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Case Report

Laparoscopic treatment for median arcuate ligament syndrome in children: A case report^{☆,☆☆}

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ABSTRACT

In median arcuate ligament syndrome (MALS), the median arcuate ligament compresses the celiac trunk and surrounding nerves leading to chronic functional abdominal pain and vague gastrointestinal symptoms. MALS can be effectively treated by dividing the arcuate ligament through open surgery or laparoscopy. This is a rare vascular condition and mostly encountered in adult patients. We hereby report a case of a pediatric patient diagnosed with MALS and treated successfully by laparoscopic approach. An 11-year-old girl presented with severe abdominal cramps for 3 months, accompanied by nonbilious vomiting. Computed tomography (CT) angiography demonstrated clear images of celiac trunk compression suggesting MALS. Laparoscopic surgery to cut the ligament and decompress the celiac artery was performed. The patient was discharged on day 7 postoperative with no recurrence of symptoms after 12 months of follow-up. This report suggested the diagnostic value of CT scan, and the safety and the feasibility of laparoscopic surgical techniques to treat MALS in children.

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Introduction

Median Arcuate Ligament Syndrome (MALS) is a rare condition caused by the compression of the celiac artery by the median arcuate ligament (the muscle and fibrous structure that wraps around the aorta at the diaphragmatic opening). This compression leads to transient localized ischemia whenever the artery is compressed during abdominal muscle movement. The consequence is postprandial pain, nausea, vomiting, and weight loss. MALS can be effectively treated by dividing the median arcuate ligament [1,2]. Laparoscopic surgery to divide the median arcuate ligament is increasingly common owing to the advancements in laparoscopic techniques [3–6]. MALS typically presents in adult individuals and there are limited reports on the laparoscopic treatment of this syndrome in children [5–7]. This manuscript was prepared following the CARE guidelines.

Case presentation

An 11-year-old girl with unremarkable medical history was admitted due to epigastric pain lasting for 3 months. She experienced severe, colic abdominal pain 2–3 times a day. Each crisis lasted 1–2 hours and typically aggravated after meals. This was accompanied by occasional nonbilious vomiting. She had normal bowel movements and no fever or weight loss. Previous evaluations suggested gastritis and gastroesophageal reflux, for which Esomeprazole was prescribed for 3 months but did not help improving the patient's symptoms. At admission, her general condition was stable. She showed no remarkable

findings on chest and abdominal examination. There was no associated medical conditions or congenital anomalies. She weighed 44 kg (BMI = 19,3 Kg/m²) with normal psychomotor development.

At the time of admission, the complete blood cell analysis shows normal values of RBC, WBC, and PLT, with the hemoglobin (Hgb) level of 13.1 g/dL. Tests for inflammatory reaction, liver and kidney function, and electrolyte were all within normal limits. The abdominal ultrasound did not reveal any abnormalities. To further investigate the cause of the pronounced abdominal pain, contrast-enhanced abdominal computed tomography (CT) scan was indicated. The CT angiography images showed a clear narrowing of the celiac artery just after its branching point from the aorta, and a prominent poststenotic dilatation of the celiac artery, which were consistent with MALS (Fig. 1). The patient was diagnosed with MALS and scheduled for laparoscopic division of the median arcuate ligament. The patient experienced relief from abdominal pain immediately after surgery. Oral intake was resumed at day 1 and the patient was discharged at day 7 post-operatively. There was no recurrent symptom during the 12-month follow-up.

Surgical technique

The procedure involved 5 trocars: a 10 mm trocar at the umbilicus and 2 working trocars of 5 mm on the right and left abdomen, and 2 subcoastal supporting trocars of 5 mm. Carbon dioxide was insufflated at 10 mmHg. A suspending suture was placed on the lesser omentum facilitating left liver lobe retraction and left gastric artery identification (Fig. 2A). The left gastric artery was isolated and dissected to its origin and the celiac trunk was identified (Fig. 2B). Thick fibrous tissue of

