Ultrasonographic diagnosis of median arcuate ligament syndrome: a report of two cases

Alper Ozel¹, Guzide Toksoy¹, Osman Ozdogan², Abdullah Soydan Mahmutoglu¹, Zeki Karpat¹

¹ Radiology Department, Sisli Etfal Training and Research Hospital, Istanbul, Turkey
² Gastroenterology Department, Sisli Etfal Training and Research Hospital, Istanbul, Turkey

Abstract
The compression of the proximal part of the celiac trunk by median arcuate ligament of the diaphragm during expiration is defined as median arcuate ligament syndrome. It is a rare cause of chronic mesenteric ischemia. We report two cases with this syndrome, primarily diagnosed by Doppler ultrasound. The diagnosis was confirmed with digital subtraction and computed tomography angiography in both cases. The role of ultrasound in the diagnostic work up of this syndrome is discussed with regard to the recent literature.

Keywords: celiac artery, median arcuate ligament syndrome, Doppler ultrasound.

Introduction
Median arcuate ligament syndrome (MALS) is an uncommon cause of chronic mesenteric ischemia. In this syndrome, the proximal part of the celiac trunk is compressed by the median arcuate ligament of the diaphragm during expiration [1,2]. Eventually, the patients present with symptoms such as postprandial epigastric pain and weight loss. Here, we report two cases with MALS diagnosed primarily by ultrasonography.

Case 1
A 49 year-old man was admitted to the gastroenterology department of our hospital due to epigastric pain after meals and weight loss (5 kilos during the last six months). He was anterior investigated (abdominal ultrasound, upper and lower gastrointestinal endoscopy, and hemograms) in another medical center, but all the tests were within normal range. On physical examination, a mild epigastric tenderness and bruit was noted. He was referred to our radiology department for abdominal ultrasonography including mesenteric Doppler ultrasound. Real-time color Doppler examination of the abdominal aorta and its major branches was performed with the patient in supine position and during 8 hours fasting state. The celiac artery appeared narrowed and showed turbulent flow. Peak systolic and end diastolic velocities of the
during expiration- peak systolic and end diastolic velocities are elevated (308/81 cm/s); b) inspiration peak systolic and end diastolic velocities decrease to normal values (135/46 cm/s).
celiac artery with angle correction performed on deep expirium were 308 cm/s and 81 cm/s, respectively. On maximum inspirium, peak and end-diastolic velocities decreased to 135 cm/s and 46 cm/s, respectively (fig 1). MALS was suspected, and abdominal computed tomography (CT) with angiography was performed. CT angiograms showed a focal narrowing with “hooked” appearance on the superior surface of the celiac artery (fig 2). The patient was treated conservatively.

Case 2

A 47 year-old woman complaining of abdominal pain and weight loss was referred to radiology department for mesenteric Doppler investigation. The Doppler examination performed on supine position with the patient on fasting state, showed increased peak systolic and end-diastolic velocities on both inspiration and expiration. On deep inspiration peak systolic and end-diastolic velocities were measured 276 cm/s and 134 cm/s, respectively. Peak systolic and end-diastolic velocities on expiration were 430 cm/s and 199 cm/s, respectively (fig 3). When the patient was told to stand up, both peak systolic and end-diastolic velocities were measured again. In an erect position, these values returned to normal (fig 4). Diagnostic subtraction angiography of the aorta on lateral projection showed a concave stenosis on the superior part of the proximal celiac trunc, and MALS was diagnosed.

Discussion

MALS was first described by Harjola and Dunbar et al. in 1963 and 1965, respectively [3,4]. The characteristic clinical triad include postprandial pain, diarrhea and weight loss. The median arcuate ligament is a band of fibrous tissue that crosses over the aorta, usually above the origin of the celiac artery. In patients with the MALS, the celiac artery is compressed by the median arcuate ligament during expiration [5]. However, it has been reported that approximately 13% to 50% of asymptomatic individuals would have, to a variable degree, compressive features of the celiac artery at their angiographic studies especially during expiration [6,7]. Therefore, the debate on the use of the term MALS continues and it is generally used when the compression of the celiac artery by the median arcuate ligament causes a significant clinical picture.

The pathophysiology of the MALS is not yet fully elucidated. One explanation of the MALS emphasizes the mesenteric ischemia due to celiac artery compression.
Mesenteric ischemia results either directly from foregut ischemia or, alternatively from midgut ischemia [8,9], which is caused through postprandial setal via collaterals from the superior mesenteric artery to the celiac bed. The other explanation of the symptoms of the MALS is compression of the celiac ganglion [10].

The diagnosis of the MALS is made traditionally by the catheter angiography, in which a characteristic superior indentation is noted along the proximal part of the celiac trunc, which becomes more pronounced on expiration [2,11]. With the advent of and use of multidetector CT technology, the ability to obtain high resolution images of the abdominal aorta and its branches. In consequence of this, several cases of the MALS with CT have been reported in the last decade [11,12]. CT angiography demonstrates a characteristic focal narrowing in the proximal celiac axis. This focal narrowing has a characteristically hooked appearance, which can be useful in differentiating MALS from other causes of celiac artery stenosis such as atherosclerosis [13].

Doppler ultrasound can be used as a diagnostic test for the MALS. The advantages of Doppler US over catheter angiography and CT are that it is noninvasive, less expensive and does not expose patients to ionizing radiation or iodinated contrast. The diagnosis of MALS can be achieved with the Doppler flow measurements through the celiac artery made during inspiration and expiration. Doppler flow velocity measurements made at the compressed segment of the celiac trunc reveals variation of peak systolic velocity during respiration with a marked increase during expiration in PSV to greater than 200 cm/s. A greater than 3:1 ratio of PSV in the celiac artery in expiration compared with the PSV in the abdominal aorta immediately below the diaphragm is another criterion to diagnose MALS [14,15]. However, some authors have suggested that increased PSV’s may be encountered during both inspiration and expiration at Doppler examinations with the patient in a supine position. They emphasized that with the additional of erect views the PSV’s returned to normal and that allowed the correct diagnosis of the MALS to be made [15]. In our second case, the peak systolic velocity decreased substantially on inspiration than on expiration, but it still remained high. To increase the specificity of our diagnosis, we examined the patient in an erect position and found the PSV at celiac trunc within the normal reference velocity levels. We believe that velocity measurements at the compressed segment of the celiac trunc could be inconsistent due to rapid respiratory changes. Therefore, it seems to be more valuable, that the patient is re-examined also in an erect position after supine examination.

In a recent article, Gruber et al. proposed that functional ultrasound, which demonstrates flow changes of the celiac trunc and functional-geometric changes such as celiac trunc deflection, should be the first line in screening for MALS. Albeit the small number of patients in their study cohort, using peak systolic expiratory flow velocity of the celiac artery greater than 350 cm/s and celiac trunc deflection angle greater than 50°, they found a sensitivity of 83% and a specificity of 100% in differentiating MALS from volunteers [16].

The treatment options for MALS include surgical or laparoscopic division of the median arcuate ligament, celiac ganglion destruction and bypass surgery [10].

In conclusion, MALS is a rare cause of chronic mesenteric ischemia, which is caused through compression of the proximal celiac trunc by median arcuate ligament of the diaphragm during expiration. Doppler ultrasound is an useful modality in the diagnostic work-up of patients with MALS. It provides the initial diagnosis through Doppler measurements of the celiac artery during both phases of respiration, and during sitting and erect positions. The definite diagnosis of this rare condition could be achieved with CT angiography or diagnostic subtraction angiography.

References