Case Study

Median Arcuate Ligament Syndrome: An Unusual Cause of Chronic Abdominal Pain

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Received 1 May 2015; Revised 20 December 2015; Accepted 12 January 2016

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Abstract Chronic abdominal pain is one of the most frequent complaints of patients visiting the gastroenterologist office and a large majority of these subjects have irritable bowel syndrome (IBS). Median arcuate ligament syndrome (MALS) is a rare cause of chronic abdominal pain that results from compression or ischemia of the celiac artery by the median arcuate ligament. Characteristic features include weight loss and postprandial pain, and the diagnosis can be confirmed by radiographic imaging. Surgical treatment can be curative. MALS is a diagnosis of exclusion and requires a high level of suspicion. Herein we present three unique cases of patients with chronic abdominal pain discovered to have MALS and we review the literature.

Keywords celiac artery; ischemia; abdominal pain; median arcuate ligament

1. Introduction

Abdominal pain is the leading gastrointestinal complaint causing patients to seek medical care in the United States [1], accounting for an estimated 15.9 million office visits per year in 2009 [2]. Many experienced medical practitioners may concur that the etiology of the abdominal pain is sometimes cumbersome and often times is left undetermined. The spectrum of diseases that can manifest as chronic abdominal pain is wide, ranging from psychosomatic etiology to malignancy with more than one third being diagnosed with a functional gastrointestinal disorder, which encompasses irritable bowel syndrome (IBS) chronic idiopathic constipation and functional dyspepsia [3]. The etiologies of abdominal pain are expansive and include gastrointestinal disturbance, genitourinary pathology, infections, inflammatory disorders or vascular phenomena. Variables that assist in diagnosis of abdominal pain include age, gender, and location of symptoms. Many patients presenting with the complaint of chronic abdominal pain undergo extensive laboratory, endoscopic, and radiographic testing making the evaluation of this condition challenging for physicians who are trying to provide a comprehensive evaluation in a cost-effective manner. Often after an exhaustive and thorough evaluation, both patients and physicians are left frustrated and without a satisfactory diagnosis.

This case series highlights the importance of including median arcuate ligament syndrome (MALS) in the differential diagnosis when evaluating chronic abdominal pain patients. MALS is a rare pathological entity that is well described and requires a high clinical suspicion as well as a thorough history and physical exam in order to make an accurate diagnosis. Appropriate diagnostic testing is necessary to identify MALS since there are multiple treatment options available that may significantly improve a patient’s quality of life.

1.1. Pathophysiology of MALS

MALS, also known as celiac artery compression syndrome (CACS) or Dunbar syndrome, was first characterized by Dr. David Dunbar in 1965 when he wrote a case series including fifteen young patients [4]. In his article, he described isolated partial obstruction of the proximal celiac artery manifested clinically as postprandial abdominal discomfort, weight loss, and audible epigastric bruit [4]. Anatomically, the crura of the diaphragm are bridged by a fibrous tissue that overlies the aorta just proximal to the trunk of the celiac artery, the median arcuate ligament, and this anatomic feature is the culprit causing MALS [5]. The pathophysiology is incompletely understood and thought to be related to both neuropathic [6] and ischemic [4] mechanisms. One hypothesis is that the median arcuate ligament has a compressive effect on the celiac trunk causing pain and ischemia [7]. Another proposed mechanism for the pain in MALS is direct irritation of the celiac plexus, which lies in close proximity to the celiac artery [8]. Stimulation of the bundle of nerves that comprise the celiac plexus may either prompt inappropriate stimulation of pain fibers resulting in abdominal discomfort [9] or induce fibrotic changes such as celiac plexus neuromas that subsequently constrict the celiac trunk [10].
1.2. Clinical evaluation

Most patients with MALS will have undergone an extensive evaluation of abdominal pain by the time they are referred to a gastroenterologist. Common symptoms of patients with MALS include weight loss, postprandial abdominal pain, nausea, and vomiting. According to a systemic review on MALS, 80% of patients present with abdominal pain and up to 48% of patients experience weight loss [11]. The physical exam is usually unremarkable other than the objective weight loss and an epigastric bruit that has been reported in 35% of patients with MALS [11]. A thorough investigation of common etiologies of abdominal pain and weight loss should be performed (Table 1), prior to diagnosing MALS.

