

CHRONIC ABDOMINAL PAIN, AN OVERLOOKED DIAGNOSIS OF MEDIAN ARCUATE LIGAMENT SYNDROME (MALS)

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ABSTRACT

Median arcuate ligament syndrome (MALS) is a rare condition that is often overlooked as a result of its nonspecific symptoms. It is usually presented with nausea, bloating, abdominal postprandial pain, and weight loss. The diagnosis of MALS is usually delayed and made by excluding other causes for the symptoms. The diagnosis of this syndrome is based on clinical presentation and radiological findings on computer tomography angiography (CTA) or magnetic resonance angiography (MRA). Surgery is treatment of choice, with promising results from laparoscopic surgery.

Keywords: median arcuate ligament syndrome, MALS, abdominal pain

INTRODUCTION

Median arcuate ligament syndrome (MALS) also known as celiac artery compression syndrome (CACS) or Dunbar syndrome is a rare syndrome with incidence of 10-24% [1]. It is a condition in which the median arcuate ligament compresses the celiac artery, possibly leading to ischemia and disease symptoms. MALS is more common in females, with a female to male ratio of 4:1 [2, 3]. When symptomatic, it is usually presented with nausea, bloating, abdominal postprandial pain and weight loss. As a result of its nonspecific symptoms numerous investigations are performed before the diagnosis is made. The diagnosis of this syndrome is usually made via computer tomography angiography (CTA) or magnetic resonance angiography (MRA) [4]. We present the case of a

patient with MALS, manifested with chronic abdominal pain that was resolved with open surgery.

CASE REPORT

A 45 year old woman presented with abdominal bloating and long-lasting abdominal pain, predominant in the upper part of the abdomen. The symptoms intensified in the last two months and were more pronounced postprandial. This patient had no previous medical conditions and did not report any loss of appetite, loss of weight or change in bowel habit.

On physical examination the patient had epigastric pain to deep palpation. All laboratory findings, including amylase, lipase, and an *Helicobacter pylori* test, were within normal ranges. The abdominal ultrasound did not reveal any abnormalities. The patient underwent upper and lower endoscopy. Esophagogastroduodenoscopy (EGD) showed small hiatal hernia. Several biopsies from the esophagus, stomach and duodenum were taken and the pathohistological report was normal. Colonoscopy revealed normal ileal mucosa and grade II internal hemorrhoids.

Abdominal computer tomography angiography (CTA) was performed, which revealed stenosis of the celiac trunk with compression of 60% of the artery because of low lying of the arcuate ligament (figure 1). There were no signs of atherosclerosis of the blood vessels. With this the diagnosis of median arcuate ligament syndrome (MALS) was made.



Figure 1. Sagittal CTA image of compression of celiac trunk by the median arcuate ligament (white arrow)

The patient underwent an open surgery. The origin of the celiac artery was identified, the median arcuate ligament was circumcised and resection of the celiac ganglion was done (figure 2 and 3).

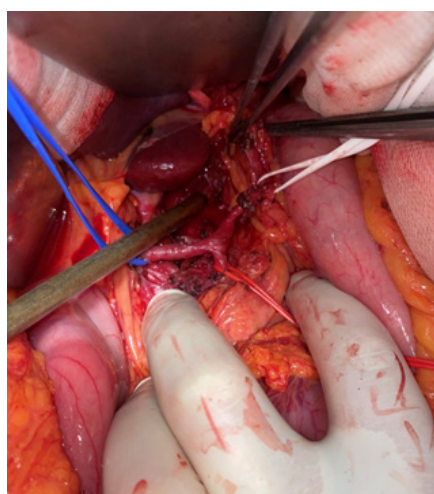
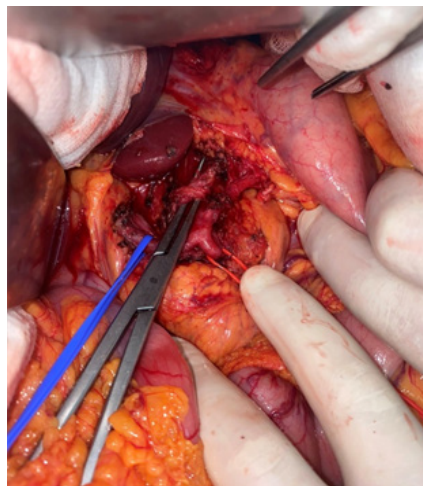


Figure 2 and 3. Intraoperative image of the median arcuate ligament compressing the celiac trunk and its circumcission

The postoperative period was without any complications and after 5 days the patient was discharged. At the two weeks and one month follow-up, the patient was feeling well and had complete resolution of the symptoms. One month later, control CT was performed. This showed a normal celiac trunk without stenosis (figure 4 and 5). The patient was still symptom free and had gained 3kg in the last month.



