Aneurysmal celiac trunk dissection caused by median arcuate ligament syndrome successfully treated by endovascular technique: a case report

Sen Li[1#], Jiaqing Luo[2#], Li Yin[1], Chao Yan[1], Yuhua Zhu[2], Jian Wang[1], Zhiwei Gao[1], Zhenjie Liu[1], Bing Chen[1]

[1]Department of Vascular Surgery, The Second Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China; [2]Department of General Surgery, Changxing People Hospital, Huzhou, China

[#]These authors contributed equally to this work.

Correspondence to: Bing Chen, MD, PhD. Department of Vascular Surgery, The Second Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China. Email: 2114008@zju.edu.cn.

Abstract: Median arcuate ligament syndrome (MALS) is a chronic pathogenic process, resulting from the compression of the celiac axis by fibrous attachments of the diaphragmatic crura which named median arcuate ligament. In clinical, isolated spontaneous superior mesenteric artery (SMA) is common. Whereas, isolated celiac trunk (CA) dissection is rare, it is usually associated with aortic dissection. In this case report, the CA is not only dissected, but also the aneurysm formed. Because the long-time compression of the MALS, relative narrowing of the celiac artery causes the high pressure at the beginning of the CA and that may contribute to the aneurysmal celiac trunk dissection (ACTD). This case described a male patient who underwent hypertension and abdominal pain as their main clinical manifestation. Contrast-enhanced computed tomography angiography (CTA) showed that a slightly stenosed root of the celiac artery during inhalation and severe stenosis of the root during expiration. Moreover, a small intimal tear at the site of celiac artery proximal end with nonruptured aneurysmal dilation located on the celiac trunk. After our carefully examinations, the patient was diagnosed as ACTD caused by MALS. And we applied endovascular technique-stenting in this case. It could not only cover the dissection, but also could obstruct the aneurysm. The patient was in a stable condition after management of ACTD by stenting. Stenting is a safe and effective treatment for ACTD caused by MALS.

Keywords: Aneurysmal celiac trunk dissection (ACTD); median arcuate ligament syndrome (MALS); stenting; celiac artery; case report

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Introduction

MALS is a rare vascular pathology (1,2). It occurs when the median fibrous arcuate ligament and muscular diaphragm fiber have a relatively low insertion, and cause extrinsic compression and luminal narrowing of the celiac trunk (3). Aneurysmal celiac trunk dissection (ACTD) caused by MALS is a much more rarely entity. Recently, Hans published a similar paper concerned about MALS with a coincident celiac artery aneurysm and dissection. However, conservative management was adopted in this case (4). To the authors' knowledge, the invasive treatment of ACTD caused by MALS is yet to be reported. Here, we report a case in which the endovascular technique is a feasible, efficient and safe technique for the treatment of ACTD.

Case presentation

Written informed consent was obtained from the patient. A 54-year-old man, experienced 3-day abdomen painfully swollen, was admitted to the hospital on June 22, 2017. It was noted that he had suffered from hypertension for almost one year without medication. The highest blood pleasure was