1.3. Radiographic imaging

MALS is a diagnosis of exclusion and as such, one must investigate ordinary causes of abdominal pain and weight loss before settling on MALS (Table 2). The diagnosis requires radiographic imaging to confirm celiac artery compression by the median arcuate ligament. Evidence of celiac artery stenosis is typically identified by the demonstration of vessel narrowing and postcompression dilatation, with either ultrasonography with Doppler [12,13,14], CT angiography, or on angiography [15,16]. In a study by Gruber et al., criteria of measured peak intraluminal systolic expiratory velocities higher than 350 cm/s and deflection angles of the celiac trunk greater than 50 degrees yielded an 83% sensitivity and 100% specificity for diagnosing MALS [12]. Unfortunately the diagnosis is complicated by the fact that 10–24% of normal, asymptomatic individuals may have radiographic evidence of median arcuate ligament compression of the celiac artery [16] while only 1% of these individuals endorse symptoms of MALS, highlighting the importance of clinical correlation in making the diagnosis. According to the published literature, the classic demographics of those affected with this syndrome are typically thin women between the ages of 20 and 40 years old. Once the diagnosis is established there are three treatment options available: decompression of the celiac artery, removal of the celiac ganglia or celiac revascularization, which can be done independently or in combination.

2. Case presentations

2.1. Case 1

A 56-year-old female with a past medical history of rheumatoid arthritis, anxiety, depression, anaplastic ependymoma status post resection with shunt placement, presented with a chief complaint of abdominal pain for two years duration. The abdominal pain was located in the mid-epigastric status post resection with shunt placement, presented with a chief complaint of abdominal pain for two years duration. The abdominal pain was located in the mid-epigastric chief complaint of abdominal pain for two years duration. The abdominal pain was located in the mid-epigastric chief complaint of abdominal pain for two years duration. The abdominal pain was located in the mid-epigastric chief complaint of abdominal pain for two years duration. The abdominal pain was located in the mid-epigastric

Table 1: Common etiologies of abdominal pain and weight loss.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Right upper quadrant</th>
<th>Left upper quadrant</th>
<th>Right lower quadrant</th>
<th>Left lower quadrant</th>
<th>Epigastric</th>
<th>Nonspecific/diffuse</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastrointestinal</td>
<td>Biliary colic, cholelithiasis, cholecystitis, pancreatitis, hepatitis</td>
<td>Peptic ulcer disease, gastritis, pancreatitis</td>
<td>Inflammatory bowel disease (ileitis), appendicitis</td>
<td>Diverticulitis, inflammatory bowel disease (pseudomembranous)</td>
<td>Gastroesophageal reflux disease, peptic ulcer disease, gastritis, pancreatitis, diffuse esophageal spasm</td>
<td>IBS, functional dyspepsia, celiac disease</td>
</tr>
<tr>
<td>Vascular</td>
<td>Budd-Chiari syndrome</td>
<td>Mesenteric ischemia, splenic infarct</td>
<td>Mesenteric ischemia, ischemic colitis</td>
<td>Mesenteric ischemia, ischemic colitis</td>
<td>Aortic dissection, MALS, myocardial infarction</td>
<td>Polyarteritis nodosum</td>
</tr>
<tr>
<td>Genitourinal</td>
<td>Pyelonephritis, nephrolithiasis</td>
<td>Pyelonephritis, nephrolithiasis</td>
<td>Cystitis, urinary retention, nephrolithiasis, pyelonephritis</td>
<td>Nephrolithiasis, pyelonephritis</td>
<td>Pericarditis</td>
<td></td>
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<tr>
<td>Inflammatory (non-GI)</td>
<td>Pleuritis, pneumonitis</td>
<td>Pleuritis, pericarditis, pneumonitis</td>
<td>Splenic abscess, Typhilitis</td>
<td>Ectopic pregnancy, salpingitis, pelvic inflammatory disease, endometriosis</td>
<td>Salpingitis, ectopic pregnancy, pelvic inflammatory disease, endometriosis</td>
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<tr>
<td>Infectious</td>
<td>Liver abscess, pneumonia, ascending cholangitis</td>
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<tr>
<td>Gynecologic</td>
<td>Splenic abscess</td>
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Table 2: Etiology of abdominal pain by gender.

