Normalization of Flow in the Common Hepatic Artery after Decompression of Median Arcuate Ligament Syndrome with Diminution of a Pancreatoduodenal Arcade Aneurysm

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Keywords
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Abstract
Median arcuate ligament syndrome (MALS) is caused by constriction of the celiac artery (CA) by the median arcuate ligament of the diaphragm. Ligament release improves perfusion of the CA, resulting in resolution of abdominal symptoms. A 51-year-old female had postprandial abdominal pain for 10 years and underwent computed tomography (CT) scan showing severe stenosis of the CA with pancreatoduodenal arcade aneurysm formation. MALS was diagnosed, and open median arcuate ligament incision was performed to decompress the CA. Intraoperative ultrasonography showed bidirectional turbulent flow in the common hepatic artery (CHA). The median arcuate ligament was uneventfully incised, and compression of the CA released. The perfusion in the CHA was changed to an antegrade direction, and the flow increased. Seven days after the laparotomy, the patient was discharged uneventfully. Follow-up CT scan 20 days after operation showed a diminished pancreatoduodenal arcade aneurysm and inferior pancreatoduodenal artery. Epigastric pain and postprandial distress symptoms were improved. In conclusion, perfusion of the CHA became normalized after median arcuate
ligament release. Surgical intervention for MALS not only improved blood flow in the tributaries but also diminished the pancreatoduodenal arcade aneurysm.

Introduction

Median arcuate ligament syndrome (MALS) is caused by celiac artery (CA) constriction by the median arcuate ligament of the diaphragm. Although several case series were reported, the true incidence of MALS remains unknown. Of asymptomatic subjects, 3.4–7.3% subjects are reported to have CA stenosis. MALS is more prevalent in females and people with a low BMI [1]. In MALS, the superior mesenteric artery “steals” blood flow from the common hepatic artery (CHA) resulting in visceral ischemia (the “steal” phenomenon) that is aggravated postprandially. Reversal of direction of flow in the CHA may result in a pancreatoduodenal arcade aneurysm due to elevated intravascular pressure [2]. Postprandial abdominal pain and pain during deep expiration are characteristic symptoms in patients with MALS.

Ligament release improves abdominal symptoms in 40–60% of patients [3]. Intraoperative Doppler sonography is important to evaluate not only severity of the stenosis but also improvement in the rate of perfusion. Although improvement of CA flow has been reported [4], there are no reports regarding improved CHA flow after surgical intervention. We present a patient who underwent open median arcuate ligament release with evaluation of CHA flow and a pancreatoduodenal arcade aneurysm.

Case Report

A 51-year-old female had postprandial abdominal pain for 10 years and underwent contrast-enhanced computed tomography (CT) scan. The CT scan showed severe stenosis at the CA take-off 0.8 mm in diameter in smallest dimension with formation of a pancreatoduodenal arcade aneurysm accompanied by a dilated inferior pancreatoduodenal artery (Fig. 1a). Endoscopic ultrasonography showed bidirectional turbulent perfusion of both the CHA and CA. Laboratory findings showed no abnormalities. MALS was diagnosed, and open median arcuate ligament incision was planned to decompress the CA.

During laparotomy (Fig. 2), vascular flow was evaluated with Veri-Q (Medistim, Oslo, Norway) before exposing the CA (Fig. 3). Bidirectional turbulent flow was observed in the CHA, and the mean flow rate was 3 mL/min in a retrograde direction with a systemic blood pressure of 85/49 mm Hg and pulse of 74 per minute. The median arcuate ligament was incised and compression of the CA released. The perfusion was evaluated again 10 min after decompression to avoid the influence of vasospasm. The perfusion in the CHA changed to an antegrade direction, and the flow increased to 38 mL/min with a systemic blood pressure of 89/52 mm Hg and pulse of 70 per minute (Fig. 3). Seven days after laparotomy, the patient was discharged uneventfully. Follow-up CT scan 20 days after surgery showed diminution of the pancreatoduodenal arcade aneurysm (118.9–82.3 mm³) and the inferior pancreatoduodenal artery (3.5–1.9 mm in maximal diameter) (Fig. 1b).

The patient’s abdominal symptoms before and after surgery were assessed using the Izumo scale score with a Likert scale, a self-reporting questionnaire reflecting various abdominal symptoms including epigastric pain and postprandial distress domains [5]. Using a Likert scale, a higher domain-specific score indicates more severe symptoms. In the epigastric pain
and postprandial distress domains, the score improved from 5 to 2 and from 9 to 4, respectively. The patient was satisfied with this symptomatic amelioration.

