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Case Report

Internal Medicine Section

Dunbar Syndrome-A Rare Cause of Foregut Ischemia

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ABSTRACT

Median Arcuate Ligament Syndrome (MALS) is a condition that can result from an anatomical aberration. If the median arcuate ligament is located too inferiorly in relation to the celiac axis, it can impede circulation and lead to vascular compromise. Here, we present the case of a 63-year-old woman, who came to the hospital complaining of continuous epigastric pain and who was ultimately found to have MALS. Her epigastric pain could be the result of the ischemia caused by MALS that made it difficult for the duodenal ulcer to heal properly. This case report documents an unusual presentation of an already rare condition.

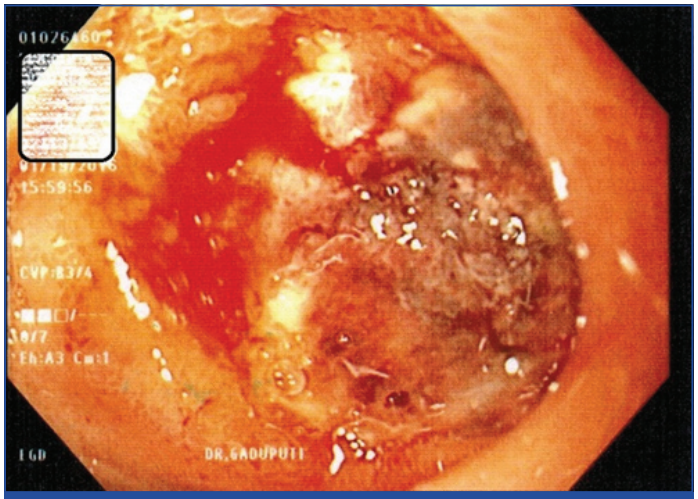
Keywords: Celiac axis compression syndrome, Celiac territory ischemia, Median arcuate ligament syndrome, Peptic ulcer disease

CASE REPORT

A 63-year-old woman with multiple medical conditions including type-2 diabetes mellitus, schizophrenia and hypertension, presented to the emergency department with abdominal pain for two months. Pain was located in the epigastrium, continuous in nature and with severity of 7/10 on a pain scale of 0-10. Patient also reported exacerbation of pain with eating but with no relieving factors. Patient also reported anorexia and 15 lb weight loss over two months period. She reported poor response of her symptoms to high dose Proton-Pump Inhibitors (PPIs) prescribed by her primary care physician. Patient denied using non-steroidal anti-inflammatory drugs. Physical examination was significant for epigastric abdominal tenderness and abdominal bruit. Laboratory values were significant for hemoglobin of 9 gm/dl, mean corpuscular volume of 76. Computed Tomography Angiography (CTA) abdomen was performed which revealed antral and duodenal bulb thickening and focal stenosis of the origin of the celiac axis with the vessel demonstrating a hook like appearance on the sagittal view, as typically seen in cases of Median Arcuate Ligament Syndrome (MALS) [Table/Fig-1].

A diagnostic Esophagogastroduodenoscopy (EGD) was performed and revealed an oozing, crated duodenal bulb ulcer [Table/Fig-2], which was treated with epinephrine injection and cautery. Biopsies were not found to have *Helicobacter pylori* infection. Patient was discharged on high dose of PPIs and was eventually referred for surgical correction of MALS in view of refractory symptoms. This is a very rare case where MALS resulted in celiac territory ischemia with a complicated and poorly healing duodenal ulcer.

[Table/Fig-1]: Computed Tomography Angiography (CTA) showing hook like morphology of celiac trunk consistent with MALS.



[Table/Fig-2]: Endoscopy showing oozing cratered duodenal bulb ulcer.

DISCUSSION

The median arcuate ligament is a fibrous structure that connects the two crura of the diaphragm at L1, which are on either side of the aorta. This ligament typically passes superior to the origin of the celiac axis as the celiac trunk migrates caudally during embryogenesis. However, in some instance, the ligament is positioned low, crossing over the proximal portion of the celiac axis and compressing it. In MALS, if the celiac axis is compressed enough, vascular flow can be disrupted, producing ischemic symptoms [1]. This phenomenon, also known as CACS or Dunbar syndrome, is a rare condition that tends to affect females in their fourth and fifth decades of life [2].

The characteristic triad of postprandial abdominal pain, epigastric bruit and weight loss is thought to be indicative of MALS. Other symptoms include: nausea, vomiting, bloating, diarrheoa and a

reduced appetite, all which result from blood being shunted away from the gastrointestinal tract. An estimated 10%-24% of patients with CACS experience a significant enough disruption of celiac blood flow that they exhibit these symptoms. Patients with CACS often do not display this triad though because of collateral supply from the superior mesenteric artery circulation [3].

The CACS diagnosis can be made through Doppler ultrasound, spiral CTA, selective catheter angiography and magnetic resonance angiography. Doppler US has been cited as the best initial test for CACS, whereas selective angiography and CTA are the gold standard diagnostic methods and can be used to create three-dimensional reconstructions [4]. When trying to differentiate between MALS and atherosclerosis, it is important to look for arterial calcifications and a characteristic hook like appearance. Presence of hook like appearance with absence of arterial calcifications is characteristic of MALS [1].

Given the overall rarity of MALS or CACS, some of the most helpful information comes from patient case studies in different settings. In one case study, a 46-year-old man was admitted complaining of exercise-induced lower retrosternal pain that traveled to the epigastric region for the past two years. His symptoms were likely exacerbated in states of increasing oxygen demand due to the arterial compression and he cited increasing pain following consumption of food. The CTA revealed a localized stricture or narrowing in the proximal celiac artery with excessive fibers originating from the diaphragm. In addition, three-dimensional CTA reconstructions confirmed the diagnosis of CACS. A laparoscopic release of the median arcuate ligament was performed and the patient was no longer symptomatic only one month after surgery [4].

Another study used MDCT and found that in those with MALS, the characteristic 'hooked appearance' was identified in 44% of patients with MAL compression. This refers to the focal narrowing of the proximal celiac artery. Stenosis of the celiac artery was between 35%-50% and 51%-80% stenosis in 15 and 35 total patients, respectively. Although rare, MALS must be considered in any patient with unexplained gastrointestinal symptoms. [5].

There is currently no medical treatment for MALS. Surgery is the only effective approach to release the compression by the median arcuate ligament, although it is not without controversy. In more extreme cases, those with post-stenotic dilatation or collateral vessels, surgery is recommended. Surgeons perform both laparoscopic and open surgical techniques to relieve the compression. More recently, the laproscopic approach has gained greater traction. Future directions for treatment of MALS include the possibility of performing celiac angioplasty or endovascular stenting [6].

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