

Median Arcuate Ligament Syndrome Confirmed With Vascular Ultrasound

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Abstract:

Introduction: The median arcuate ligament syndrome (MALS) is a rare abdominal vascular compression condition of the coeliac trunk caused by a fibrous ligamentous band that, in different phases of respiration, restricts blood flow to the gastrointestinal tract, resulting in a clinical presentation of abdominal pain, mostly postprandial, and weight loss.

Case report: We report a case of a female patient with chronic epigastric pain which was initially investigated by computed tomography angiography (CTA) with ambiguous result. On duplex scan, a non-atherosclerotic stenosis with a hook-like appearance was identified, causing exacerbated velocity increase in expiration at the coeliac trunk origin. Disturbed flow was also noted distally into the common hepatic and splenic artery.

Discussion: Duplex scan has the advantage of assessing the appearance and haemodynamics of the coeliac trunk at different phases of the respiratory cycle and with patient in either supine or orthostatic position, to identify changes in blood flow velocities with the median arcuate ligament compression relieve.

Conclusion: Vascular ultrasound is a first line imaging modality to diagnose MALS, which was confirmed in this case with the haemodynamic information of the duplex scan investigation.

Keywords: Median arcuate ligament syndrome, Dunbar syndrome, coeliac stenosis, Doppler Ultrasound.

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I. Introduction

The median arcuate ligament syndrome (MALS), also called Dunbar syndrome, is defined as a combination of both clinical and imaging features which typically occurs in adolescents and young adults, being more common in thin women and has an incidence between 1.8-4.0% [1]. It is caused by an extrinsic compression of the coeliac trunk (CoeT) with typical clinical presentation including frequent epigastric pain, nausea and vomiting resulting in weight loss. It has first been described by Harjola in 1963 and Dunbar *et al.*, in 1965 on their study of 15 symptomatic patients that underwent conventional angiography, with 13 being submitted to surgery which resulted in a relief of their symptoms [2].

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes superior to the origin of the CoeT however in 10-24% of healthy people it may be atypically low positioned, crossing over its proximal segment and causing a characteristic indentation. In some cases, the ligament can externally compress the CoeT to the point of compromising blood flow causing symptoms which can be exacerbated in expiration. While abdominal pain may be associated with eating, this is not always described which makes clinical diagnosis more challenging. A physical examination may demonstrate an audible abdominal bruit in the mid epigastric region, originated from the turbulent flow in the compressed CoeT, that varies with respiration [3].

Angiography and computed tomography angiography (CTA) are considered the gold standard diagnostic methods for MALS, with possibility to create three-dimensional reconstructions. A focal stenosis of the origin of the CoeT with the vessel demonstrating a hook-like appearance and the absence of arterial calcification, to differentiate from atherosclerosis, are the typical characteristics for MALS diagnosis. Doppler ultrasound has also been considered one of the main diagnostic exams and best initial test to screen for MALS due to its non-invasiveness, safety and haemodynamic information [4].

II. Case Report

A 53 year old woman presented to the vascular clinic with regards to chronic epigastric pain. Discomfort was constant and not specific to an action or time of day, whilst the patient reported large weight loss. Previous gastroscopy and colonoscopy were completed as normal and CTA imaging performed two years earlier did not show any abnormality, however, a recent repeat CTA was suggestive of a CoeT stenosis (Figure

1). A duplex vascular ultrasound imaging of the abdominal aorta and the visceral arteries was then requested by the vascular surgery clinic. On duplex scan, B-mode imaging demonstrated obstruction of the CoeT origin, which was not atherosclerotic in nature, but rather a median arcuate ligament compression of the CoeT with a typical hook-like appearance (Figure 2). Colour Doppler demonstrated narrowing with notable aliasing associated with stenosis (Figure 3) whilst spectral Doppler demonstrated elevated velocities, both in inspiration and expiration, with peak systolic velocities (PSV) of 342cm/s at the CoeT origin (Figure 4) and above 400cm/s more distally. Beyond the origin of the CoeT Doppler waveforms showed disturbed haemodynamics, with turbulent and mildly damped waveforms into the common hepatic artery (Figure 5) and splenic artery (Figure 6).

The patient was placed on a low FODMAP diet by the multi-disciplinary team, in light of her co-existing irritable bowel syndrome. As the patient's weight was now stable and the pain was managed well with analgesics, the decision was made not to pursue vascular surgical options for the time being and to review her in six months' time.



