Dunbar Syndrome: A Rare Case Report of Celiac Artery Compression Syndrome

Kripa Shankar Tiwari¹, Vartika Jain¹, A. Ghanghoria¹
¹(Department of General Surgery, M.G.M.M.C. & M.Y.H. Hospital, India)

ABSTRACT:
AIM: To present a case of Dunbar syndrome and the associated surgical experience.
BACKGROUND: Dunbar Syndrome or otherwise also known as Celiac artery compression syndrome or Median Arcuate Ligament Syndrome is a rare cause of abdominal pain and weight loss. It is caused by compression of the celiac artery and possibly the celiac ganglion by median arcuate ligament (MAL). The incidence is 10-24% but approximately 1% of these individuals exhibit symptoms of severe compression by median arcuate ligament. The diagnosis is difficult as it presents with non-specific symptoms which overlap with other more common causes of chronic intermittent abdominal pain. The diagnosis is often made by exclusion. Laparoscopic approach is considered to be an optimal therapeutic approach.
METHOD: We describe a case of a 42 year old female who presented with a 4 year long history of postprandial abdominal pain, occasional vomiting and severe weight loss in the last one year. An abdominal CT angiography demonstrated an external compression of the celiac trunk and the patient underwent laparoscopic decompression by division of the MAL.
RESULT: Postoperative course was uneventful and the patient was discharged on the 5th postoperative day.
CONCLUSION: Laparoscopic approach gave a magnified operative view assisting the dissection of MAL fibers along with the advantage of early recovery, less postoperative pain and excellent cosmetic outcome. This procedure is safe and feasible approach and restores the quality of life of the patient.
KEY WORD: Median Arcuate Ligament Syndrome, Dunbar Syndrome, Celiac Artery Compression Syndrome, Median Arcuate Ligament.

I. Introduction
Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome, consists of chronic and recurrent abdominal pain due to the compression of the celiac trunk by the median arcuate ligament (MAL). The syndrome was first described by Harjola in 1963 and Dunbar in 1965.[1,2] Dunbar syndrome is more commonly seen in women (4:1) of 40-60 years age group, mostly with a thin body built. Some patients remain asymptomatic due to development of collateral blood supply from the superior mesenteric artery, however symptoms may develop due to celiac artery or celiac ganglion compression and consist of the classical triad seen in mesenteric ischemia characterized by postprandial abdominal pain, nausea and vomiting and subsequent weight loss.[3,4]

The diagnosis is a difficult one and often made by excluding differential diagnoses of intermittent abdominal pain. Selective angiography in expiration or magnetic resonance (MR) angiography identifies the stenosis of the proximal segment of the celiac artery and helps to confirm the diagnosis. The treatment includes the release of the median arcuate ligament along with the fibres of the celiac plexus which can be done by percutaneous transluminal angioplasty or surgery.[1,4] In surgery, the laparoscopic approach is preferred over the open approach.[5,6]

II. Case Report
A 40-year-old female, presented with a history of moderate postprandial pain in epigastrium, occasionally associated with nausea, vomiting and dyspepsia since 4 years. The patient also had a history of unintentional weight loss of approximately 15 kg in the past one year. She had no history of any chronic disease or drug use and was not a smoker. She had no family history for neoplasm and her only past surgical history was of a laparoscopic appendectomy. Her physical examination was mostly normal, except for a mild epigastric tenderness on palpation. Laboratory tests, abdominal ultrasound and gastroscopy were performed which were also within normal limits. The symptoms persisted despite therapy with prokinetic drugs. Hence, an abdominal CT-angiogram was performed that showed a proximal stenosis of the celiac trunk, with post stenotic dilatation...
and characteristic “hooked appearance” due to indentation of celiac trunk on its superior surface. [Figure 1,2] The finding was suggestive of compression by median arcuate ligament.

