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CASE REPORT

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Median arcuate ligament syndrome in an old male: a case report with occlusion of celiac artery after stenting

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Abstract

Background Median arcuate ligament syndrome (MALS), also called celiac artery compression syndrome or Dunbar syndrome, is a rare disorder caused by the compression of the celiac trunk by the median arcuate ligament, which results in patients presenting with bloating, vomiting, nausea, weight loss, and postprandial abdominal pain.

Case presentation A 77-year-old male was admitted to our center with irregular abdominal pain over the epigastric region for the past 5 months. The pain occurred with no apparent causes, which had intensified in the last 10 days, without nausea, vomiting, and other symptoms. The physical examination, laboratory examination, abdominal ultrasound, and gastroenterological endoscope showed no obvious abnormalities. The angiography showed that the celiac artery was 90% narrowed, so revascularization was performed, leading to a resolution of the symptoms. After 6 months, the patient presented with a recurrence of abdominal pain. Computed tomography angiography showed the stent in the ostial celiac artery was compressed and deformed, which obstructed the vessel. Finally, due to the advanced age, and high surgical risk, the patient was not willing for the decompression of the celiac artery, and the post-dilation was performed, resulting in < 50% residual stenosis in the ostial celiac artery and resolution of pain.

Clinical discussion The current diagnosis of MALS is still based on postprandial abdominal pain and imaging modalities. However, due to the atypical symptoms and imaging manifestation, MALS is diagnosed precisely only after extensive evaluation and exclusion. In our case, celiac artery stenosis was initially diagnosed based on the symptoms and the results from angiography, so the revascularization of the celiac artery was conducted, leading to the deformation of the stent and a recurrence of abdominal pain 6 months later. MALS and decompression of the celiac trunk were finally considered. Although he refused to undergo celiac artery decompression because of the high surgery risk, the abdominal pain was relieved by post-dilation during the follow-up of 8 months.

Conclusion Due to the vague manifestation, MALS should be considered after excluding intestinal disorders using different imaging modalities. Once diagnosed, the goal of treatment was centered around the decompression of the celiac artery.

Keywords Median arcuate ligament syndrome, Celiac artery compression syndrome, Dunbar syndrome, Vascular compression syndrome, Case report

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Introduction

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome or Dunbar syndrome, is an exceptionally rare syndrome resulting from the compression of the celiac artery and celiac plex by the median arcuate ligament (MAL) and diaphragmatic crura [1]. The MAL, a tendinous fascial band, serves to connect anteriorly the diaphragmatic crus of either side surrounding the aortic hiatus [2]. This structure is usually superior to the origin of the celiac artery. However, MALS may develop in some patients because of the higher origin of the celiac artery or lower insertion of diaphragmatic crura [3]. Furthermore, the neuropathic pain associated with the MALS was caused by the extrinsic compression of the MAL on the celiac plex, which is located next to the celiac artery [1, 4]. As a result, the patients may experience a constellation of nonspecific symptoms, such as exercise-induced or postprandial abdominal pain, nausea, vomiting, diarrhea, subsequent food aversion, and unintentional weight loss [5, 6]. Due to the poorly understood pathophysiology of MALS and the variable presentations among patients, MALS remains a diagnosis of exclusion, and extensive investigations are performed to exclude the common causes of abdominal pain [5]. The optimal treatment for MALS aims at decompression of the celiac artery through open laparotomy, retroperitoneal endoscopy, laparoscopy, or robot-assisted surgery, either with celiac gangliectomy and angioplasty or not [6]. We herein report a case of MALS in a 77-year-old male who presents with the recurrence of abdominal pain after a failed stenting. Our case report has been reported in line with the SCARE 2023 criteria [7].

Case report

A 77-year-old male was admitted to the local hospital with a chief complaint of irregular epigastric pain for the past 5 months, which was worse 10 days before admission. The pain was epigastric in location, mild in intensity, not associated with eating and physical activities, and persistent despite antacid use. The patient reported chest tightness and shortness of breath but had no nausea, vomiting, bloating, or alteration in bowel habits. The patient had a history of chronic obstructive pulmonary disease (COPD) and smoking but did not have a history of hypertension, diabetes, or gastric ulcer. There was also no history of drug use or alcohol intake. He underwent coronary angiography, which revealed severe stenosis (85%) in the proximal to mid part of the left anterior descending (LAD), severe stenosis (80%) in the mid part of the left circumflex (LCX), and mild stenosis in the right coronary artery (RCA). Computed tomography angiography (CTA) was also performed, showing the initial portion of the celiac trunk was approximately 70% narrowed.

