1*). Additional care must be taken to avoid injury of the branches of the celiac trunk, especially the common hepatic and left gastric arteries as described in the article of Gülmez (3). Careful dissection of adhesions using laparoscopic sealer and hook cautery gives access to the fibrous tissue of the MAL as shown in the video. Finally, when the musculo-fibrous structure of MAL is seen it must be divided until the celiac trunk is freed completely and its branches are seen. Surgical time was 1 hour 33 minutes long with no blood loss. Postoperative care was uneventful (Clavien-Dindo: 0) and the patient was discharged within 48 hours from admission. During the follow-up, the patient presented adequate oral intake without epigastric postprandial pain. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the editorial office of this journal.

### Discussion

The pathophysiology of MALS is not well understood but there are two accepted hypotheses: the first one is a neurogenic theory that means that MALS originates from direct sympathetic pain or local irritation or indirect splanchnic stenosis due to external compression of the celiac trunk. In other words, a downward located arcuate ligament produces compression of the celiac trunk with added local irritation. This would explain why some authors describe the association of celiac trunk lymphadenectomy to the arcuate ligament division. The second theory is known as the steal phenomenon, with means that there is movement of the blood flow towards the celiac artery from collaterals between the superior mesenteric artery (SMA) and the celiac trunk (3). Thus, generating blood steal to the organs being vascularize by the SMA, which then produces the symptoms. There is still debate between which one is the most accepted theory, but some authors state that the neurogenic theory would explain better the symptoms in MALS patients.

The treatment of MALS consists in the release of the compression of the celiac axis by dividing the median arcuate ligament as seen in the video. This restores the adequate blood flow through the compressed celiac trunk. Moreover, this can be achieved in open, laparoscopic or robotic surgery. In our case, laparoscopic surgery has several advantages such as less morbidity, less post-operative pain, shorter recovery time with shorter hospital stay, less blood loss and faster return to normal activities as well as more cosmetic result (2). On the other hand, robotic approach offers the same advantages as laparoscopic surgery but it is not available in all centers, requires more economic investment as well as specialized training, without counting on that there are a limited number of cases.

Coelho *et al.* described the laparoscopic procedure in 6 patients with similar symptoms as in this case report. They

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reported successful laparoscopic treatment for all 6 patients and asymptomatic during 3 months follow-up (1).

In about 10-25% of normal people, the median arcuate ligament passes through a lower level at the aorta, compressing the celiac trunk, but only a small subset of patients develop symptoms (2).

Moreover, Sahm and colleagues reported 16 procedures for MALS during a two-year period. In 13 patients (72.2%), a nutcracker syndrome of the left renal vein was additionally diagnosed to MALS. Sixteen patients underwent laparoscopic arcuate ligament release. Postoperatively, 12.5% of the patients still complained of abdominal pain. About half of the patients were completely asymptomatic during followup, and the other half had some detectable abdominal pain afterwards, but 75% of these patients reported a reduction of pain compared with preoperative symptoms (2).

MALS remains a clinical challenge in both diagnosis and treatment. Sahm *et al.* also described as their experience that an exaggerated lumbar lordosis might be a triggering mechanism for the development of a MALS. This may also explain why many patients with MALS suffer from other vascular compression syndromes, such nutcracker syndrome (1,2).

On the other hand, Fernstrum *et al.* described the robotic procedure in 23 patients. At initial follow up with 25 patients (2 patients lost during follow-up), 20 patients (80%) had full and 3 (12%) partial symptom resolution. This gave an initial treatment response rate of 92% (4).

#### Conclusions

In conclusion, minimally invasive approach for median arcuate ligament release is feasible, effective and safe following key steps and paying attention to vascular the structures near the celiac trunk.

## **Acknowledgments**

Funding: None.

#### Footnote

*Reporting Checklist*: The authors have completed the CARE reporting checklist. Available at https://asj.amegroups.com/article/view/10.21037/asj-21-116/rc

*Conflicts of Interest:* All authors have completed the ICMJE uniform disclosure form (available at https://asj.amegroups.com/article/view/10.21037/asj-21-116/coif). The authors

have no conflicts of interest to declare.

*Ethical Statement:* The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the editorial office of this journal.

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# doi: 10.21037/asj-21-116

**Cite this article as:** Jeri-McFarlane S, Bianchi A, Pujol-Cano N, Gil-Catalan A, Martinez-Córcoles JA, Pagan-Pomar A, Gonzalez-Argente FX. Minimally invasive approach & key aspects for the treatment of median arcuate ligament syndrome: case report. AME Surg J 2023;3:20.