**Discussion**

Incising the median arcuate ligament for MALS in the present patient normalized the direction of flow in the CHA due to decompression of the CA. This improved perfusion has the potential to prevent aneurysm formation or aggravation. Further, the present patient also showed symptomatic amelioration.

Fig. 1. **a** Preoperative 3D-CT scan. The median arcuate ligament compresses the CA (13 mm in length, 0.8 mm in diameter) (white arrow). There is an inferior pancreatoduodenal arcade aneurysm (118.9 mm\(^3\)) (blue arrow). **b** Postoperative 3D-CT scan showed decompression of the CA up to 2.4 mm in diameter. The inferior pancreatoduodenal arcade aneurysm is diminished to 82.3 mm\(^3\) (blue arrow). 3D-CT, three-dimensional computed tomography.

Fig. 2. Surgical procedure for MALS. **a** The median arcuate ligament (white arrow heads) is incised anterior to the abdominal aorta after controlling the CHA (white arrow), SA (yellow arrow), and LGA (blue arrow). **b** After the incision, the CA becomes enlarged and pulsatile (green arrow). SA, splenic artery; LGA, left gastric artery.
improvement of both epigastric pain and postprandial distress symptoms. To the best of our knowledge, this is the first report that incising the median arcuate ligament normalized the direction of flow in the CHA resulting in diminution of a pancreatoduodenal arcade aneurysm.

MALS was first reported in 1963 as extrinsic compression of the CA by the median arcuate ligament which connects the right and left crura at the aortic hiatus [6]. In 1965, median arcuate ligament incision was reported to improve persistent abdominal symptoms including abdominal pain, nausea, vomiting, and weight loss. The sustained symptomatic relief rate after operation was reportedly 81% [7]. Complication by psychiatric disorders or alcohol abuse was associated with negative long-term outcomes after surgery [7]. Severe CA occlusion (>70%) may be a positive predictor for good long-term outcomes after surgery [8].

Pancreatoduodenal arcade aneurysms are believed to form as a result of elevated intra-vascular pressure in the pancreatoduodenal arcade resulting from proximal CA stenosis. This aneurysm has a comparatively high rupture rate reportedly ranging between 20 and 80% [9]. Although ligament incision accompanied by endovascular intervention is sometimes selected in patients with large aneurysms [2], surgical decompression of the CA can normalize the direction of flow, resulting in spontaneous diminution of the aneurysm. High-risk visceral aneurysms are characterized by large size (>2 cm), rapid growth, the presence of symptoms, and multiple aneurysms [9]. The aneurysm in the present patient did not have these characteristics. The clinical presentation of a ruptured pancreatoduodenal arcade aneurysm includes gastrointestinal bleeding, abdominal pain, and gastric-outlet obstruction [10]. In the present patient, antegrade perfusion of the CHA was confirmed after incising the ligament, obviating the need for further intervention with close follow-up for the pancreatoduodenal arcade aneurysm. If the direction of flow is not normalized after incising the ligament, intravascular intervention or revascularization surgery should be considered.

In conclusion, evaluation of the CHA using intraoperative ultrasound revealed normalization of the direction of flow after median arcuate ligament incision. A postoperative CT scan showed diminution of a pancreatoduodenal arcade aneurysm and the inferior pancreatoduodenal artery. Therefore, endovascular intervention for the aneurysm can be avoided with improved perfusion of the CHA after surgery as long as the aneurysm is small. Future studies are necessary to confirm this preliminary result.

**Fig. 3.** Schema of visceral perfusion. **a** Before median arcuate ligament incision, the CHA perfusion is turbulent, with a mean rate of 3 mL/min in a retrograde direction. Flow in both the SA (66 mL/min) and LGA (17 mL/min) is antegrade. **b** After incision, the flow in the CHA changed to an antegrade direction at a rate of 38 mL/min. Antegrade flow is observed in the SA (48 mL/min), LGA (15 mL/min), and CA (40 mL/min). SA, splenic artery; LGA, left gastric artery.
Statement of Ethics

Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images. This study protocol was reviewed and approved by the Ethics Committee of Saiseikai Utsunomiya Hospital (ID#2021-61).

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Kenji Matsumoto and Hiroharu Shinozaki drafted the manuscript and acquired the data. Satoshi Shinozaki and Alan Kawarai Lefor drafted and wrote the manuscript. Toshiaki Terauchi and Naohiro Sata revised it critically for important intellectual content. All the authors read and approved the final version of the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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