The patient was advised a laparoscopic exploration and release of the MAL. After obtaining an informed consent, surgery was commenced. Under general anaesthesia with the patient in supine position with open legs and the first surgeon between the patient’s legs, pneumoperitoneum was created. A camera port was inserted periumbilical and 4 more trocars were placed subsequently: two in the left subcostal area, 1 in the right subcostal area and 1 in the subxiphoid location (for the elevation of left lobe of the liver). Following abdominal exploration, the Left Gastric Artery and the Common Hepatic Artery were isolated. The fibrotic structures impinging on the origin of the celiac trunk were visualized and divided. A drain was placed near the aorta which was removed on the 2nd postoperative day. The postoperative course was uneventful. The patient tolerated soft diet well on postoperative day 1 and was discharged on day 5. On regular follow up at POD 14 and then after two months and after three months, the patient had no complaints of pain and tolerating feeds well with insignificant discomfort.

### III. Discussion

Dunbar syndrome or median arcuate ligament syndrome (MALS) is a rare vascular compression syndrome with its etiology and pathophysiology, still not fully understood. The MAL appears as a fibrous arch passing over the aorta at the level of L1 just above the origin of the celiac trunk. The celiac ganglia lie closely, just below and lateral to the celiac trunk. In some patients a low insertion of MAL can compress the celiac trunk.

Asymptomatic compression or stenosis of the celiac trunk is common, probably due to hemodynamic compensation mechanisms. While for the symptomatic cases, vascular and neurological theories have been suggested.

Compression of the celiac trunk by MAL causes symptoms of mesenteric ischemia when the arterial blood flow is reduced below 60-75%, since there is an extensive collateral blood supply to the mesentery from other blood vessels (superior and inferior mesenteric artery). The effective stenosis of the celiac trunk can vary during the respiration due to change in the position of celiac trunk and MAL during respiration. The aorta with its branches move cranially during expiration, consequently MAL moves over the celiac trunk to cause its maximal compression during expiration, as was proved in the study by Lee et al.

MALS may be suspected in middle-aged (40 to 60 years), thin built female patients presenting with a clinical triad of postprandial epigastric pain, nausea or vomiting and weight loss. Physical examination is usually normal and epigastric tenderness may be present, but these clinical features are not specific of the syndrome. Angiography confirms the diagnosis by revealing vascular compression. The gold standard in the diagnosis is the selective arteriography which should be performed during both inspiration and expiration, although the first approach is often with an abdominal CT-scan or MR.

In our case, we made the diagnosis with an abdominal CT-angiogram, showing the proximal stenosis of the celiac trunk at the origin and post stenotic dilatation. The diagnosis is very difficult in most cases, with an average duration of symptoms of 34 months before a diagnosis is obtained.
Doppler Ultrasound has been reported to have a high sensitivity for the diagnosis of MALS: ultrasound may show an elevated systolic velocity at the origin of the celiac trunk during expiration and normal velocities in inspiration.

The mainstay of treatment of MALS is decompression of celiac trunk to restore blood flow by releasing the extrinsic compression on the vessel. In 2000, the first laparoscopic decompression was published by Roayaie et al. [11]

Both open and minimally invasive approaches can be used, but we suggest laparoscopy because of its superadded advantages of smaller incisions, decreased pain and postoperative morbidity, quicker recovery and a magnified view of the operative field which is critical in this case considering the major vessels involved.[12]

Exposure of the aorto-celiac axis is considered the most challenging part of the surgical procedure. Confirmation of the total release of celiac trunk can be obtained by arteriography or intraoperative doppler ultrasound. In our case, we considered the visual inspection of an increased celiac flow to be sufficient.

Angioplasty and stenting of celiac trunk have been described as reasonable tool in the treatment of MALS, but we still believe that the treatment is primarily surgical.

IV. Conclusion

MALS is a rare condition that should be considered as a differential diagnosis in patients with nonspecific postprandial abdominal pain, particularly in middle-aged females. The diagnosis is challenging and often doubtful. Laparoscopic surgical approach is preferred as it is feasible and safe with satisfying results, leading to significant resolution of symptoms and an improved quality of life.

References