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Median Arcuate Ligament Syndrome (MALS)

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Abstract: *Median Arcuate Ligament Syndrome(MALS) is a rare disorder characterised by postprandial intestinal angina due to coeliac artery compression. The median arcuate ligament is fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus, passes superior to the origin of the celiac artery and is a continuation of the posterior diaphragm that wraps over the aorta. If it lies too low on the aorta, the ligament may cause symptoms of abdominal pain related to compression of the celiac artery.*

Keywords: *celiac artery, celiac artery compression, median arcuate ligament*

I. Introduction

Median Arcuate Ligament Syndrome(MALS) was first observed by Benjamin Lipshutz in 1917[(1)] and first described by Pekka- Tapani Harjola in 1963[(2,3)] and subsequently by J.David Dunbar and Samuel Marable in 1965 (Harjola-Marable Syndrome or Marable Syndrome[(1)]or Dunbar Syndrome or Coeliac Artery Compression Syndrome). It typically occurs in young (20-40 years), mostly lean women [(4,5,6)], presenting with epigastric pain, sitophobia (food fear) and weight loss[(7)] and have had extensive work-ups for other sources of abdominal pain. Pain is associated with eating but not always [(8)]. At physical examination, an abdominal bruit that varies with respiration may be audible in mid- epigastric region in approximately 83% cases [(5,6)].

Median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of aortic hiatus, arising from the anterior surface of L_1-L_4 vertebral bodies on the right and L_1-L_3 vertebral bodies on the left and from the intervertebral discs and anterior longitudinal ligament [(9,10)]. This ligament usually passes over the aorta superior to the origin of celiac axis, however, in 10-24% of people it may be low and cross over the proximal portion of celiac axis causing characteristic indentation [(4,9,11,12)]. A small subset of these individuals exhibit severe compression associated with symptoms of MALS [(4)]. The incidence of this syndrome is reported to be between 1.76-4% [(13)]. Complications of MALS include gastroparesis [(14)] and aneurysm of pancreatico-duodenal artery [(15)]. Pathophysiology includes increased compression during expiration as diaphragm moves caudally resulting in visceral ischemia and pain, or steal phenomenon from blood flow being diverted away from superior mesentric artery via collaterals to celiac axis causing midgut ischemia [(5,6)]or overstimulation of celiac ganglion causing chronic pain.

MALS is a diagnosis of exclusion [(4,16)]. Other causes of abdominal pain like biliary sources, ulcer disease, should be excluded.

Case History: A 35-year-old woman with no significant past medical history presented to the hospital with a 2 years history of intermittent epigastric abdominal pain. The pain was associated with nausea and bloating. The nausea worsened with any oral intake and relieved with bowel rest. She had rare diarrhoea and denied any radiation of the pain to other locations in her body. There were no similar complaints in other family members. She denied any drug or tobacco product use. She had lost 9kgs weight. Her physical examination revealed mild epigastric tenderness to palpation but no other abnormalities.

Electrolyte, *H. pylori* titres, liver function tests, amylase, lipase, and complete blood count all were within normal limits. Right upper quadrant ultrasound showed an insignificant liver cyst and no evidence of cholelithiasis. Gastric emptying study showed minimal delay. A hepato-biliary scan was normal. Colonoscopy showed granularity of terminal ileum. A mesentric ultrasound showed elevated celiac artery velocity that augmented with inspiration. Superior mesentric artery velocities matched those of aorta and indicated no abnormalities. CT scan of abdomen showed a small subcentimetric hypodense lesion in segment8 of liver (likely suggestive of simple cyst) and significant narrowing at the origin of celiac axis (by a shelf-like projection superiorly, likely hypertrophied Median Arcuate Ligament) was also noted. Post-stenotic dilatation of celiac axis was seen and there was no evidence of any sclerotic plaque Fig. [1,2,3]. The patient underwent laparotomy which revealed MAL impingement which was released. At follow-up patient's postprandial pain had significantly improved and tolerated a diet without difficulty.

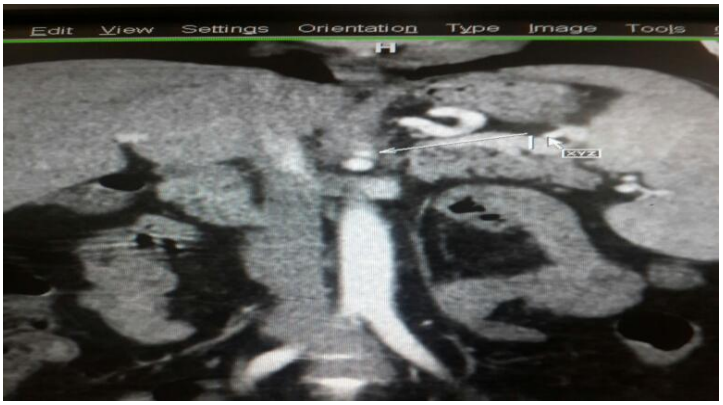

Figure 1: Coronal section showing narrowing at the origin of celiac trunk.

