

Case Report

A case report on median arcuate ligament syndrome: Need for a high index of suspicion

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ABSTRACT

The Median Arcuate Ligament Syndrome (MALS) is a rare clinical entity occurring due to abnormally low insertion of the ligament across the origin of the coeliac axis resulting in varying degrees of celiac artery compression. Its symptoms mimic those of chronic mesenteric ischemia and its diagnosis requires a high index of clinical suspicion coupled with appropriate imaging. We report the case of a young male with pain abdomen, nausea, and vomiting in whom routine evaluation for pain abdomen was inconclusive. He was diagnosed with MALS after multi-modality imaging. The patient was suspected to have MALS by using ultrasound and color Doppler imaging. These findings were confirmed using Computed tomography angiography (CTA) of the abdomen. MALS should be considered as an important differential in patients with symptoms resembling post-prandial angina. A combination of a high index of suspicion with the use of appropriate imaging techniques can clinch this diagnosis. We describe the diagnostic challenges faced in this case with a brief review of the literature.

Keywords: Median arcuate ligament syndrome, Celiac artery constriction, Low insertion of median arcuate ligament, Abdominal angina, Dunbar syndrome

INTRODUCTION

The median arcuate ligament is a fibrous arch under the diaphragm that connects both the crura of the diaphragm. This arch usually shows variations in the level of insertion but in a majority of individuals, it is inserted at L1 vertebral level and is cranial to the origin of the celiac trunk. In some individuals, it has an abnormally low insertion across the origin of the coeliac axis and can result in varying degrees of celiac artery compression. This can be further exacerbated by the abnormally high origin of the celiac trunk.^[1] This is known as median arcuate ligament syndrome (MALS). This condition is also known as celiac artery compression syndrome, Dunbar syndrome, or Harjola-Marable syndrome.

A small number of patients with MALS show symptoms, which are similar to those of chronic mesenteric ischemia. The best method of the evaluation of this entity is CT with three-dimensional reconstruction where the low insertion of the median arcuate ligament, the high origin of the celiac trunk, and the hook or J-shaped indentation on the celiac artery, all can be demonstrated.^[2] Hemodynamically significant compression of the celiac trunk can lead to post-

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stenotic dilatation of the celiac artery and formation of collaterals from the superior mesenteric artery (SMA). The diagnosis can be suspected on the Doppler which reveals raised peak systolic velocities in the celiac artery (>200 cm/s) and velocity change in various phases of respiration along with a drop on erect scans.^[3] It is usually a challenging diagnosis that requires a high index of clinical suspicion followed by the use of appropriate imaging modalities.

We present the case of a young male who presented with pain abdomen, nausea, and frequent episodes of vomiting, and on evaluation which was found to have MALS. This case illustrates the classic clinical and imaging findings of this condition with an emphasis on the need for a high index of suspicion. This case is also unusual due to the young age of the patient.

CASE REPORT

An 18-year-old man with no significant medical or surgical history presented to the outpatient department of the gastroenterology department of our tertiary care center with an 8-month history of intermittent epigastric pain. The pain was acute in onset, moderately severe in intensity, and non-radiating. It was associated with nausea, non-bilious vomiting, and bloating. The pain used to aggravate after taking meals and got relieved with bowel rest. This phenomenon was more common with fatty meals. He had no alterations of bowel habits. He gave a history of 7 kg weight loss in the past 3 months due to these symptoms. He did not give any history of significant medical conditions in the past. There was no history of alcohol or tobacco consumption or drug use. The family history was non-contributory.

On physical examination during the episode of pain, he only had mild epigastric region tenderness. There was no organomegaly. Bowel sounds were normal. His routine hematological and biochemical parameters were within normal limits. Esophagogastroduodenoscopy (EGD) showed

a small hiatal hernia with no mucosal erosions. The tissue samples from the stomach were negative for *Helicobacter pylori*.

The patient underwent B-mode ultrasound imaging, color Doppler, and spectral Doppler ultrasound examination of the mesenteric vessels due to clinical suspicion of MALS. The B-mode ultrasound images showed kinking of the celiac trunk origin with a dilated segment distal to the stenosed part. The color Doppler of this vessel showed color aliasing [Figure 1] at the origin of the celiac trunk which was exacerbated in the supine position. Spectral Doppler examination revealed the elevated peak systolic velocity (PSV) of around 200 cm/s (normal – 90–100 cm/s) at inspiration which was further augmented at expiration to around 350 cm/s. The higher velocities in the celiac trunk are a marker of stenosis and a velocity of more than 200 cm/s is consistent with approximately 70% stenosis. On erect scanning, the velocity dropped to around 205 cm/s in expiration. The spectral Doppler of SMA showed no abnormality. Based on these ultrasound findings, we suspected the diagnosis of MALS.

The patient underwent a computed tomography angiography of the abdomen to confirm the diagnosis of MALS. The CT protocol consisted of an arterial phase at 35 s duration and a venous phase at 65 s. A 16-slice multidetector CT (Somatom Emotion, Siemens AG, Erlangen, Germany) was used. Coronal, sagittal, and axial reconstructions of the celiac artery [Figure 2] revealed a focal constriction at the origin of the celiac trunk associated with dilatation of the post-stenotic segment. The thickness of the median arcuate ligament on axial images was 6 mm against a normal range of up to 4 mm.^[3] These findings confirmed the diagnosis of MALS.

The patient was taken up for laparoscopic surgery. The surgical findings confirmed the diagnosis. Surgical decompression was performed by dividing the median arcuate ligament.