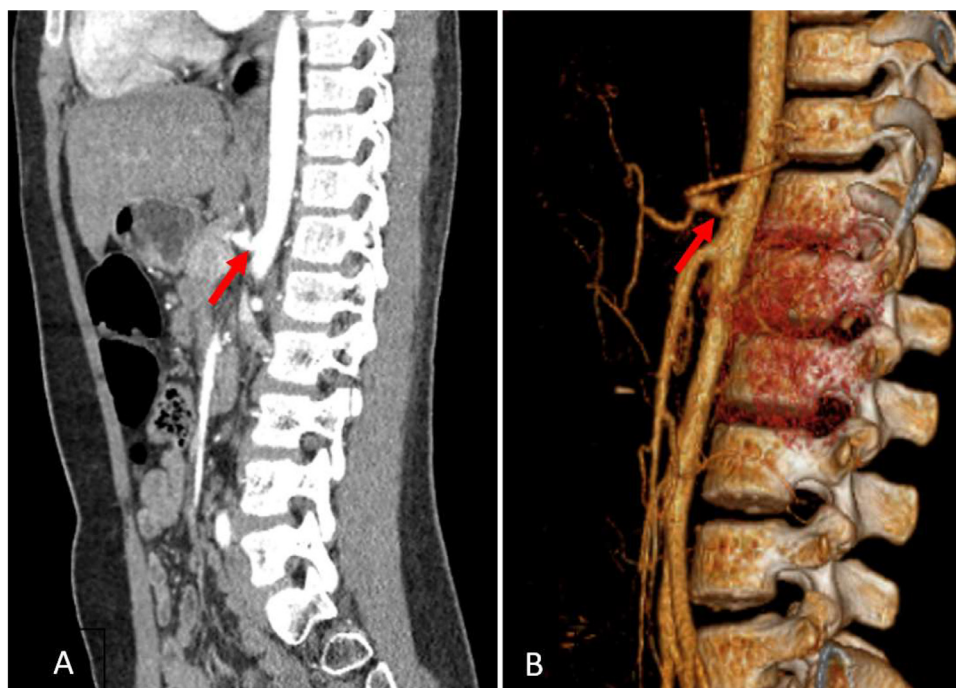


Fig. 1 – The CT angiography images (A-sagittal slice, B-vascular reconstruction) showed the narrowing of the celiac artery (red arrow).

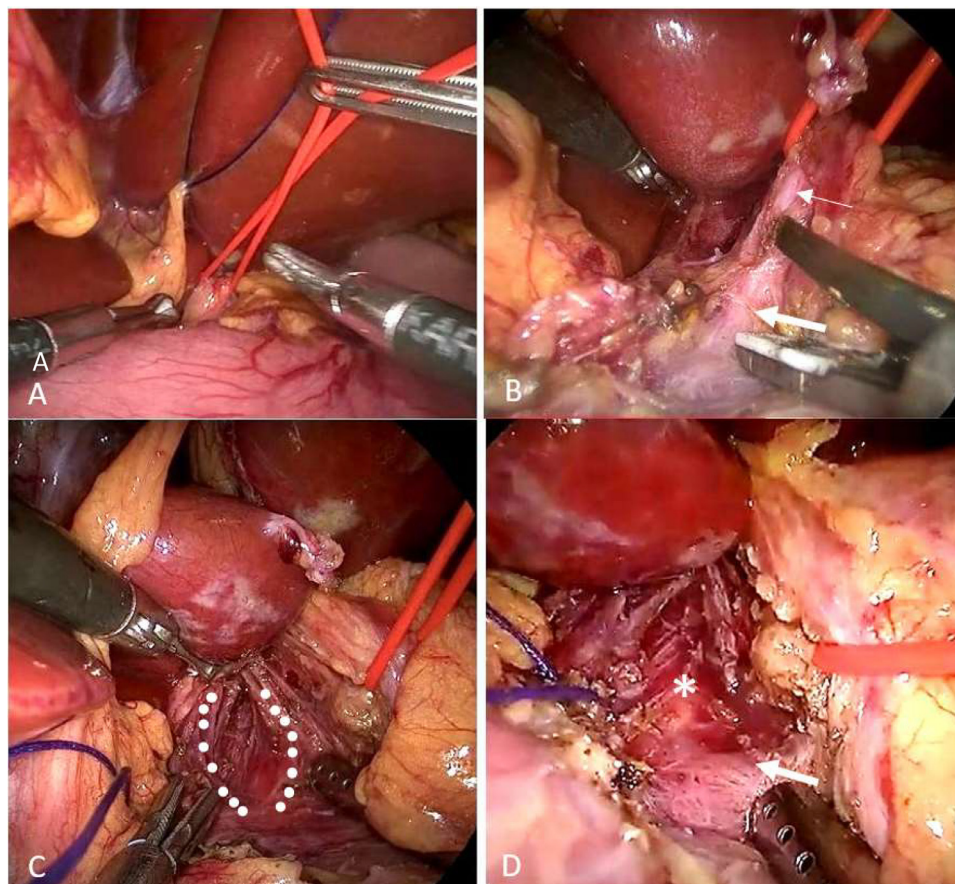


Fig. 2 – Laparoscopic surgical steps in dividing median arcuate ligament.

the median arcuate ligament and nerve plexus was revealed. They were then meticulously divided exposing approximately 4 cm of the aorta, releasing the stenotic celiac trunk (Fig. 2C). Adequacy of the division of the median arcuate ligament was confirmed with no arterial flow restriction through direct visualization during patient's breathing movements (Fig. 2D).