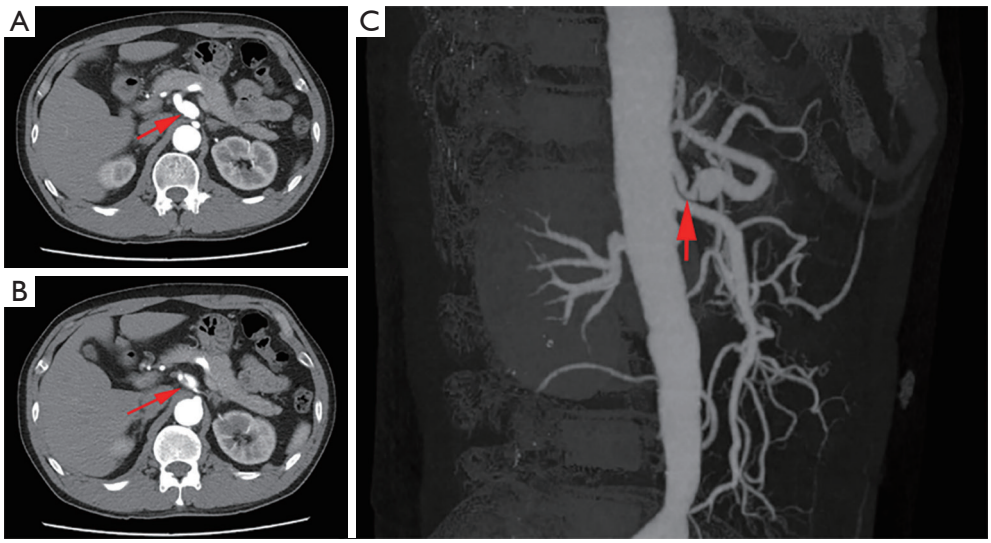

Figure 1 The CTA of ACTD caused by MALS. CTA demonstrated that the aneurysm was 15 mm in diameter and 22 mm long, 20 mm away from the origin of the celiac trunk without signs of calcification or thrombus, and the dissection did not extend beyond the celiac trunk. Arrow: (A) celiac artery aneurysm; (B) a small intimal tear at the site of focal celiac artery; (C) narrowing of the celiac artery. CTA, computed tomography angiography; ACTD, aneurysmal celiac trunk dissection; MALS, median arcuate ligament syndrome.

180/140 mmHg. He had a smoking history for almost 30 years with 20 cigarettes per day. He denied any genetic disorder hereditary disease, diabetes mellitus, stroke and coronary heart disease. Moreover, he had no such medical history before. Physical exam was positive for abdominal tenderness, whereas the blood test was negative. Computed tomography angiography (CTA) documented a small intimal tear at the site of focal celiac artery (CA) with nonruptured aneurysmal dilation located on the celiac trunk. This aneurysm was 15 mm in diameter and 22 mm long, 20 mm away from the origin of the celiac trunk without signs of calcification or thrombus, and the dissection did not extend to the collaterals (*Figure 1*). Moreover, the celiac axis was severely compressed by the arcuate ligament. Since this disease is rare, it is easy to be missed. There were other diagnosis considered, such as celiac artery dissection, celiac artery aneurysm and so on. However, after our carefully examinations, the patient was much more complex, it is diagnosed as ACTD caused by MALS. A favorable medical prognosis will be achieved once the aneurysm and the dissection were covered.

There are several strategies could be done to deal the disease, such as endovascular treatment, open surgery, laparoscopy, and so on. After our carefully considering patient's situation and will, we performed endovascular surgery for the patient on June 26, 2017. The patient was placed under vertebral canna injection anesthesia in the supine position with the legs apart in reverse Trendelenburg position. The pigtail catheter was inserted into the abdominal aorta by the 6 Fr introducer sheath through the right femoral according to the Seldinger method. Abdominal aorta angiography was performed at the 12 thoracic vertebrae level. The image showed a slightly stenosed root of the celiac artery during inhalation and severe stenosis of the root during expiration. Moreover, a 22×15 mm aneurysm dissection was noticed at the beginning of the celiac artery (*Figure 2*). Considering the difficulty of the stent implantation due to the acute angle of the CA and high tension, we positioned the guide wire through the left branchial artery. The Viabahn 7×50 mm stent covered stent was implanted with the help of road map (*Figure 2*). Distal position of the stent was at the forking of the CA and proximal position was at beginning of the CA. After the stent was released, we could see the stent was in good shape, the dissection and aneurysm were disappeared, and there was no blood flow limitation of the vessels. After surgery, we applied Rivaroxaban 20 mg once a day, Norvasc 5 mg and Aprovel 150 mg once a day for one year in that patient. And the patient was discharged on the third postoperative day.

After one-year follow-up, the patient came to the clinic without any discomforts. The CTA showed that the stent was on the position of celiac artery without any narrowing. There were no signs of calcification, thrombus or deficient visceral perfusion (*Figure 3*). And there are no postoperative

