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MEDIAN ARCUATE LIGAMENT SYNDROME WITH SUPERIOR MESENTERIC ARTERY THROMBOSIS

Dr. Mandapati Mallikarjuna Reddy Associate Professor, Department of General Surgery, NRI General Hospital, Chinakakani, Guntur, Andhra Pradesh, India

Dr. Jukuri Sanjeev Kumar Postgraduate, Department of General Surgery, NRI General Hospital, Chinakakani, Guntur, Andhra Pradesh, India

Dr. Nasika Nageswara Rao Professor, Department of General Surgery, NRI General Hospital, Chinakakani, Guntur, Andhra Pradesh, India

ABSTRACT

Median arcuate ligament syndrome (MALS) is a rare etiology of chronic abdominal pain, clinically characterized by the triad of postprandial abdominal pain, weight loss, and often an abdominal bruit due to compression of the celiac artery by the median arcuate ligament. Given the nonspecific symptoms, this is a rare and difficult diagnosis to obtain. Symptomatic Superior Mesenteric Artery(SMA) thrombosis most often accompanies celiac occlusion. Thrombosis of the SMA (approximately 25% of cases) is usually associated with pre-existing chronic atherosclerotic disease leading to stenosis. History consistent with chronic mesenteric ischemia (CMI), including postprandial pain, weight loss, or "food fear", and thus a systematic history is important when evaluating a patient suspected to have AMI. We present a patient with postprandial abdominal pain with fear of food intake, vomitings and weight loss, in whom etiology was ultimately determined to be median arcuate ligament syndrome with SMA thrombosis. Decompression of median arcuate ligament and revascularization is done.

KEYWORD

Median arcuate ligament syndrome, chronic mesenteric ischemia, SMA thrombosis

*Corresponding Author Dr. Jukuri Sanjeev Kumar

Postgraduate, Department of General Surgery, NRI General Hospital, Chinakakani, Guntur, Andhra Pradesh, India sanjeevjukuri@gmail.com

Introduction:

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome, celiac axis syndrome or Dunbar syndrome, is often misdiagnosed due to its relative scarcity.1 Median arcuate ligament is a fibrous arch that usually passes just superior to the celiac artery near the first lumbar vertebra. In 10% to 24% of the general population, the ligament crosses anterior to the celiac artery, and a few of these patients may have hemodynamically significant stenosis causing abdominal pain.2 The classic triad of post-prandial abdominal pain, weight loss and epigastric bruit is likely to be incomplete. Due to a wide differential diagnosis, including peptic ulcer, gallbladder disease, appendicitis, IBD etc., most patients will have undergone multiple radiologic investigations or procedures including oesophagoduodenoscopy or even diagnostic laparoscopy.1 Typically, the compression of the celiac trunk is usually seen on the sagittal view of a computerized tomography scan where it appears prominent on expiration; however, in severe cases, it may be shown as a persistent compression on inspiration.3 Traditionally, the treatment of this syndrome is surgery such as a classic opening or a novel laparoscopic division of the median arcuate ligament to relieve the extrinsic compression.1 Percutaneous endovascular treatment is an alternative technique and may be considered in selected cases, for which the traditional surgery failed or was a treatment option. We report a median arcuate ligament syndrome case with a severe compression of the celiac trunk, and with SMA thrombosis presenting with postprandial abdominal pain, vomitings and weight loss.

Case Report:

A 30-year-old male, who had not any past medical history, presented with severe upper abdominal pain since 3 days, associated with vomitings. Patient has chronic intermittent abdominal pain for the last 5 months, during which he had experienced a weight loss of 8 kilograms. On clinical examination, Mild tenderness in the epigastric and umbilical region, no palpable masses, no guarding and rigidity, bowel sounds were present. His whole abdomen sonogram and oesophagogastroscopy were within normal limits. However, the contrast enhanced computer tomography (CECT) of the abdomen revealed, compression of celiac axis by the median arcuate ligament and short segment complete SMA thrombosis with segmental bowel ischemia of jejunal loop and prominent collaterals between SMA and IMA. According to his clinical symptoms and imaging findings, the patient was diagnosed with MALS with SMA thrombosis. The patient was treated by exploratory laparotomy, Intraoperatively there was Jejunal stricture with ischemic changes and Median arcuate ligament is constricting the celiac plexus. Releasing i.e., narrowing of the medial arcuate ligament, resection of a segment of jejunum including stricture with end to end anastomosis and revascularization for the SMA thrombosis was done. Postoperative period is uneventful.