A-Suspending the left lobe of the liver (suture), opening the lesser omentum and identifying the left gastric artery (vessel loop). B-Dissecting the left gastric artery (thin arrow) and the celiac trunk (thick arrow). C- Exposing and dividing fibrous tissue (arcuate ligament) and nerve plexus in half (dotted line). D- Verifying celiac artery (thick arrow) flow through direct inspection following the complete division of the arcuate ligament. The aorta (asterisk) also comes clear in to view.

Discussion

The celiac artery branches off from the abdominal aorta at the level of vertebrae T11 to L1. Normally, the median arcuate ligament (MAL), connecting the diaphragmatic crura at the aortic hiatus, curves up above this aorto-celiac bifurcating level [5]. However, in some variations, the celiac artery originating from a higher position can lead to celiac compression during exhalation by the ligament [8,9]. Chronic compression causes fibrosis and progressive narrowing of the celiac artery [9,10]. This

is associated with local organ ischemia due to reduced blood flow, as well as damage to nerve ganglia. The consequences include recurrent upper abdominal pain, nausea, vomiting, weight loss, and occasionally chronic diarrhea [9,10]. While the compression of the celiac artery by the median arcuate ligament occurs in 10%-25% of the general population, only about 1% of these cases develop into clinically symptomatic MALS, necessitating intervention [11].

Due to the vague manifestation of the syndrome, a wide range of etiologies including gallstones, adhesions in the small bowel, gastroenteritis, gastric ulceration, and incomplete bowel rotation, etc. [9] need to be excluded and this could lead to delay in definite diagnosis. Our patient has been experiencing recurrent and prolonged upper abdominal pain for over 3 months, with an initial diagnosis of gastroenteritis. We decided to proceed with an abdominal contrast-enhanced CT scan to identify the cause of the chronic abdominal pain. In the literature, imaging studies including abdominal ultrasonography and contrast-enhanced CT angiography have showed the value in diagnosis [9]. Doppler ultrasonography is performed during both inspiration and expiration to measure the flow velocity across the celiac artery. Significant narrowing is suspected when the velocity exceeds 200 cm/s [9]. CT angiography is considered the gold standard as it allows direct visualization of the arterial stenotic segment. The image of the arterial compression could be obtained from conventional sagittal plane or on vascular reconstruction

as demonstrated in our case [12]. The primary objective of treatment is effective decompression through the division of the median arcuate ligament. Surgical approaches may involve open surgery or laparoscopy [13]. However, in certain cases, the endovascular or vascular surgical interventions is required. Laparoscopic median arcuate ligament release was first introduced in 2000 and represents a newer approach in the management of celiac artery compression [3,8]. The advantages of the laparoscopic approach include better magnification, less postoperative adhesion formation, smaller cosmetic incisions, quick recovery, and shorter hospitalization [9,12]. In our case, we opted for the laparoscopic approach considering these benefits. Symptomatic improvement is reported in up to 90% of cases within the first year after surgery. Unsatisfactory response to surgical decompression can be attributed to chronic extrinsic compression causing structural changes in the celiac artery, including intimal hyperplasia, elastic fiber hypertrophy, and loss of medial structure. In such cases, combined endovascular dilatation, with or without stent placement, or bypass surgery may be considered. In our case, the outcome of surgical decompression proved to be satisfactory. Although level of evidence of case report suggesting that arterial decompression alone might be adequate for pediatric patients with MALS. This aligns with findings from other reports on pediatric cases [9].

Conclusion

Median arcuate ligament syndrome is uncommon in children and manifests with nonspecific gastrointestinal signs and symptoms. CT angiography is crucial for a conclusive diagnosis. Performing laparoscopic division of the median arcuate ligament in children is both feasible and safe, providing favorable outcomes.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Author's contributions

Trinh-Nguyen HV and Tran TT: Case file retrieval and case summary preparation. Trinh-Nguyen HV and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

Our institution does not require ethical approval for reporting individual cases or case series. Written informed consent was obtained from legal guardians of the patient(s) for their anonymized information to be published in this article.

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