On admission to our hospital, physical examination was unremarkable, with no peritoneal irritation signs and normal bowel sounds on auscultation. His initial blood investigations, including a complete blood count, liver functions, kidney functions, electrolytes, thyroid function tests, B-type natriuretic peptide (BNP), and tumor markers, were within the normal limits. Both abdominal ultrasound and gastroenterological endoscope revealed no abnormal findings. The electrocardiogram showed T-wave flattening at the anterior lead, which showed the pain may be associated with angina pectoris, combined with the results from the prior angiogram. Therefore, the patient was then referred for coronary angiography, which revealed severe stenosis (95%) in the mid part of the LAD and severe stenosis (90%) in the mid part of the LCX, as depicted in Fig. 1A and B. Percutaneous coronary intervention (PCI) was then successfully performed for LAD by Resolute 2.5*24 mm stent and Resolute 2.75*18 mm stent (Fig. 1C). PCI was also performed on the RCA using EXCEL 2.5*33 mm stent, as shown in Fig. 1D. Subsequent angiography revealed TIMI grade 3 flow in both target vessels and demonstrated complete expansion and apposition of the stent struts. Based on the unrelieved symptoms and results from the prior CTA, the decision was made to perform an abdominal angiography, which revealed significant stenosis (95%) in the ostial part of the celiac trunk and moderate narrowing in the common hepatic artery, the left gastric artery, and the splenic artery. Given the possibility of celiac trunk narrowing caused by atherosclerosis, an EV3 8*33 mm stent was implanted in the ostial celiac trunk, as depicted in Fig. 2. After the operation, he was treated with indobufen and clopidogrel for antiplatelet therapy and rosuvastatin for lipid modulation. The patient reported significant relief postoperatively and was discharged on day 3 post-operation.

Six months following the operation, the patient experienced a recurrence of abdominal pain. CTA revealed that the stent in the ostial celiac artery was compressed and deformed, which obstructed the vessel (Fig. 3A and B). The patient also underwent the angiography via the left radial artery, and balloon angioplasty was performed in the stent by 2.0*20 mm, 4.0*15 mm, and 6.0*40 mm ballon, with < 50% residual stenosis in the ostial celiac artery (Fig. 3C, D, and E). These findings confirmed the diagnosis of MALS, and the arcuate ligament release was proposed. Given the high anesthesia-associated risk and high surgical risk, the patient refused the surgical treatment. He recovered well after post-dilation and was discharged on the third day. Due to poor pulmonary function, duplex ultrasonography (DHS) was used 1 week postoperatively, revealing that the flow velocity varies with breathing maneuvers, 272 cm/s in inspiration and 366 cm/s in expiration (Fig. 4A and B). After 4 months, DHS revealed