Figure 1 (left):CTA imaging of coeliac trunk hook-like stenosis (yellow arrow). **Figure 2** (right): B-mode image demonstrating a non-atherosclerotic obstruction of the coeliac trunk origin (yellow arrow), suggesting median arcuate ligament compression with a hook-like appearance

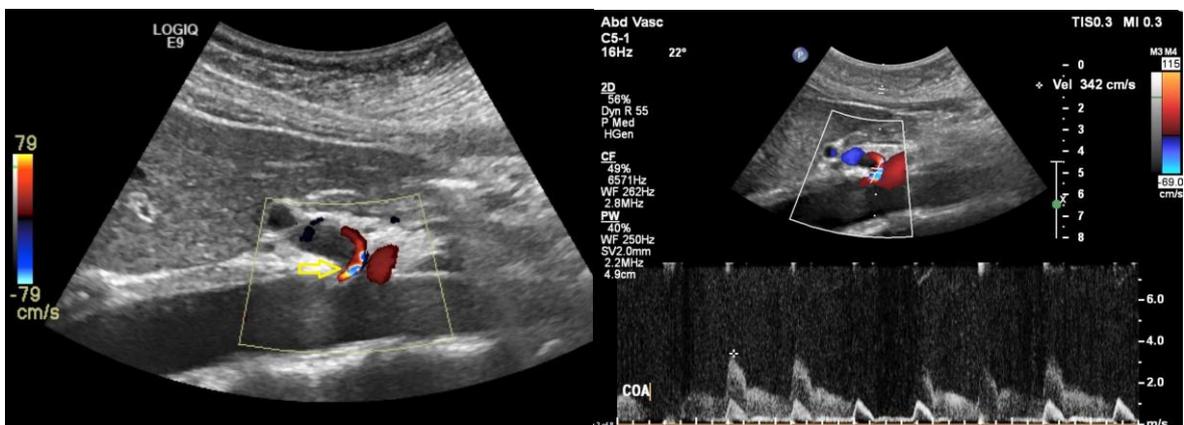


Figure 3 (left): Colour Doppler demonstrated narrowing with notable aliasing associated with stenosis of the coeliac trunk (yellow arrow). **Figure 4** (right): Spectral Doppler demonstrating elevated velocities (PSV of 342cm/s) at the coeliac trunk origin.

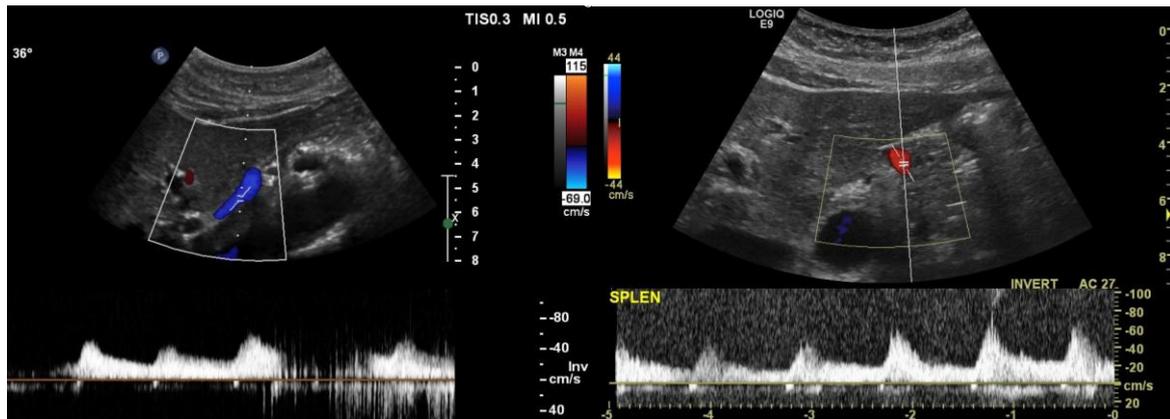


Figure 5 (left) and **Figure 6** (right): Spectral Doppler demonstrating damped waveforms in the common hepatic artery (left) and in the splenic artery (right).

III. Discussion

The pathophysiology of MALS is not fully understood. A variable degree of compression of the CoeT has been identified in 13-50% of asymptomatic individuals in angiographic studies, mainly detectable in expiration [5]. A possible explanation may be that the ischemia resulting from the CoeT compression causes a concomitant mesenteric ischemia. This is due to postprandial superior mesenteric artery (SMA) flow being collateralised towards the coeliac vascular bed territory which results in foregut or midgut ischemia [6]. Because of this possible associated involvement of the SMA, Akan *et al.*, suggested that this artery should also be included in the duplex scan protocol assessment of MALS [7]. Another accepted explanation implicates the compression or ischaemia of the coeliac ganglion.

The treatment possibilities for patients suffering from MALS consist of surgical or laparoscopic division of the median arcuate ligament, coeliac ganglion removal and bypass surgery [5].

A typical hook-like appearance on CTA and similarly as a membrane in ultrasound is a distinctive feature to characterise MALS. The resulting focal narrowing can be distinguished from atherosclerosis which is another differential cause for CoeT stenosis but should not be mistaken with a dissection on B-mode imaging. The sonographer must pay attention to its anatomical positioning, integrity of the aorta and absence of flap.