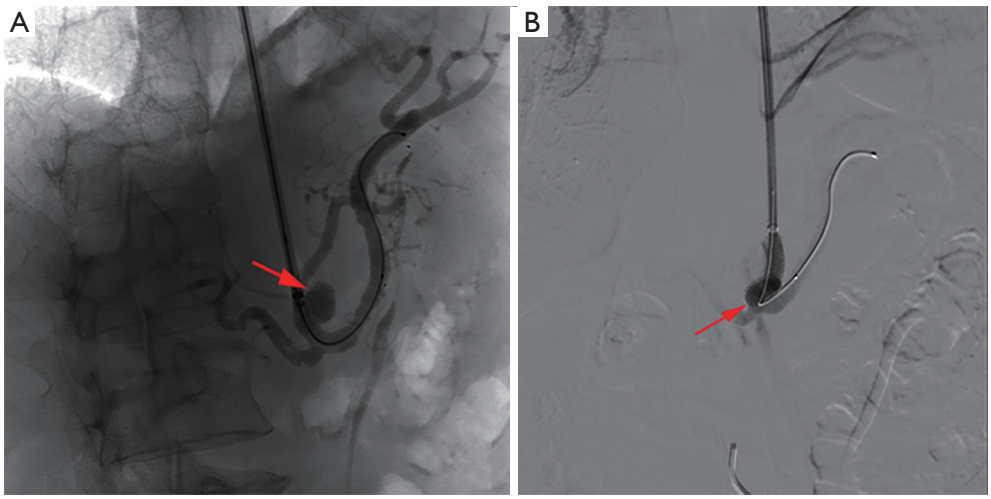

Figure 2 The angiography of ACTD caused by MALS. (A) Angiography showed that the aneurysm was 15 mm in diameter and 22 mm long, 20 mm away from the origin of the celiac trunk, and the dissection did not extend beyond the celiac trunk. (B) Viabahn 7×50 mm stent covered stent was implanted with the help of road map. Distal position was at the forking of the celiac artery and proximal position was at beginning of the CA. Arrow: celiac artery aneurysm. ACTD, aneurysmal celiac trunk dissection; MALS, median arcuate ligament syndrome; CA, celiac artery.

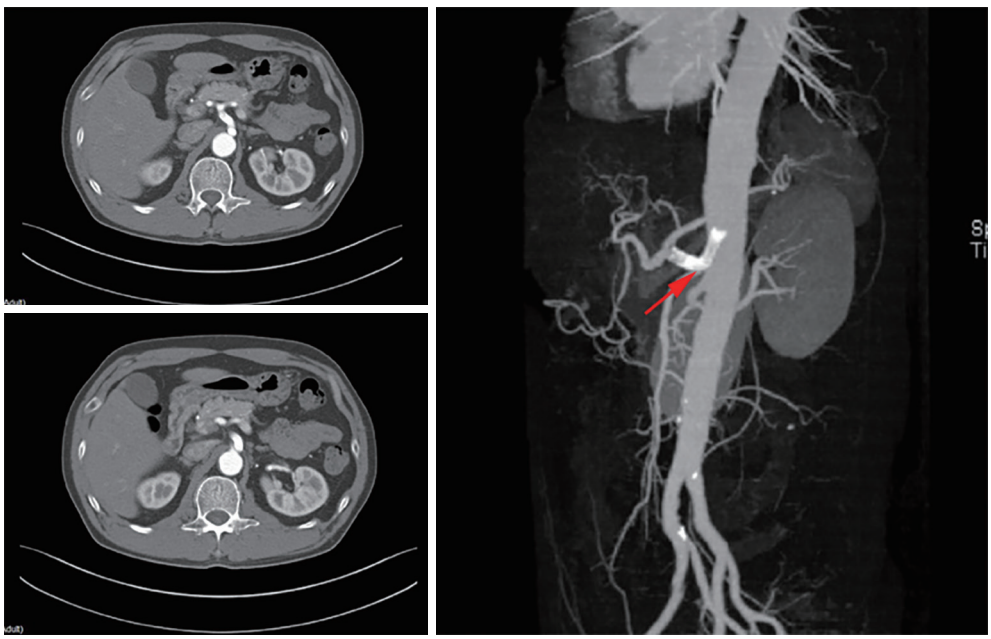

Figure 3 One year follow-up CTA of ACTD caused by MALS. CTA showed that after one-year follow-up, the stent was in good shape, the dissection and aneurysm was disappeared, and there was no blood flow limitation of the vessels. Arrow: celiac artery stent. CTA, computed tomography angiography; ACTD, aneurysmal celiac trunk dissection; MALS, median arcuate ligament syndrome.

complications appear, such as abdominal pain, stent occlusion and relevant artery dissection. The timeline of the whole treatment and follow-up process was present in (*Figure 4*).