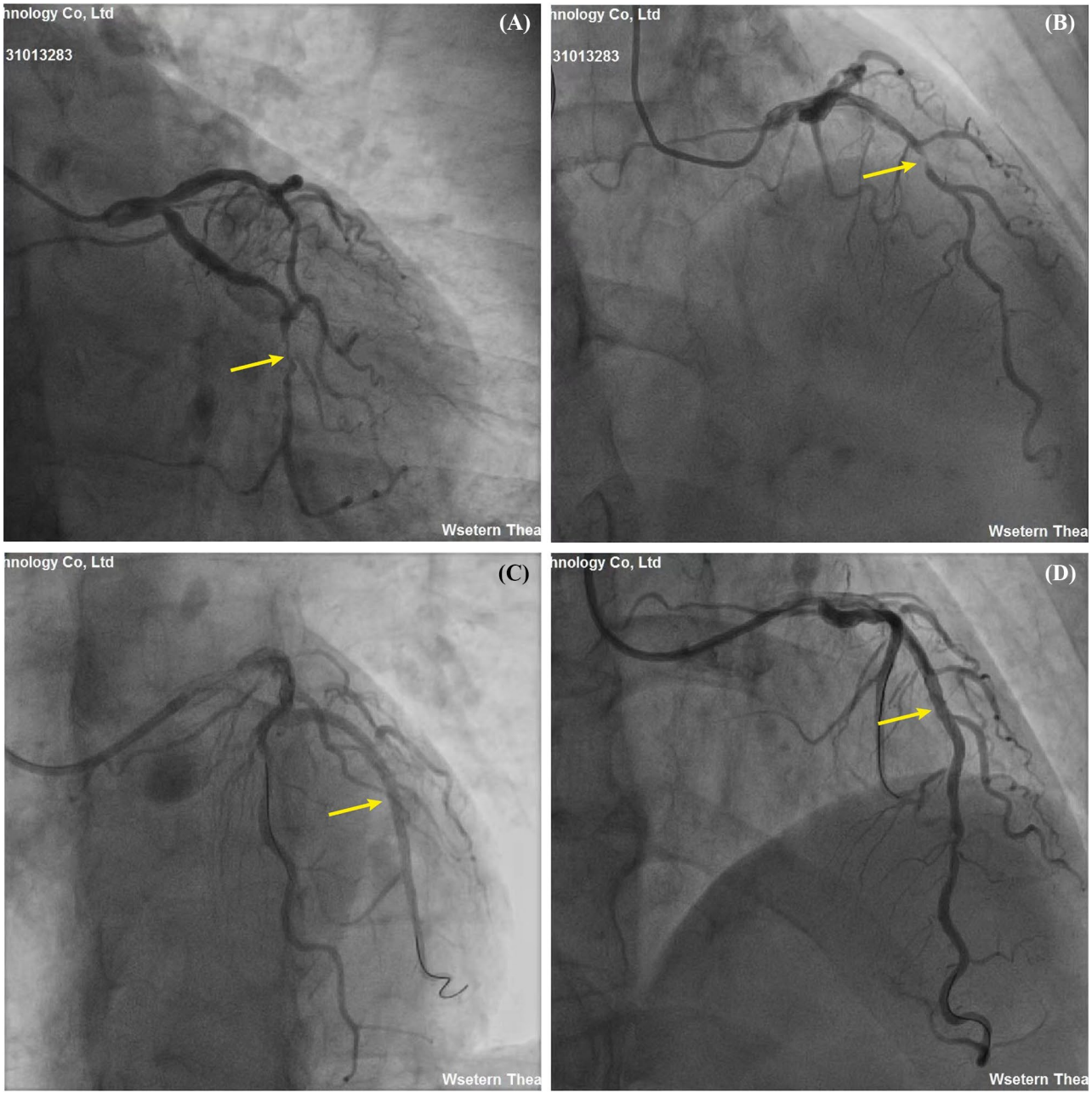

Fig. 1 Coronary angiography image showing stenosis of the LAD and LCX. (**A**) significant lesion in the LAD. (**B**) significant lesion in the LCX. (**C**) PCI on LAD. (**D**) PCI on LCX

the peak systolic velocity of 216 cm/s at the end of deep inspiration and 0 cm/s at the end of deep expiration, as shown in Fig. 4C and D. With the follow-up of 8 months, DHS showed a blood flow velocity of 203 cm/s at the end of inspiration and 322 cm/s at the end of expiration (Figure E and F). The abdominal pain has subsided during the follow-up of 8 months.

Discussion

MALS is an uncommon disorder primarily caused by the compression of the celiac axis by the MAL and the diaphragmatic crura [1]. The disorder affects approximately 2 per 100,000 people and shows a predilection for women aged between 30 and 40 years with thin body habitus [2]. The clinical presentations of MALS are variable and are characterized by chronic postprandial abdominal pain, nausea, vomiting, diarrhea, and unintentional weight loss [1, 6]. Physical examination may reveal epigastric tenderness and abdominal bruits, which are exaggerated upon