Diagnosing MALS with ultrasound has the advantage of assessing haemodynamics at the narrowed segment of the CoeT at different phases of the respiratory cycle, and with patient in either supine or orthostatic position. It has been observed that different velocities may be obtained according to the respiratory cycle, where higher velocities are typically seen in expiration. A small study cohort conducted in 2012 by Gruber *et al.*, proposed that a reliable ultrasound diagnostic criteria to define subjects with high probability of suffering from MALS was a combination of maximum expiratory PSV $>350\text{cm/s}$ and a functional-geometric deflection angle of the CoeT greater than 50° , obtaining a sensitivity of 83% and a specificity of 100% in differentiating MALS from volunteers [8]. Additionally, a 3:1 PSV ratio between the CoeT and the proximal abdominal aorta (immediately below the diaphragm) during either inspiration or expiration may also indicate stenosis [9]. A larger study performed in 2012 by AbuRahma *et al.*, including 151 SMA and 150 CoeT, proposed that a PSV $\geq 240\text{cm/s}$ would indicate $\geq 50\%$ stenosis and PSV $\geq 320\text{cm/s}$ a $\geq 70\%$ stenosis [10].

As some authors have suggested that raised velocities can be detected during both inspiration and expiration with the patient in a supine position, Wolfman *et al.*, suggested that the CoeT should also be assessed with the patient in orthostatic position. A more vertical orientation of the CoeT often relieves the compression caused by the median arcuate ligament and reverts velocities to normal range as the CoeT descends into the abdominal cavity during inspiration and even further in orthostatic position [9]. Therefore, enhanced confidence in diagnosing MALS can be attained if PSV returns to normal with patients examined while standing.

In this case, the duplex scan was able to provide anatomical and haemodynamic information and was more clarifying than CTA. Despite our patient only being assessed in supine position, severely raised velocities were detected during both respiration phases, being more exacerbated in expiration. The CoeT to aorta ratio of 4:1 was obtained as the abdominal aorta PSV was 105cm/s . Additionally, there was clear B-mode imaging suggestive of CoeT origin narrowing caused by a membrane like appearance instigating the compression which resulted in the increased velocities. This caused the subsequent disturbed post-stenotic flow observed distally in the CoeT and becoming mildly damped towards the common hepatic artery and splenic artery. This may be in keeping with a possible collateralisation requirement from the SMA, as this widely patent and unobstructed vessel had raised PSV $>300\text{cm/s}$. Nonetheless, an orthostatic assessment is recommended as part of a MALS duplex scan protocol, particularly when all criteria are not met or are confounding.

IV. Conclusion

Vascular ultrasound constitutes an important and first line imaging modality to diagnose MALS. It allows a non-invasive haemodynamic and functional assessment of the CoeT during both phases of respiration, as well as in distinct patient positioning, which can help diagnostic confidence and guide treatment options. Additionally, assessment of the CoeT branches and SMA should also be performed in order to obtain a full spectrum of the haemodynamic repercussions of this condition.

References

- [1] R. Tembey, A. Bajaj, P. Wagle, and A. Ansari, "Real-time ultrasound: Key factor in identifying celiac artery compression syndrome" *Indian J. Radiol. Imaging*, vol. 25, no. 2, p. 202, 2015.
- [2] P.T. Harjola, "A rare obstruction of the coeliac artery. Report of a case" *Ann. Chir. Gynaecol. Fenn.*, vol. 52, pp. 547–550, 1963.
- [3] K. M. Horton, M. A. Talamini, and E. K. Fishman, "Median Arcuate Ligament Syndrome: Evaluation with CT Angiography" *RadioGraphics*, vol. 25, no. 5, pp. 1177–1182, 2005.
- [4] T. Sunkara, M. E. Caughey, K. C. Zhen, B. Chiong, and V. Gaduputi, "Dunbar syndrome-A rare cause of foregut ischemia" *J. Clin. Diagnostic Res.*, vol. 11, no. 7, pp. OD13–OD14, 2017.
- [5] A. Ozel, G. Toksoy, O. Ozdogan, A. S. Mahmutoglu, and Z. Karpat, "Ultrasonographic diagnosis of median arcuate ligament syndrome: A report of two cases" *Med. Ultrason.*, vol. 14, no. 2, pp. 154–157, 2012.
- [6] Bech FR., "Celiac artery compression syndromes" *Surg. Clin. North Am.*, vol. 77, no. 2, pp. 409–424, 1997.
- [7] D. Akan, A. Ozel, O. Orhan, E. Bozdog, and M. Basak, "Doppler ultrasound diagnosis of an unusual variant of median arcuate ligament syndrome: concomitant involvement of celiac and superior mesenteric arteries. A case report" *Med. Ultrason.*, vol. 17, no. 4, pp. 557–560, Dec. 2015.
- [8] H. Gruber, A. Loizides, S. Peer, and I. Gruber, "Ultrasound of the median arcuate ligament syndrome: a new approach to diagnosis" *Med. Ultrason.*, vol. 14, no. 1, pp. 5–9, 2012.
- [9] D. Wolfman, E. Bluth, and J. Sossaman, "Median Arcuate Ligament Syndrome" *J Ultrasound Med*, vol. 22, pp. 1377–1380, 2003.
- [10] A. F. AbuRahma *et al.*, "Mesenteric/cealic duplex ultrasound interpretation criteria revisited" *J. Vasc. Surg.*, vol. 55, no. 2, pp. 428–436, 2012.

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