Discussion:

This syndrome was first described by Harjola4 in 1963. The incidence of MALS has been found in 10–24%.3 Even though the etiology of MALS is not well understood, there are 2 main theories, mainly mesenteric ischemia and nerve dysfunction.1,5 One of the proposed theory is compression of the celiac artery by a median arcuate ligament that causes mesenteric ischemia, which leads to classical symptoms of MALS.1 However, It is not well known how celiac artery

compression alone can cause mesenteric ischemia since generally there is extensive collateral blood supply to the mesentery from other blood vessels.6 Another proposed mechanism for MALS is due to celiac plexus nerve dysfunction.5 Isolated vascular compression of the celiac artery as the sole etiologic factor seems unlikely. Symptomatic SMA thrombosis most often accompanies celiac occlusion.7 First, in 10%–24% of the population the MAL can cause asymptomatic compression.8 Second, collateral circulation by the superior mesenteric artery provides adequate blood supply; therefore, postprandial abdominal pain should not be expected with celiac artery compression alone. The cause of MALS is likely multifactorial, including compressive effects on the celiac artery and surrounding neurogenic structures. In our patient, CECT Abdomen demonstrated focal narrowing of the celiac axis at the origin with a post-stenotic dilation along with some degree of superior compression of the celiac axis suggestive of MALS and short segment complete SMA thrombosis with segmental bowel ischemia of jejunal loop and prominent collaterals between SMA and IMA. Although conventional angiography is considered that gold standard for the diagnosis of MALS, angiography has largely been replaced by CTA and MRA.9 Sagittal view CTA or MRA will be able to detect focal narrowing of the proximal celiac axis.9 In celiac artery compression, it has been noted that either the celiac artery is located slightly higher or the MAL is located lower than expected.10 In a large series, significantly higher symptomatic relief was achieved through combined release of the MAL and revascularization.11 If celiac artery compression alone is corrected, evidence suggests up to 53% will be asymptomatic on long-term follow up.11 Combined release and revascularization however increases the long-term success to 79%.11 Therefore some patients may require additional revascularization procedures such as mesenteric artery stenting or bypass to provide long-term symptomatic relief. Since delay of revascularization is unlikely to adversely affect outcome, most choose ligament release first, followed by revascularization via stenting or bypass if symptoms persist. Endovascular celiac artery stenting alone without release of the ligament is discouraged because of clinical failure and recurrent stenosis.12 Surgical decompression of the median arcuate ligament is the treatment of choice for symptomatic MALS patients. There are different modalities of surgery. Open surgery with decompression of the celiac artery and celiac plexus by division of the median arcuate ligament fibres is the most common treatment. Endovascular treatment is beneficial in some selected cases such as those with a failure of traditional surgery, or contraindication to surgery. It is also demonstrated, resolution of postprandial pain in 81% of patients. If symptoms persist, revascularization of the celiac artery by either endovascular stenting or bypass can be considered as secondary procedures.

Conclusion:

This case illustrates that MALS is an often missed diagnosis due to its nonspecific symptoms. Postprandial abdominal pain should not be expected with celiac artery compression alone, as collateral circulation by the superior mesenteric artery provides adequate blood supply. So, it should be remembered that Median arcuate ligament syndrome along with superior mesenteric artery thrombosis in a case of a severe mesenteric ischemia. Patients who are diagnosed should be referred to surgery for decompression of the median arcuate ligament and revascularization for long term success.

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