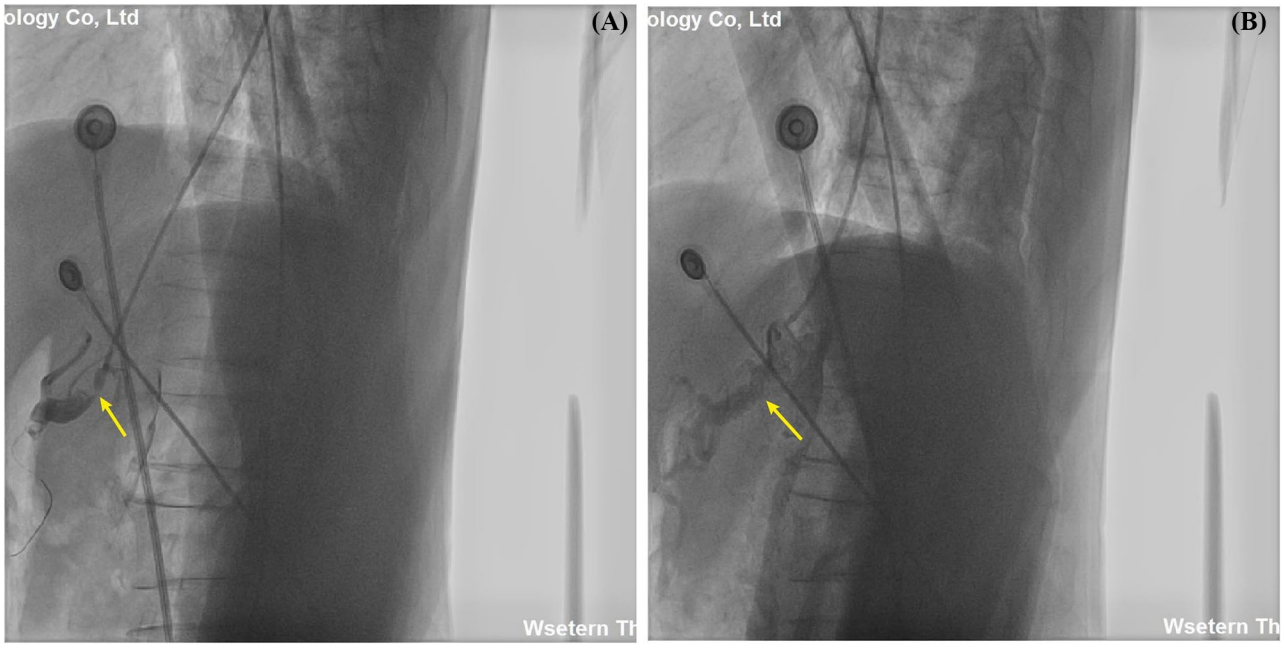

Fig. 2 Celiac angiogram image of aorta showing focal narrowing at the celiac artery. (**A**) celiac angiogram showing severe stenosis in the ostial celiac trunk. (**B**) an EV3 8*33 mm stent was implanted in the ostial celiac trunk, with complete expansion of the stent

expiration [6]. The underlying pathophysiology behind the spectrum of symptoms of patients with MALS remains unknown. Still, it has been postulated that the disorder occurs when the origin of the celiac artery is slightly higher than usual or the insertion of the MAL is somewhat lower than normal [8]. Another possible cause of the pain may be the combination of chronic compression and overstimulation of celiac ganglion, which results in the direct irritation and stimulation of sympathetic pain fibers and splanchnic vasoconstriction and ischemia [9]. Furthermore, these symptoms of celiac artery compression in patients with occluded or compressed celiac trunk may be due to the vascular steal of blood flow by larger collateral vessels [10].

Due to the nonspecific clinical presentations, MALS is usually a diagnosis of exclusion, and multiple modalities are needed to rule out the alternative causes of abdominal pain before arriving at a diagnosis. DHS is a typical initial investigation in patients with suspected celiac artery compression, as it is noninvasive and inexpensive, and it doesn't expose patients to high doses of radiation [2]. Additionally, DHS is reliable in demonstrating the exact configuration of a dynamic celiac artery and showing that the celiac axis tracks cephalad during expiration, which leads to external compression and elevated flow velocities with post-stenotic dilation [1]. However, it is operator-dependent and may also be limited by the body habitus of patients and overlying bowel gas [3]. Additional modalities, such as CTA, magnetic resonance angiography (MRA), or digital subtraction angiography (DSA), can be used to visualize the location of the celiac trunk. CTA helps in diagnosis, allowing the three-dimensional reconstruction and visualization of the compressed artery from different angles and showing changes like post-stenotic dilation [3, 10]. CTA has a high resolution and is also used to identify the concomitant abdominal pathology in addition to findings of MALS [1]. MRA provides results similar to those of CTA and can be used in patients with intravenous contrast allergy and renal dysfunction, which are contraindications for CTA [2]. In addition, angiography with breathing maneuvers is the gold standard investigation and reveals the cephalad movement of the celiac axis during expiration increases the celiac artery compression and post-stenotic dilatation [3, 5].

