Doppler ultrasound diagnosis of an unusual variant of median arcuate ligament syndrome: concomitant involvement of celiac and superior mesenteric arteries. A case report.

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Abstract:
Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome is a rare condition characterized by chronic mesenteric ischemia, secondary to the compression of the celiac artery by the median arcuate ligament. Occasionally, in addition to the celiac artery, the superior mesenteric artery may be partially compressed by the median arcuate ligament. We report a case with complaints of chronic abdominal pain from compression of both the celiac artery and the superior mesenteric artery due to MALS, which was primarily detected by Doppler ultrasound. The diagnosis was confirmed with CT-angiography.

Keywords: median arcuate ligament syndrome, celiac artery, superior mesenteric artery, hepatic artery, Doppler ultrasound

Introduction
Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome is an uncommon cause of chronic abdominal pain and weight loss, secondary to compression of the proximal part of the celiac artery by the median arcuate ligament. Rarely, in addition to the celiac artery, the superior mesenteric artery (SMA) may be partially affected by the extrinsic compression of the median arcuate ligament [1,2]. Here, we report a 30 year-old male with complaints of chronic abdominal pain from compression of both the celiac artery and the SMA due to MALS.

Case report
A 30 year-old male patient, with no past medical history, was admitted to the general surgery department with recurrent epigastric postprandial pain. The pain increased while lying down and caused fear of eating. The patient did not complain about weight loss. Nausea, with or without vomiting, was not reported. On physical examination, a mild abdominal tenderness at upper quadrants was noted. Initial laboratory investigations revealed normal values for complete blood count, erythrocyte sedimentation rate, liver and kidney function tests, amylase, and lipase. Endoscopy of the upper and lower gastrointestinal tract was normal. The patient was referred to the radiology department for abdominal ultrasonography.

Color Doppler ultrasonography demonstrated focal narrowing of the celiac artery with a turbulent flow at the origin. Gray-scale ultrasound examination excluded atherosclerotic arterial wall changes. The celiac trunk had a cranially deflected appearance. In the supine position, angle-adjusted peak-systolic and end-diastolic velocities within the celiac artery were significantly elevated during expiration (563.7 cm/sec) and inspiration (510.4 cm/sec). MALS was suspected and erect views were obtained: the celiac artery caliber returned to normal and peak-systolic velocities within the celiac artery decreased to 149 cm/sec, which was very suggestive for MALS (fig 1).

Considering the probability of the SMA to be influenced by the extrinsic compression of the median arcuate
ligament, angle-adjusted peak-systolic velocities within the SMA in the supine and erect position were measured: 252.6 cm/sec and 187.2 cm/sec, respectively, which was likely due to a tolerable extrinsic compression (fig 2a,b). Peak-systolic and end-diastolic velocities of the inferior mesenteric artery were within normal range.

In order to investigate secondary findings caused by the narrowing of the celiac artery, the flow pattern of the hepatic artery was also examined. Peak-systolic and end-diastolic velocities within the hepatic artery in the supine position were measured: 41.9 cm/sec and 22.5 cm/sec, respectively and spectral evaluation disclosed a significantly decreased resistive index (0.46), consistent with poststenotic flow changes (fig 2c).

CT angiography confirmed focal narrowing at the origin of the celiac artery with a characteristic hooked appearance for MALS and poststenotic dilatation. The stenosis did not appear to be associated with an atherosclerotic plaque. The proximal part of the SMA was involved as well, due to the partial extrinsic compressive effect of the median arcuate ligament (fig 3). No other vascular abnormalities were identified.

The patient was recommended a laparoscopic surgical division of the median arcuate ligament but he refused surgical treatment, and was followed up conservatively.

**Discussions**

MALS is a rarely encountered condition, which was initially described by Harjola [3] in 1963 and Dunbar et
al [4] in 1965. Patients are typically young (20-40 years) and thin women [5]. Signs may include abdominal pain, weight loss, nausea, diarrhea and fear of eating [6]. Physical examination of the mid epigastric region may reveal an audible bruit which varies with respiration.

The median arcuate ligament usually crosses the aorta superior to the origin of the celiac axis. However, in some individuals the median arcuate ligament inserts at a lower level, thus compressing the proximal portion of the celiac axis at varying degrees, causing the median arcuate ligament syndrome to occur [7]. Occasionally, the SMA may also be affected by the extrinsic compression of the ligament. Reilly et al published a case series in which 4 of the 51 patients with MALS had both celiac artery and superior mesenteric artery compression [8]. Typically, the compression exerted on the celiac artery becomes more pronounced with expiration and diminishes during inspiration. This is secondary to a physiologic anterior and inferior displacement of the aorta with inspiration, which usually relieves the constricting effect of the median arcuate ligament, whereas with expiration, this physiological mechanism runs in the exact opposite way [9]. However, in severe cases a persistent compression may be observed during deep inspiration [10]. When the patient stands up, the aorta descends further in the abdominal cavity, resulting in subsequent dissolution of the compression. The definition of the syndrome relies on a combination of both clinical and radiographic features. This is because it has been reported that angiographic studies may demonstrate compression of the celiac artery with varying degrees in approximately 13% to 50% of asymptomatic individuals [11].