Discussion

Isolated visceral artery aneurysm and dissection is a rare

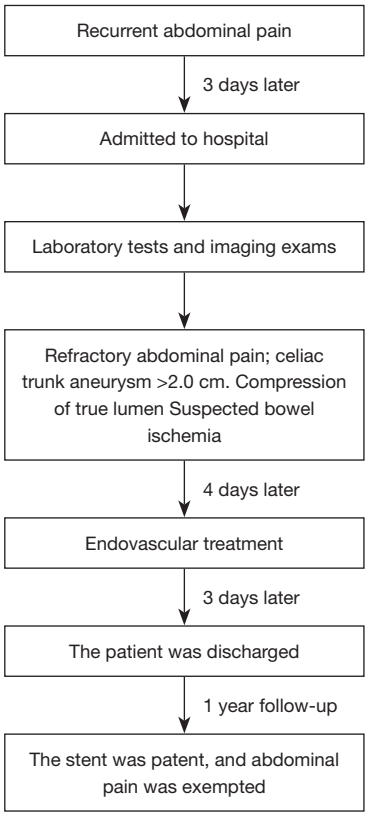

Figure 4 Timeline of the whole treatment and follow-up process.

clinical condition and its physiopathology is still unknown (5-8). Moreover, this condition is most commonly located in the superior mesenteric artery (SMA), and it is extremely rare in the celiac trunk (9-11).

The formation mechanism of the aneurysmal dissection of the CA by MALS is as follow: (I) the celiac artery is compressed by the Median arcuate ligament; (II) narrowing of the CA caused the relative high pressure at the beginning of the CA; (III) the patient had hypertension for almost one year without medication treatment, which also imposes enhanced hemodynamic stress on CA; (IV) the CA is not very thick, and it cannot bear much blood pressure; (V) fibromuscular dysplasia, and connective tissue disease may also cause the disease. All these reasons could promote aneurysmal formation and dissection. Spontaneous dissection of the celiac trunk is most often related to the extension of aortic dissection (12-14). ACTD caused by MALS is rare. Expansion of a false lumen due to mechanical compression of true lumen flow, aneurysmal dilation, or rupture may cause malperfusion. Moreover, it could propagate into adjacent vessels, which results in the splenic and renal infarcts. So timely treatment is necessary.

There are several choices of treatment for MALS: [open (15,16), laparoscopic approach (17,18), robot-assisted (19) or endovascular treatment (14)]. Although no optimum treatment for ACTD has been established until now, all those treatments are feasible for this patient. In this case, we performed endovascular treatment to cure the disease. Because endovascular management has numerous potential advantages: the intervention can be achieved through local anesthesia; collateral circulation is easily assessed with selective splanchnic arteriography during the procedure, fewer postoperative complications occur and hospital stays are shorter (14). Moreover, stenting of the CA could not only cover the tear, but also could block the aneurysm. In our opinion, for both celiac trunk aneurysm and spontaneous dissection, endovascular technique may be the first-choice of treatment. Roh and LeSher (20) reported a rupture that resulted in the death of the patient before any endovascular intervention. Our patient was fortunate. He has not only avoided the CA rupture, but also has been well after one-year follow-up. The CA is still patent and he did not have any discomforts.

Covered-stent placement is a safe and effective treatment for ACTD caused by MALS and can provide immediate symptom relief. Since the large clinical trials are unavailable due to the rarity of the syndrome, such case was carried out to demonstrate the efficacy and advantages of the covered-stent placement approach.

There are still some limitations in this case. Firstly, a longer duration of follow-up may further elucidate the feasible and efficient of the endovascular treatment to the disease. Secondly, the sample size of that patient was small and only endovascular treatment was applied in that disease. Thirdly, it will be better if more cases and treatment strategies was collected and propensity match model was conducted to rule out some irrelevant factors.

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Footnote

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uniform disclosure form (available at https://dx.doi.org/10.21037/acr-20-123). 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 investing and resolved. All procedures performed in studies involving human participants 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).

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