The treatment of MALS centers around the decompression of the celiac artery to restore the blood supply and pain management by neurolysis. Decompression of the celiac artery was traditionally achieved by using an open approach to the surgery, which involves the dissection and the separation of the diaphragmatic crura from the celiac axis [2, 4]. With the advancement in technology, the laparoscopic approach is becoming the standard surgical option owing to its several benefits, such as the shorter length of stay, decreased time to feeding, less blood loss, more negligible risk of complications, more excellent postoperative relief, improved cosmetic results, and faster recovery time [10, 11]. Nevertheless, the disadvantages of the laparoscopic method involve hemorrhage, pneumothorax, and aortic injury, resulting in a high rate of conversion to open surgery (9.1%) [12]. Robot-assisted surgery is another minimally invasive approach with optic enhancements and operator-based

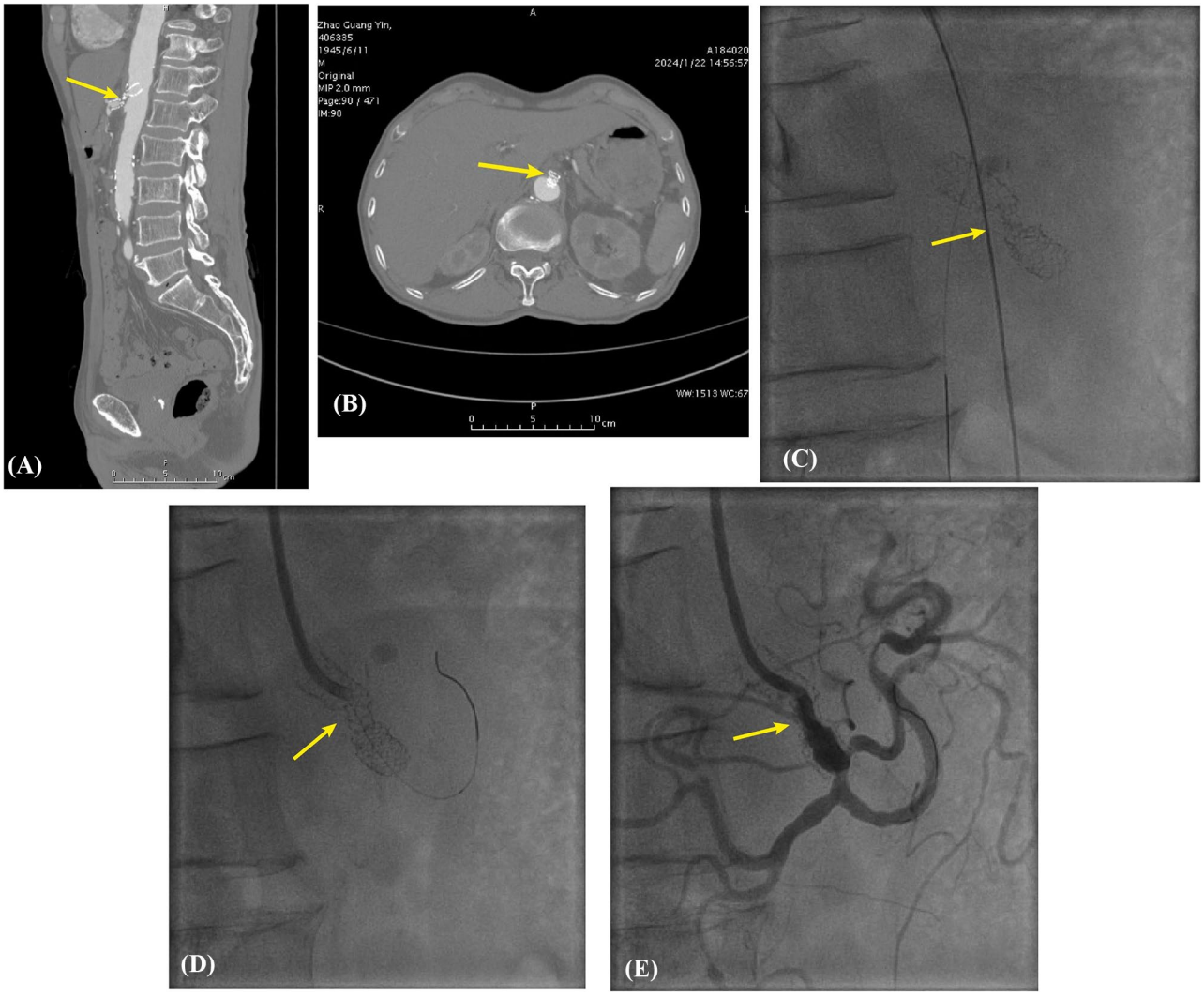

Fig. 3 Image of aorta showing the compressed and deformed stent in the ostial celiac artery. (**A**) CTA image of the aorta showing the stent in the ostial part of the celiac artery was compressed. (**B**) Axial image at the level of celiac artery origin showing the compression of the stent. (**C**) celiac angiogram showing the full compression and deformation of the middle part of the stent. (**D**) balloon angioplasty was performed in the stent by a 6.0*40 mm balloon. (**E**) celiac angiogram showing < 50% residual stenosis in the middle part of the stent after post-dilation