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</tr>
<tr>
<td>Genitourinary</td>
<td>Cystitis, urinary tract infection, nephrolithiasis, pyelonephritis, sexually transmitted infection</td>
</tr>
<tr>
<td>Gynecological</td>
<td>Ectopic pregnancy, salpingitis, pelvic inflammatory disease, endometriosis, ovarian torsion, ruptured ovarian cyst, uterine perforation</td>
</tr>
<tr>
<td>Vascular</td>
<td>Hernia, MALS, mesenteric ischemia, ischemic colitis, Budd-Chiari syndrome, aortic dissection, myocardial infarction</td>
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patient reported postprandial bloating, nausea, indigestion, and heartburn. Over the last six months, the patient reported a 10 lb unintentional weight loss. The evaluation had included multiple esophagogastroduodenoscopies (EGD), colonoscopies, computerized tomography (CT) scan of the abdomen/pelvis, and magnetic resonance (MR) enterography, all of which had been negative. Additionally, a gastric emptying study and porphyria screen were negative. An abdominal ultrasound with Doppler analysis revealed focal elevation of peak systolic velocity in the celiac axis on expiration as compared to inspiration, which was compatible with MALS.

On exam, BMI was 22.17 and vital signs were stable. The patient was well developed, well nourished, well hydrated, and in no acute distress. Her physical exam was normal including the abdominal exam with normal bowel sounds and no tenderness with deep palpation. There was no organomegaly, palpable masses or lymph nodes present and no audible bruits in the abdomen. Exam was negative for guarding, rebound tenderness, and ascites. There was no halted inspiration due to pain with palpation of the right upper quadrant, consistent with a negative Murphy’s sign.

The patient’s laboratory results were unremarkable with pertinent negatives including normal thyroid stimulating hormone (TSH) and negative anti-tissue transglutaminase (anti-TTG) antibody.

CT angiography of the abdomen and pelvis were performed to further characterize the location of compression. The CT angiogram of the abdomen and pelvis revealed slight kinking of the proximal celiac artery distal to the takeoff from the abdominal aorta with expiration that likely reflected celiac artery compression syndrome (Figures 1(a) and 1(b)). The patient underwent a laparoscopic assisted median arcuate ligament release, after which the patient reported improvement in symptoms and decreased pain.

2.2. Case 2
A previously healthy 25-year-old male presented to the emergency room with abdominal pain unresponsive to a proton pump inhibitor, reflux symptoms, and hemoptysis. The patient stated that symptoms began after receiving treatment with steroids for a salivary gland infection. Upon further questioning, the patient reported unintentional 35 lb weight loss and postprandial abdominal pain intermittently over the past six months. The abdominal pain was located in the lower mid right quadrant and migrated to the left abdomen below the umbilicus. He also reported excruciating bilateral anterior rib pain. A chest X-ray was performed and
revealed pneumonia and the patient was prescribed an antibiotic. A CT scan of the abdomen and pelvis revealed a narrowing of the proximal celiac artery with poststenotic dilatation and suggestion of mild increased collateral flow consistent with MALS (Figures 2(a) and 2(b)). The patient was referred to the outpatient gastroenterology clinic for further work up and management.

On exam, the patient was thin with a BMI of 19.57. He had normal vital signs. The patient was well developed, well nourished, well hydrated, and in no acute distress. The patient’s physical exam was unremarkable including a soft, nontender, nondistended abdomen, without hepatosplenomegaly, bruits or masses and normal bowel sounds. The patient had a normal TSH and anti-TTG, ruling out hypo/hyperthyroidism and celiac disease as etiologies of abdominal pain and weight loss, respectively.

An abdominal ultrasound with Doppler was performed to further investigate the findings seen on CT scan, as well as an EGD to further work up the intractable heartburn symptoms. The endoscopy with biopsies was negative for celiac disease. The ultrasound findings revealed elevated peak systolic velocities and narrowing at the origin of the celiac artery, which decreased with standing yielding straightening of the celiac artery. The patient underwent an interventional radiologic celiac plexus blockade using bupivacaine, for diagnostic confirmation. The celiac block confirmed improvement of symptoms and he then underwent surgical release of the median arcuate ligament with resolution of symptoms.

2.3. Case 3
A 17-year-old male with a history of chronic and constant midabdominal pain since childhood, but worse over the past four months, presented for evaluation. The symptoms were worse in the morning and aggravated by meals, and alleviating factors included playing soccer and sleeping. The patient had undergone an EGD and small bowel-follow through study, which revealed an ulcer in the body of the stomach, for which he was started on an H2 blocker with some relief. He had recently presented to an emergency room with complaints of abdominal pain and a CT scan that showed a “blockage of the bowel” for which he was given a prescription for MiraLax and instructions to followup with the gastroenterology clinic as an outpatient. The exam revealed a well-developed, well-nourished, well-hydrated male with a BMI of 21.03 in no acute distress. His exam was unremarkable and the abdomen was soft, tender, nondistended, and free of hepatosplenomegaly or masses. Normal bowel sounds without bruits appreciated. The patient’s serologies were negative for celiac disease and thyroid abnormalities.

