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A variant of the median arcuate ligament syndrome: are sagittal images enough for diagnosis?

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Variante des Truncus-coeliacus-Kompressionssyndroms. Reichen sagittale Bilder für die Diagnose?

Zusammenfassung Das Truncus-coeliacus-Kompressionssyndrom ist eine seltene Ursache für abdominales Unbehagen, postprandialen Schmerz und Gewichtsabnahme. Um diese Anomalie zu identifizieren, können Doppler Ultraschall, Computertomographie, Magnetresonanz und selektive Katheterangiographie eingesetzt werden. Klassischerweise sieht man den „Angelhaken" auf den sagittalen Bildern. Bei unserem Fall sahen wir eine milde (20 %ige) Stenose am Ursprung der Arteria coeliaca ohne den typischen hakenförmigen Verlauf in der sagittalen Bildgebung. In der axialen Schnittführung konnte die Stenose als deutlich höhergradig (55 %ige) gezeigt werden. Wir stellen einen seltenen Fall eines Truncus-coeliacus-Kompressionssyndroms vor, welches durch prominente Crura des Diaphragmas bedingt war.

Schlüsselwörter: Truncus-coeliacus-Kompressionssyndrom, Multidetektor Computertomographie, Hypertrophie des Diaphragmas

Summary Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome, is a rare cause of abdominal discomfort, postprandial pain and weight loss. Doppler ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), and selective catheter angiography can be used to identify this abnormality. Classically, the "fish hook" appearance is seen on sagittal images. In our case, mild stenosis (20 %) was seen at the origin of the celiac artery without typical "fish hook" appearance. Higher degree stenosis (55 %) was demonstrated on axial images. We are presenting a rare case of a MALS caused by prominent crura.

Keywords: Median arcuate ligament syndrome (MALS), Celiac artery compression syndrome, Multidetector computed tomography (MDCT), Hypertrophy of diaphragm

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Introduction

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome, is a rare cause of abdominal discomfort, postprandial pain, and weight loss [1]. MALS occurs when the median arcuate ligament (MAL) and the origin of the celiac artery are in abnormally close proximity. The MAL is a fibrous band that connects the left and right diaphragmatic crura across the aortic hiatus at the level of the T12/L1 vertebral bodies. Compression and tethering of the celiac artery by MAL during respiration may result in damage to the celiac nerve plexus or partial obstruction of blood flow through the celiac artery and its distributary vessels. Doppler ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), and selective catheter angiography can be used to identify this abnormality. Treatment options involve minimally invasive laparoscopic techniques or open surgical division of the median arcuate ligament.

Case report

A 65-year-old man was referred to our clinic for the evaluation of abdominal pain and routine control of previously placed abdominal aorta stent by multidetector computed tomography (MDCT) angiography. No evidence of nausea or vomiting was reported. Laboratory tests were unremarkable. In medical history, patient was treated with stent implantation because of abdominal aorta aneurysm. No complications were identified on

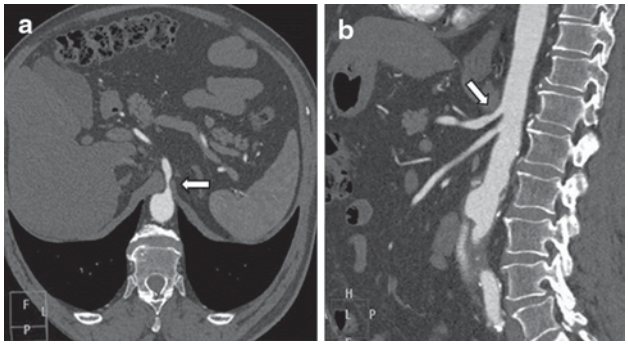

Fig. 1 Severe stenosis at the proximal segment of the celiac artery is best seen on axial reformatted multidetector computed tomography (MDCT) image (**a**). Hypertrophied diaphragmatic crura are also noteworthy. Sagittal MDCT angiogram shows mild stenosis at the proximal segment of the celiac artery (**b**) probably due to compression of the superior wall by median arcuate ligament

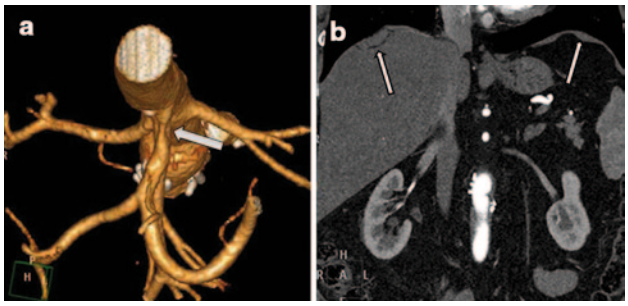

Fig. 2 3D multidetector computed tomography (MDCT) image best delineates external compression of the proximal segment of the celiac artery on both lateral sides (**a**). Coronal MDCT image reveals hypertrophy of the diaphragm on both sides (*arrows* in **b**)