improvements, although it is limited by the longer operator time and increased cost [1]. When persistent celiac flow abnormalities after decompression are noted on intraoperative ultrasonography, vascular reconstruction, like celiac artery patch angioplasty, reimplantation of the celiac artery on the aorta, and aortoceliac bypass should be employed [1, 3]. In addition, percutaneous transluminal angioplasty (PTA) with or without stenting has proved helpful as an adjunctive therapy to MAL release. However, this procedure as sole management doesn't address the extrinsic compression of the celiac artery and has thus given a poor prognosis for patients with MALS [2].

In this report, we present a case of a 77-year-old male with MALS who isn't consistent with the classic demographic of young women with thin body habitus. Advanced age, presenting symptoms of abdominal pain, chest tightness and shortness of breath, and T-wave flattening at the anterior lead noted on the electrocardiogram, may promote the search for angina pectoris, although peptic ulcer disease, inflammatory bowel diseases, and gastrointestinal tumors were also suspected. Routine laboratory examination, ultrasound findings, and gastroenterological endoscope ruled out the digestive disease, so the abdominal pain was probably associated with angina pectoris, noted on the coronary angiography. However, the symptoms remained after PCI was successfully performed for LAD and LCX. Angiography revealed significant lesions in the celiac artery, so the revascularization was performed with symptom relief in the short term. However, after the recurrence of the abdominal pain, CTA showed the stent in the ostial celiac artery