A gastric emptying study was normal. Abdominal ultrasound with Doppler revealed a kinked celiac artery on expiration at the origin of the vessel (Figure 3(a)). The
Figure 3: Ultrasound and flow velocity characteristics of MALS. (a) Ultrasound showing the hooked celiac artery at end expiration. (b) Peak flow velocity at end inspiration. (c) Peak flow velocity at end expiration I markedly increased.

narrowing improved with deep inspiration and the increased velocities were consistent with MALS (Figures 3(b) and 3(c)). A CT scan of the abdomen and pelvis was ordered and revealed a minimal impression on the celiac axis by the MAL but no associated poststenotic dilatation. The patient underwent laparoscopic celiac artery decompression with initial improvement of symptoms. Six months after the laparoscopic procedure, the patient started to have similar complaints of abdominal pain that he experienced prior to the surgery. A repeat CT scan with angiography revealed a patent celiac axis that was similar to prior exam. Specifically there was mild indentation on the celiac axis without significant poststenotic dilatation.

3. Discussion

There is no established gold-standard tool or regimen to diagnose MALS. It is a diagnosis of exclusion and therefore a thorough evaluation must be performed prior to concluding that MALS is the etiology of abdominal pain and weight loss. One author used ultrasonography to screen and diagnose patients with MALS [12]. Deflection angles and peak flow velocities were measured using ultrasound, with maximum expiratory peak flow velocities of greater than 350 cm/s and deflection angles greater than 50 degrees, being a reliable indicator for MALS [12]. The flow velocity and the angle of the celiac artery are influenced by diaphragmatic excursion during respiration and therefore were used in the proposed diagnostic criteria of MALS. Another noninvasive modality for diagnosis of MALS includes a single injection, high-pitch dual-source computed tomography angiogram during both inspiratory and expiratory phase [17]. This technique produces images similar to conventional angiography but with a lower contrast load and less risk of contrast induced nephropathy. Other proposed diagnostic tools include fractional flow reserve and intravascular ultrasound (IVUS) [18], gastric exercise tonometry [19], and magnetic resonance angiography; however, the optimal diagnostic study remains unknown. The use of IVUS was described by Vasquez de Lara and colleagues, as a confirmatory diagnostic tool when CT and MRA results were inconclusive [20]. In their report [20], the MRA showed 40–50% stenosis of the celiac artery, while the ultrasound showed 72% stenosis. Sufficient data is lacking concerning the sensitivity of IVUS; however, the aforementioned case report suggests that IVUS may have increased ability to detect the narrowing compared to traditional CT or MRA imaging. Measuring intestinal perfusion via gastric tonometry has been utilized to predict whether surgical intervention might improve a patient’s symptoms [21]. Due to the obscurity and under-diagnosis of the disease, the epidemiology remains poorly delineated and at-risk populations are not defined.

Optimal treatment of MALS is difficult because the precise mechanism involved with the manifestation of abdominal pain is unknown. It remains unclear to what extent ischemia plays a role in the symptomatology of MALS. Some patients demonstrated significant improvement after revascularization of the celiac artery to treat MALS, suggesting that a contributing component of the pain associated with MALS is due to ischemia. Response of this condition to celiac plexus blockade or celiac ganglion sympathectomy, emphasizes the fact that a neurological component may also be contributing to the abdominal pain in MALS [22]. The first minimally invasive treatment of MALS occurred in 2000 [23], and since then there have been multiple case series describing the efficacy of this approach.
compared to open surgical technique [24]. There are likely multiple contributing factors that cause the symptoms of MALS and once more is learned about its pathophysiology, treatment can be optimized.

In all three patients, treatment helped confirm the diagnosis. The resolution of pain after nerve block suggests that the irritation of the celiac plexus plays a predominant role in the pathophysiology of this syndrome, as does celiac artery decompression, both of which decrease stimulation of the celiac plexus. It should be noted that the third patient who presented did have recurrence of his abdominal pain after laparoscopic release of the median arcuate ligament. Given the questionable findings on imaging, the authors suspect that the patient did not have MALS and was in the 10–24% of patients who have evidence of median arcuate ligament compression of the celiac artery but without symptoms [16], highlighting the importance of ruling out other etiologies before making the diagnosis. Our cases demonstrate three patients with diverse demographics and clinical presentations who had different treatment approaches with two out of three patients experiencing complete resolution of symptoms.

Conflict of interest The authors declare that they have no conflict of interest.

References