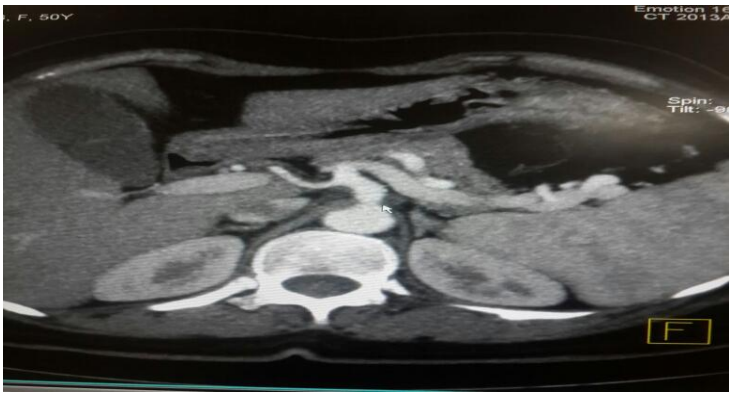

Figure 2: Transverse section showing significant narrowing.

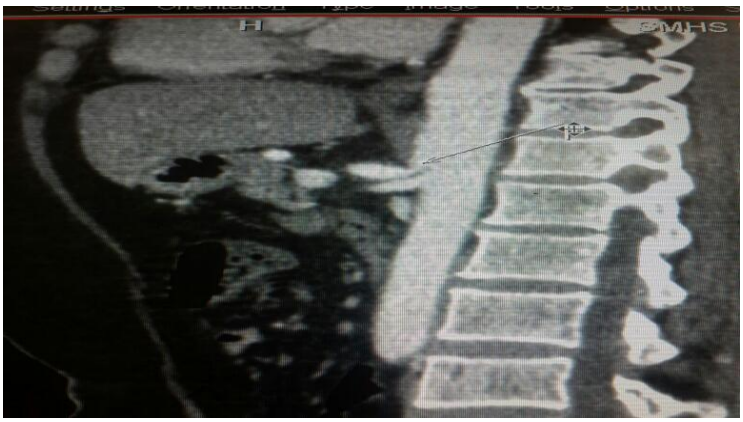

Figure 3: Narrowing with post-stenotic dilatation

II. Discussion

The definition of MALS relies on both clinical and radiographic features. CT angiography [7,9,17] with 3D imaging demonstrates a characteristic focal narrowing in the proximal celiac axis (hooked appearance) during inspiration, with associated post-stenotic dilatation or collateral vessels. Doppler ultrasound or Mesentric Ultrasound and MRI Angiography can also be used. It reveals variation of peak systolic velocity (PSV) during respiration with a marked increase during expiration to more than 200cm/s. A greater than 3:1 ratio of PSV in celiac axis in expiration compared with PSV in abdominal aorta immediately below diaphragm can be used as other criterion [18,19,20]. Reversal of flow in hepatic artery may also be seen [6].

Decompression of celiac artery is the general approach to treatment of MALS [11]. Either open surgical [11] or laparoscopic [3,21,22] approach is used. The other interventions include celiac gangliectomy, celiac artery revascularization (aorto-coeliac bypass, patch angioplasty) [11], percutaneous transluminal angioplasty (PTA) [5,11,23], robotic ligament release, angioplasty with stenting in recalcitrant cases[24,25].

There are few studies of long-term outcomes of patients treated for MALS. According to Duncan [(17)], the predictors for favourable outcome are;
-Age 40-60 years
-Lack of psychiatric illness
-No alcohol use
-Abdominal pain that was worse after meals
-Weight loss more than 20lb (9.1kg)
-Post-stenotic dilatation and collateral vessels [(26)].

III. Conclusion

Median arcuate ligament syndrome is a difficult diagnosis to obtain in a majority of patients. Most patients have had extensive workups or various surgical procedures for postprandial abdominal pain. A patient with suspected compression of the celiac artery should undergo a mesenteric ultrasound with evaluation of artery velocities. Confirmation of this diagnosis can be performed with conventional angiography or CT angiography. Patients who have evidence of median arcuate ligament syndrome should undergo surgical decompression, which can be accomplished laparoscopically.

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