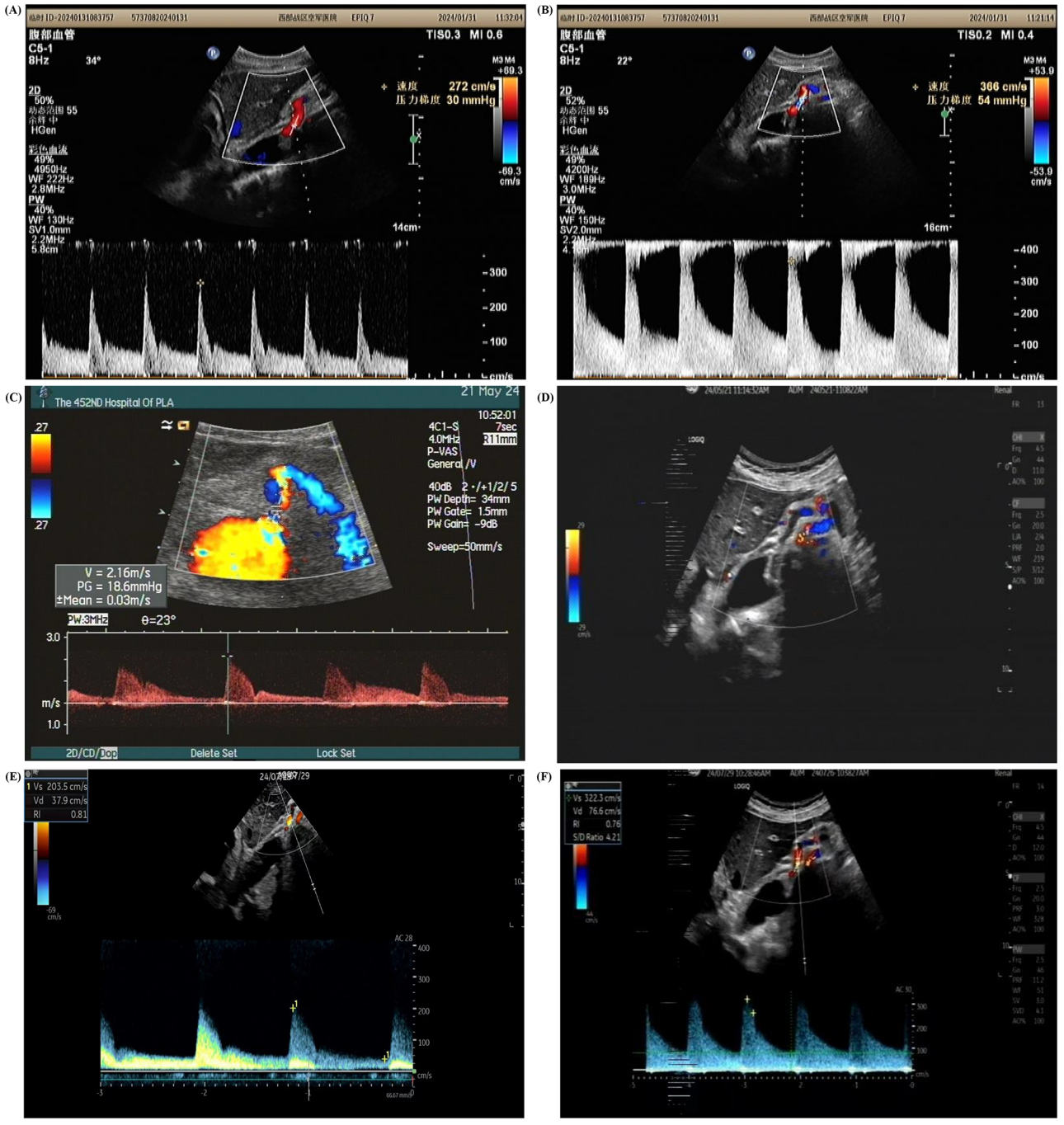

Fig. 4 DHS imaging showing the flow velocity with breathing maneuvers. DHS shows the flow velocity of 272 cm/s in inspiration (**A**) and 366 cm/s in expiration (**B**) 1 week postoperatively. DHS shows the flow velocity of 216 cm/s in inspiration (**C**) and 0 cm/s in expiration (**D**) at 4 months postoperatively. DHS shows the flow velocity of 203 cm/s in inspiration (**E**) and 322 cm/s in expiration (**F**) at 8 months postoperatively

was compressed and deformed caused by the MAL, which resulted in the obstruction of the vessel and thus led to intestinal ischemia and abdominal pain. Although the celiac artery decompression was not performed due to some limitations, symptom relief occurred immediately after the post-dilation and was maintained during the follow-up of 8 months. In this report, we hope the conditions should be thoroughly evaluated to avoid the missed diagnosis or misdiagnosis due to its unspecific clinical presentation and imaging manifestation. Furthermore, celiac artery decompression is effective for the treatment of MALS, while endovascular angioplasty alone is unsuccessful at achieving the resolution of the symptoms.

Conclusion

MALS is diagnosed only after extensive evaluation and exclusion due to its unspecific clinical manifestations and lack of consensus on a diagnostic criterion. Thus, such a disorder is prone to missed diagnosis or misdiagnosis. This case highlights the importance of keeping the condition as a differential diagnosis in patients with gastrointestinal symptoms. The primary treatment for MALS aims to decompress the celiac artery through the release of the MAL to relieve the symptoms.

Abbreviations

MALS	Median arcuate ligament syndrome
MAL	Median arcuate ligament
COPD	Chronic obstructive pulmonary disease
LAD	Left anterior descending
LCX	Left circumflex
RCA	Right coronary artery
CTA	Computed tomography angiography
BNP	B-type natriuretic peptide
PCI	Percutaneous coronary intervention
DHS	Duplex ultrasonography
MRA	Magnetic resonance angiography
DSA	Digital subtraction angiography
PTA	Percutaneous transluminal angioplasty

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Author contributions
WG, study concept and design; HQ and WG, drafting and finalization of the manuscript, preparation of the figures; ZY and YY, acquisition of data, and analysis and interpretation of data; HQ, YY and WG, critical revision of the manuscript for important intellectual content and material support.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethical approval
Case reports are exempt from ethical approval in our institution.

Informed consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Competing interests
The authors declare no competing interests.

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