For a long while, MALS has been disputed on grounds of its proposed but unproven pathophysiological mechanisms. The most-widely accepted pathophysiological concept behind MALS is the mesenteric ischemia secondary to the compression of the celiac artery by the atypically positioned median arcuate ligament. The mesenteric ischemia results either directly from foregut ischemia, or alternatively due to a celiomesenteric steal phenomenon from midgut ischemia [12,13]. But the causal relationships between the vascular stenosis and the symptoms remain still obscure. Another explanation for the symptoms is that the median arcuate ligament may impinge on the celiac artery as well as on the celiac splanchnic nerve plexus and it is suggested that the pain results from the involvement of the splanchnic nerve plexus [14].

In the presence of significant clinical manifestations, the diagnosis of the MALS is made traditionally by use of catheter angiography which demonstrates a characteristic indentation along the proximal celiac artery at its superior side which becomes more pronounced with expiration. The advance of multidetector CT technology has enabled identification of celiac artery compression by performing three-dimensional CT angiography. The characteristic focal narrowing of the proximal celiac artery with its typical hooked appearance can be helpful in distinguishing the MALS from other underlying causes of the celiac artery stenosis, such as atherosclerotic disease [6]. An additional modality that may be performed to achieve the diagnosis of MALS is MR angiography. Lack of need for application of iodinated contrast agents and lack of radiation exposure are its advantages over CT angiography, especially in the pediatric population [15].

In the present case, MALS was detected primarily by using Doppler ultrasound. Doppler US is an effective screening test, which provides quantitative evaluation of the abdominal aorta and its major branches such as celiac artery and SMA, also enables measuring in supine and erect positions during inspiration and expiration. The advantages of Doppler US over CT and catheter angiography are that it is noninvasive, easily accessible, and it does not expose patients to ionizing radiation or iodinated contrast agents. The usual protocol for evaluating the celiac artery should include both supine and erect views. Many authors state that increased expiratory-peak-systolic velocities measured within the compressed segment of the celiac artery, may not return to normal during inspiration with the patient in a supine position, which is consistent with our experience, as well. Therefore, it is recommended that quantitative evaluation of the celiac artery with the patient in an erect position to be added to the study. It is suggested that improvement of the peak-systolic velocities when the patient is placed in an erect position, allows the correct diagnosis of MALS to be made [10]. In cases of celiac artery stenosis, peak-systolic velocities measured within the celiac artery are elevated to greater than 200 cm/sec and the celiac artery-aortic ratio for peak-systolic velocities is greater than 3.0 during both inspiration and expiration [16].

While performing Doppler ultrasound to evaluate the celiac artery for MALS, it is important to look for a possible narrowing of the SMA and to obtain spectral Doppler analysis within the SMA. A peak systolic velocity greater than 275 cm/sec or no color flow within the SMA can be seen as an indicator of 70-100% percent stenosis [17]. End-diastolic velocities are also elevated in high grade stenosis and an end-diastolic velocity greater than 45 cm/sec is suggestive of ≥50% SMA stenosis [18]. In the present case, the values of peak-systolic and end-diastolic velocities in SMA met the criteria of SMA stenosis. To our knowledge, this is the first report in literature that examines concomitant compression of the celiac and the superior mesenteric arteries due to MALS accompanied by Doppler US findings.
We also recommend quantitative Doppler flow evaluation of the hepatic artery to be made which may reveal a poststenotic flow pattern secondary to the narrowing of the celiac artery. Especially when examining patients with limited acoustic access to the compressed part of the celiac artery due to air-filled gut or abdominal obesity, evaluation of the hepatic artery should be considered as a secondary source data before performing angiographic modalities. And again to the best of our knowledge there is no previous report in literature which gives information in MALS cases whether the flow pattern within the hepatic artery is affected or not.

As to the treatment of MALS, surgical release of the median arcuate ligament performed by a laparoscopic approach is a frequently preferred method [6]. Other treatment options involve transluminal dilatation, stent placement and celiac artery bypass surgery [19].

In conclusion, Doppler ultrasound, is the initial diagnostic modality for MALS, providing information about direct and indirect hemodynamics in the celiac and hepatic artery, respectively. Because of the possible rare concomitant involvement of SMA in MALS, this artery is an important part in the Doppler ultrasound protocol. The definite diagnosis could be achieved with CT angiography or diagnostic substraction angiography.

References