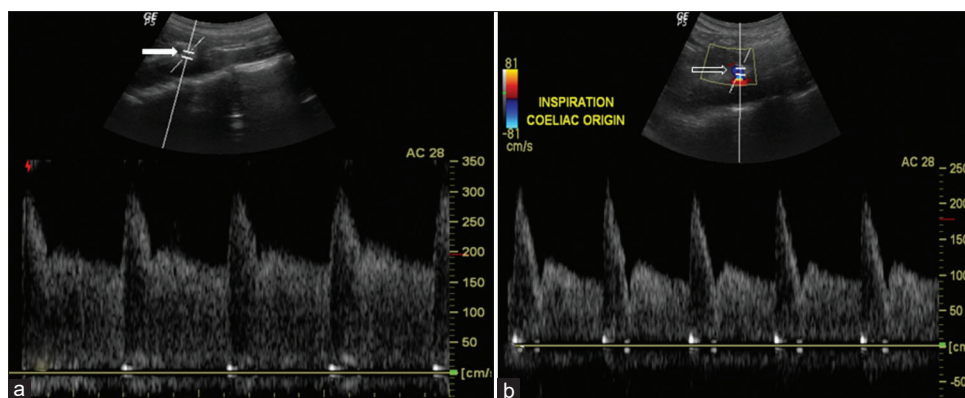


Figure 1: Spectral Doppler images with Doppler gate at the origin of celiac trunk in expiration (a) and in inspiration (b) show raised peak systolic velocity in celiac trunk which partially reduces on inspiration.

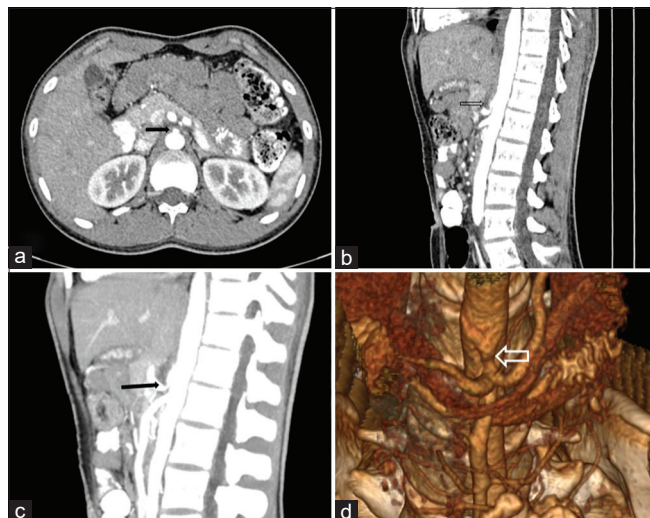


Figure 2: Computed tomography angiography images from arterial phase study. (a) Image in axial plane shows the focal narrowing at the origin of the celiac trunk (solid black arrow). (b) Sagittal reformatted and (c) maximum intensity projection image in sagittal plane show “hook-like” narrowing (solid black arrow in c) at the origin of the celiac trunk due to indentation by median arcuate ligament. (d) A three-dimensional volume rendered image illustrates the narrowing at the origin of celiac trunk (hollow white arrow) when viewed from anterosuperior direction.

DISCUSSION

The median arcuate ligament is a junction of the right and left crura of the diaphragm. The crura connect to form an arch just anterior to the aortic hiatus.^[4] Abnormally low insertion of this ligament can lead to MALS. It was first described in 1963^[5] when a patient with abdominal pain and epigastric bruit was found to have thick fibrous tissue encasing the celiac artery at the surgery.

The suffered group is usually young lean females between the age of 30 and 50 years and present with abdominal pain.^[6] Pain is located in the epigastric area and worsens after food intake, exercise, or on leaning forward.^[7] The pain is also associated with nausea, vomiting, bloating sensation, and passage of loose stools. Some patients may experience food fear and hence suffer from significant weight loss^[8] as seen in our case. Patients may get transient symptom relief by bringing their knees close to their chest. This position moves the median arcuate ligament cephalad to the artery and causes partial relief of the impingement.

Physical examination usually reveals tenderness in the epigastric region and may reveal epigastric region bruit in a significant number of patients (83%)^[9] which may increase on expiration. The patient typically undergoes extensive workup to find the etiology of abdominal pain.

The workup should exclude hepatobiliary pathology and gastric or duodenal ulcer disease because MALS is a

diagnosis of exclusion. These pathologies can be ruled out by abdominal ultrasound, EGD, and gastric emptying studies.

Ultrasound along with color and spectral Doppler imaging has emerged as a non-invasive, real-time, and dynamic screening tool that can increase diagnostic confidence. It shows raised (>200 cm/s) PSV at the origin of the celiac trunk which shows further increase on expiration and fall on assuming erect posture. Doppler imaging can also help in excluding atherosclerotic affliction at the origin of the celiac artery. Severe cases may show flow reversal in the hepatic artery.

Conventional catheter angiography was the imaging modality of the choice for MALS in the past. This could show the focal narrowing of the celiac axis, its post-stenotic dilatation, and the collaterals arising from the SMA.^[10] This modality has largely been supplanted by non-invasive multidetector CT scanners. A multiplanar CT scan can detect focal narrowing of the celiac axis with a characteristic hooked appearance.^[11] Collateral vessels arising from SMA and supplying the celiac trunk territory may also be noted.

Surgical median arcuate ligament release has been the mainstay of treatment. The patient characteristics that predict relief of symptoms after surgery are postprandial pain, age 40–60 years, female gender, and weight loss >20 pounds.^[12] Almost 80% of the patients show symptomatic relief with modern laparoscopic techniques.

CONCLUSION

This case confirms that MALS is a challenging diagnosis. Patients usually undergo extensive workup for their abdominal pain before this diagnosis is reached. It is a good choice to perform abdominal ultrasound with Doppler imaging in a patient with symptoms such as postprandial angina. Confirmation of these findings can be done on CT angiography.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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