follow-up examinations. MDCT angiography revealed a focal stenosis at the origin of the celiac artery with poststenotic dilation and also bilateral hypertrophy of diaphragm with prominent crura (Fig. 1a). On sagittal images, mild stenosis (20 %) was seen at the origin of the celiac artery without typical "fish hook" appearance (Fig. 1b). Higher degree stenosis (55 %) caused by prominent hypertrophied diaphragmatic crura was demonstrated on axial images. Narrowing of the origin of the celiac artery at both lateral sides was greatly assessed on 3D MDCT imaging (Fig. 2a). Hypertrophy of diaphragm was best demonstrated on coronal images as thick, prominent ondulations (Fig. 2b). Doppler USG imaging demonstrated moderate to severe (>50 %) celiac artery stenosis with poststenotic dilation, and increased blood flow (Fig. 3a, b). Peak systolic velocity of the celiac artery were measured as 143 and 153 cm/s during inspiration and expiration phases, respectively. The celiac artery–aortic ratio was 2.18. Patient was taken for follow-up and no treatment was performed.

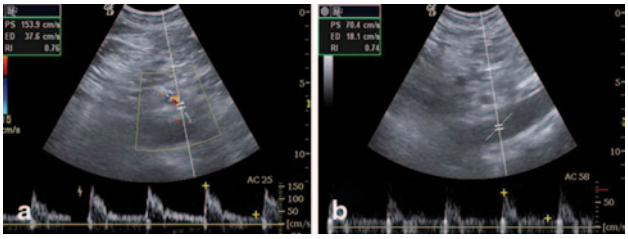

Fig. 3 Duplex Doppler image shows increased peak systolic velocity (153 cm/s) at the origin of the celiac artery (**a**) in expiration and normal peak systolic velocity (70 cm/s) at the abdominal aorta (**b**)

Discussion

MALS is thought to be a syndrome of abdominal pain caused by compression of the celiac trunk and encasement of the periarterial neural tissue by the MAL. MALS was initially described by Harjola [2] and Dunbar et al. [3], as a compression of the celiac artery thought to cause intimal fibrosis that leads to luminal stenosis and impaired splanchnic blood flow. Stenosis would result in symptoms similar to those of atherosclerotic mesenteric ischemia.

Clinical symptoms included postprandial epigastric pain, anorexia, nausea, vomiting, abdominal bruit, and weight loss [4]. The disease usually occurs in young slender patients with a female predominance. But in our case, patient is an adult obese male.

Classically, physical examination will reveal a bruit in the epigastric region that varies with respiration. The MAL is a fibrous band that connects the left and right diaphragmatic crura across the aortic hiatus at the level of the T12/L1 vertebral bodies. The celiac artery is compressed by the MAL with expiration. It descends in the abdominal cavity, resulting in a more vertical orientation in inspiration, which often relieves the compression.

In the past, MALS was detected by conventional angiography. A characteristic superior indentation is noted along the proximal celiac artery, usually about 5 mm from its origin at the abdominal aorta. Compression of the celiac artery increases during expiration and decreases during inspiration [5]. MDCT with 3D software have greatly improved the ability to obtain high-resolution images of the abdominal aorta and its branches. Typically, the sagittal plane is optimal for visualizing the proximal portion of the celiac artery [6]. In our case, we performed CT imaging in expiration and minimal compression was seen at the superior wall of the origin of the celiac artery. But on reformatted axial images, higher degree stenosis at the origin of the celiac artery was revealed. No fibrous or fibrocalcific atherosclerotic plaque formation was seen at proximal segment of the artery. In our case, we want to emphasize on the importance of the axial and especially axial reformatted MDCT images on diagnosis of the MALS. Doppler US imaging and MRI of celiac artery origin are also helpful to identify the MALS [7]. Lim et al. [8] revealed that peak sys-

tolic velocity of the celiac artery greater than 200 cm/s, and the celiac artery–aortic ratio greater than 3.0 during inspiration and expiration were significantly related to celiac artery stenosis. As Doppler USG is a real-time imaging, spectral changes occur during inspiration and expiration in the stenosis area. In our case, we observed high peak systolic velocities of the celiac artery with both inspiration and expiration due to greater degree compression of the artery at the lateral sides compared to the anterior side.

Surgical release of the MAL by laparoscopic approach and laparotomy are the most popular treatment choice. Laparoscopic approach is safer than laparotomy [9]. Alternative therapies are transluminal dilatation, stent placement, and celiac artery bypass surgery [10]. Because the etiology of MALS is not the luminal pathology, percutaneous transluminal angioplasty may not benefit and stent placement should not be undertaken because of the possible device fatigue.

In our case, no surgical or endovascular treatment were performed. Follow-up imaging was planned depending on the criteria (the celiac artery–aortic ratio <3 and peak systolic velocity <200 cm/s) based on the previous reports [8].

Conclusion

MDCT angiography plays a dominant role in the diagnosis of MALS especially with reformatted and 3D images. Beside the classical sagittal image interpretation, other axial, coronal, and reformatted images also have a significant role in the accurate diagnosis.

Conflict of interest

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in this manuscript.

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