Case report

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

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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 substraction 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 trunc 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 peformed 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

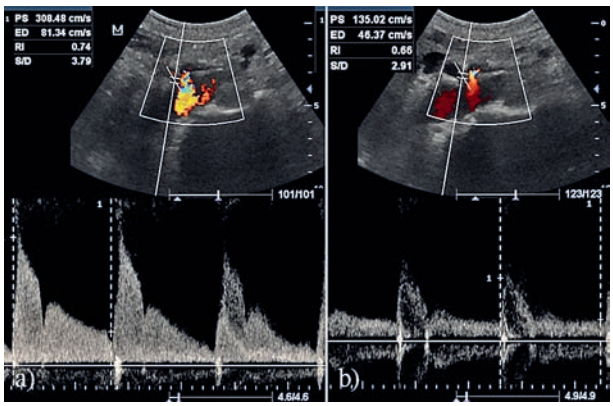

Fig 1. Doppler spectral images of the proximal celiac trunc a) during expiration- peak systolic and end diastolic velocities are elevated (308/81 cm/s); b) inspiration peak systolic and end diastolic velocities decrease to normal values (135/46 cm/s).

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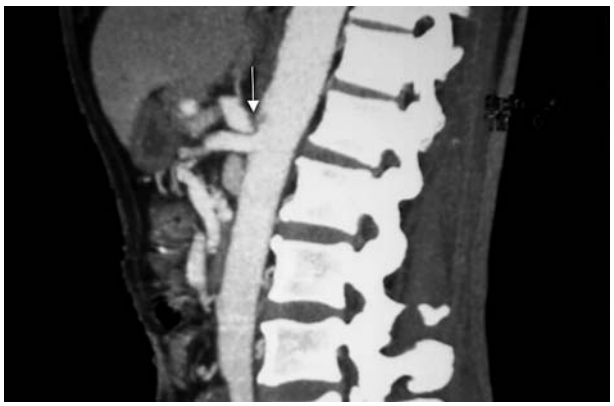

Fig 2. Sagittal maximum intensity projection (MIP) image of the CT angiogram shows significant narrowing "hooked appearance" (arrow) along the superior aspect of the proximal portion of the celiac artery.

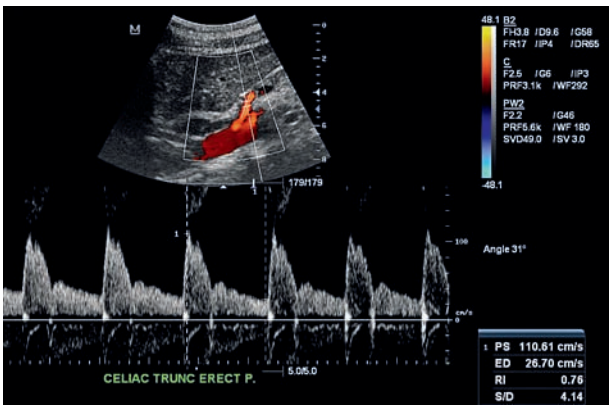

Fig 3. Doppler spectral images of the proximal celiac trunc during both phases of respiration. Peak systolic and end diastolic velocities are markedly elevated with a) inspiration (276/134 cm/s); b) and expiration (430/199 cm/s).

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 velocites 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.

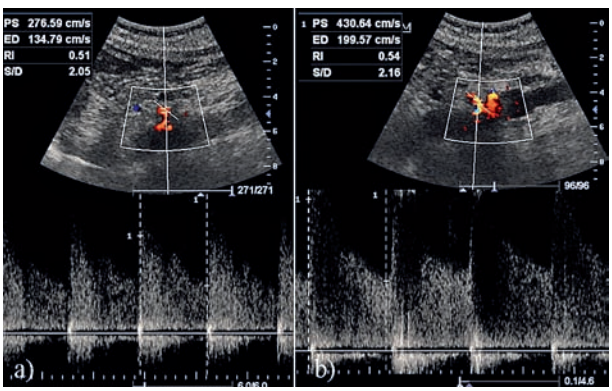

Fig 4. Peak systolic and end diastolic velocities within the celiac artery return to normal values with the patient in an erect position (110/26 cm/s).

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 substraction 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 or ischemia 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 artery 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 addition 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 substraction angiography.

References

1. Alehan D, Dogan OF. Pediatric surgical image. A rare case: celiac artery compression syndrome in an asymptomatic child. J Pediatr Surg 2004; 39: 645–647.
2. Ogino H, Sato Y, Banno T, Arakawa T, Hara M. Embolization in a patient with ruptured anterior inferior pancreaticoduodenal arterial aneurysm with median arcuate ligament syndrome. Cardiovasc Intervent Radiol 2002; 25: 318–319.
3. Harjola PT. A rare obstructıon of the coeliac artery. Report of a case. Ann Chir Gynaecol Fenn 1963; 52: 547–550.
4. Dunbar JD, Molnar W, Beman FF, Marable SA. Compression of the celiac trunk and abdominal angina. Am J Roentgenol Radium Ther Nucl Med 1965; 95: 731–744.
5. Lindner HH, Kemprud E. A clinicoanatomic study of the arcuate ligament of the diaphragm. Arch Surg 1971; 103: 600–605.
6. Szilagyi DE, Rian RL, Elliott JP, Smith RF. The celiac artery compression syndrome: does it exist? Surgery 1972; 72: 849–863.
7. Bron KM, Redman HC. Splanchnic artery stenosis and occlusion. İncidence; arteriographic and clinical manifestations. Radiology 1969; 92: 323–328.
8. Bech FR. Celiac artery compression syndromes. Surg Clin North Am 1997; 77: 409–424.
9. Tribble CG, Harman PK, Mentzer RM. Celiac artery compression syndrome. Report of a case and rewiev of current opinion. Vasc Surg 1986; 20: 120–129.

10. Loukas M, Pinyard J, Vaid S, Kinsella C, Tariq A, Tubbs RS. Clinical anatomy of celiac artery compression syndrome: a review. Clin Anat 2007; 20: 612–617.
11. Kopecky KK, Stine SB, Dalsing MC, Gottlieb K. Median arcuate ligament syndrome with multivessel involvement: diagnosis with spiral CT angiography. Abdom Imaging 1997; 22: 318–320.
12. Dordoni L, Tshomba Y, Giacomelli M, Jannello AM, Chiesa R. Celiac artery compression syndrome: successful laparoscopic treatment – a case report. Vasc Endovascular Surg 2002; 36: 317-321.
13. Horton KM, Talamini MA, Fishman EK. Median arcuate ligament syndrome: evaluation with CT angiography. Radiographics 2005; 25: 1177–1182.
14. Erden A, Yurdakul M, Cumhur T. Marked increase in flow velocities during deep expiration: A duplex Doppler sign of celiac artery compression syndrome. Cardiovasc Intervent Radiol 1999; 22: 331–332.
15. Wolfman D, Bluth EI, Sossaman J. Median arcuate ligament syndrome. J Ultrasound Med 2003; 22: 1377–1380.
16. Gruber H, Loizides A, Peer S, Gruber I. Ultrasound of the median arcuate ligament syndrome: a new approach to diagnosis. Med Ultrason 2012; 14: 5–9.