

Harjola Marable Syndrome: A Rare Case Report

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Abstract- Background: Compression of celiac artery due to unusual placement of median arcuate ligament which is ligamentous continuation of diaphragm over aorta results into symptom complex called as median arcuate ligament syndrome or Harjola Marable syndrome. All the patients who have unusually low lying median arcuate ligament may or may not be symptomatic.

Case report: We describe a case of 31 years old male patient who presented with complaints of pain in epigastric region and was diagnosed as celiac artery syndrome after extensive workup including OGD scopy, USG and CT angiography. He underwent laparoscopic release of median arcuate ligament which resulted in relief of symptoms.

Index Terms- Median arcuate ligament syndrome, celiac artery syndrome, Harjola-Marable syndrome, Celiac axis syndrome, Marable syndrome

I. INTRODUCTION

Celiac artery compression was first observed by Benjamin Lipshutz in 1917. It is also been called as Harjola-Marable syndrome, Marable syndrome, celiac artery compression syndrome, celiac axis syndrome, celiac trunk compression syndrome or Dunbar syndrome.

The median arcuate ligament is a ligament formed by joining of the left and right diaphragmatic crura join near the 12th thoracic vertebra. This fibrous arch forms the anterior aspect of the aortic hiatus, through which the aorta, thoracic duct, and azygos vein pass. The median arcuate ligament usually comes into contact with the aorta above the branch point of the celiac artery. Occasionally the median arcuate ligament passes in front of the celiac artery, below T12, compressing the celiac artery and nearby structures such as the celiac ganglia. In some of these individuals, this compression is pathologic and leads to the median arcuate ligament syndrome.

The syndrome most commonly affects individuals between 20 and 40 years old, and is more common in women.

Patients with MALS experience abdominal pain, particularly in the epigastrium, which may be associated with eating and which may result in anorexia and weight loss. Other signs are persistent nausea, and exercise intolerance. Diarrhoea is a common symptom.

Occasionally, abdominal bruit is heard in the mid-epigastrium.

Complications of MALS result from chronic compression of the celiac artery. They include gastroparesis and aneurysm of the superior and inferior pancreaticoduodenal arteries.

Median arcuate ligament syndrome is difficult to diagnose and is a diagnosis of exclusion. It is usually diagnosed after extensive workup including upper endoscopy, colonoscopy, and evaluation for gallbladder disease and gastroesophageal reflux disease (GERD).

II. CASE REPORT

31 years old male patient presented with complaints of pain in abdomen, mostly in epigastrium on and off since 1 year, aggravated since 6 months. Pain used to aggravate on food intake and after activity. Pain was associated with nausea and giddiness. Physical examination was essentially normal. Upper GI scopy was within normal limits. USG abdomen showed narrowing of celiac artery at the origin with increased in peak systolic Velocity. Diameter of celiac artery at origin 3.3 mm and distal celiac artery 6.6 mm suggestive of post stenotic dilatation.

PSV in erect position 134 cm/sec on inspiration and 204 cm/sec on expiration. On supine position, PSV increased to 304 cm/sec. Above findings were suggestive of ? celiac artery compression syndrome. Which was further confirmed with CT angiography. CT angiography showed severe narrowing of the origin of celiac artery due to thickened median arcuate ligament of diaphragm. There is post stenotic dilatation of celiac artery. Findings are suggestive of median arcuate ligament syndrome.

Patient underwent laparoscopic release of median arcuate ligament. Post op was uneventful.

5 months after surgery patient underwent repeat abdominal ultrasound which showed marked improvement in celiac artery PSV. USG with Doppler showed calibre of celiac artery at origin and in distal part to be 5-6 mm. PSV in erect position on inspiration and expiration 113 cm/sec and 124 cm/sec in supine position.

PSV and diastolic velocity of celiac artery were in expected range even in supine position and on expiration. No elevated velocity seen in supine position and on expiration which was suggestive of adequate release of median arcuate ligament.

III. DISCUSSION

Classically, MALS involves a triad of

1. Abdominal pain after eating,
2. Weight loss, and
3. An abdominal bruit.

Although the classic triad is found in only a minority of individuals. And as the symptoms are non specific it is difficult to diagnose.

There are various theories stating cause of pain in MALS.

- 1.Pain in MALS is assumed to be arising from inadequate blood flow through celiac artery due to Stenosis.
- 2.Compression of celiac ganglion.
- 3.Steal phenomenon which suggests diversion of blood to celiac artery from superior mesenteric artery through collaterals resulting in inadequate flow to intestine.

Although compression of celiac artery is widely accepted.

MALS is diagnosed using [duplex ultrasonography](#). It measure blood flow through the [celiac artery](#). Peak systolic velocities greater than 200 cm/s are suggestive of celiac artery stenosis . Further evaluation and confirmation can be obtained via [angiography](#) to investigate the anatomy of the celiac artery.

CT angiographic findings in MALS s/o

- 1.Proximal stenosis of celiac artery with post stenotic dilatation.
- 2.Indentation on superior aspect of celiac artery
- 3.Hook shaped contour of celiac artery.

These imaging features are exaggerated on expiration, even in normal asymptomatic individuals without the syndrome.

The hook-shaped contour of the celiac artery is characteristic of the anatomy in MALS and helps distinguish it from other causes of celiac artery stenosis such as [atherosclerosis](#).

Decompression of the celiac artery is the general approach to treatment of MALS. The mainstay of treatment involves an [open or laparoscopic surgical approach](#) to divide, or separate, the median arcuate ligament to relieve the compression of the celiac artery. Other modalities include stenting with dilatation although chances of recurrence are high, percutaneous angioplasty and aortoceliac bypass graft.

IV. CONCLUSION

Median arcuate ligament syndrome is a diagnosis of exclusion in a case of pain in abdomen, especially in case of post prandial fullness. It should be considered in patients complaining of post prandial pain in abdomen in whom other like GB disease, pancreatitis, acid peptic disease have been ruled out.

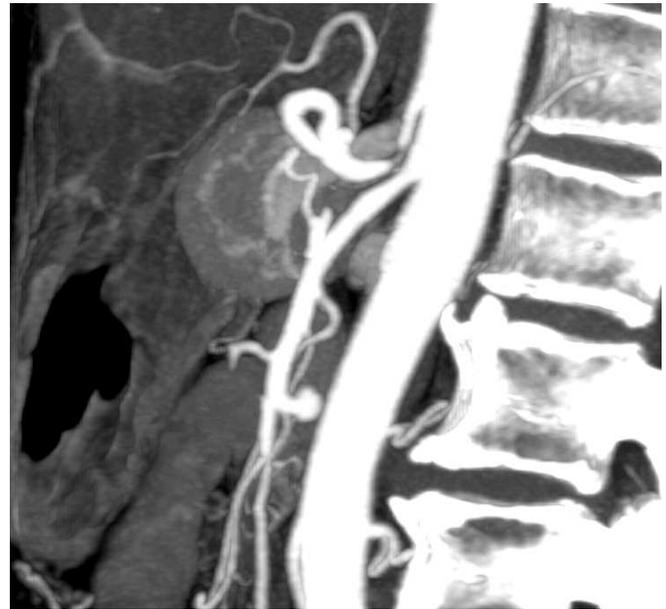


Figure ICT image on sagittal section showing narrowing at celiac artery origin and its hook shape contour.

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