Figure 4 and 5. Postoperative CT of abdomen with no sign of stenosis of the celiac trunk

DISCUSSION

Harjola first described median arcuate ligament syndrome (MALS) in 1963 and later, Dunbar in 1965 [1]. This syndrome is also known as celiac artery compression syndrome (CACS) and Dunbar syndrome. Although the etiology of this condition is uncertain, there are two theses: the vascular and neurological [5]. The pathophysiology of the vascular theses is due to the decreased blood supply and intestinal ischemia as a result of the compression of the celiac artery [6, 7]. The neurological theses suggest involvement of the surrounding nerve plexus in the etiology of the abdominal pain and discomfort [6, 8]. It is taught that the neurogenic pain is either the result of stimulation of the celiac plexus or as a result of direct sympathetic irritation [9].

Most common symptoms of MALS are postprandial pain, weight loss, nausea, vomiting, and diarrhea. Due to its non-specific symptoms, prior to diagnosis, patients undergo a series of examinations such as abdominal ultrasound, upper and lower endoscopy to exclude other causes of abdominal pain. The diagnosis is usually made with the help of different imaging methods including Doppler ultrasound, CT angiography

or MR angiography. These are all non-invasive and quick methods that can easily demonstrate the place of compression of the celiac plexus in this condition. Literature shows that in 15% to 50% of asymptomatic patients, some degree of stenosis of the celiac trunk is present on angiographic imaging reports [10, 11]. On account of such findings, the diagnosis of MALS should be made when the imaging reports correlate with the patients' clinical presentation. Serious complications of MALS, such as gastroparesis and pancreatic duodenal aneurysm, require prompt surgical treatment [5].

By using Doppler ultrasound, one can detect the discontinuation of blood flow and increased blood flow in the proximal part of the celiac trunk. H. Gruber et al. showed that an expiratory peak velocity (PV) higher than 350 cm/sec and the bending of the celiac trunk greater than 50° had a sensitivity of 83% and a specificity of 100%. Thus, it is a useful tool for diagnosing of MALS [5, 12].

In MALS, CT and MR angiography demonstrate a hook-like narrowing of the celiac artery which is a typical sign for this syndrome [5, 13]. This hooked stenosis can help distinguish MALS from other causes of stenosis in the celiac artery such as atherosclerosis [10, 14]. For a better and more precise diagnosis, sagittal CTA image combined with 3D imaging of the celiac trunk can be used [14]. With the use of CTA and MRA, the remaining vascular structures and the low lining of the ligament can be visualized as well. This is important for preoperative planning.

The treatment of choice for this syndrome is surgery. The purpose of the procedure is decompression of the artery and establishing normal blood flow, as well as reducing or eliminating the neural irritations. Although open surgery has been mostly used in the past, nowadays, laparoscopic surgery has shown equally good results [15]. Patients that are surgically treated with the resection of the celiac ganglion and have vessel reconstruction have better outcomes and better symptom relief compared to patients treated only with celiac artery decompression [9, 16].

CONCLUSION

As a result of its nonspecific symptoms, the diagnosis of MALS is hard to obtain and it

is usually made by excluding other causes. This is a rare condition and doctors should be aware of it, especially when dealing with patients with abdominal bloating and postprandial pain. Surgery is the treatment of choice for this condition, which has shown good long-term results.

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Резиме**ХРОНИЧНА АБДОМИНАЛНА БОЛКА,
ПРЕВИДЕНА ДИЈАГНОЗА НА СИНДРОМ НА СРЕДЕН ЛАЧЕН ЛИГАМЕНТ (MALS)**

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Синдромот на среден лачен лигамент (median arcuate ligament syndrome – MALS) е ретка состојба, која често се занемарува како резултат на нејзините неспецифични симптоми. Обично се манифестира со гадење, надуеност, абдоминална постпрандијална болка и губење на телесната тежина. Дијагнозата на MALS вообичаено е одложена и се поставува со исклучување на другите причини за појава на овие симптоми. Дијагнозата на овој синдром се заснова на клиничката презентација и на радиолошките наоди на компјутерска томографија ангиографија (КТА) и магнетна резонанца ангиографија (МРА). Хирургијата е третман на избор, со ветувачки резултати од лапароскопската хирургија.

Клучни зборови: синдром на среден лачен лигамент, MALS, абдоминална болка