

CASE REPORTS

Dunbar syndrome as a cause of chronic abdominal pain in a 24-year-old female

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ABSTRACT

External compression of the celiac artery by the median arcuate ligament is referred to as Dunbar syndrome, which is an eponym for celiac axis syndrome or median arcuate ligament syndrome. It is correlated with the archetypal triad of postprandial abdominal pain, weight loss of greater than 20 pounds, and an abdominal bruit on auscultation. This is often accompanied by epigastric tenderness, vomiting, and nausea. Given its lack of symptomatic specificity, Dunbar syndrome is a diagnosis of exclusion for unexplained episodic abdominal discomfort. Here, we present a unique case of a 24-year-old woman who experienced several months of chronic abdominal distress and an extensive workup prior to being diagnosed with Dunbar syndrome. The diagnosis was made via cross-sectional abdominal imaging and duplex ultrasound with respiratory maneuvers, which showed downward displacement of the celiac trunk, post-stenotic dilatation, and increased flow velocity on expiration. She underwent successful laparoscopic division of the median arcuate ligament which greatly alleviated her pain.

Key Words: Dunbar syndrome, Median arcuate ligament, Celiac trunk, Abdominal pain, Median arcuate ligament syndrome, Celiac artery compression syndrome

1. INTRODUCTION

The diaphragm is composed of bilateral musculotendinous crura that connect via the median arcuate ligament. Normally, the median arcuate ligament forms the anterior border of the aortic hiatus between T11-L1, which is superior to the emergence of the celiac trunk from the aorta.^[1] Alternate localization of the median arcuate ligament or the celiac axis makes this group of vessels prone to compression, a condition known as Dunbar syndrome.^[2] In some instances, symptoms may be due to involvement of the celiac plexus rather than blood flow.^[3] The majority of cases involve abnormal positioning of the celiac axis (90%) rather than anatomical variations in the median arcuate ligament.^[4]

Compression of the celiac axis has been found in one third of

autopsy patients.^[5] Despite that, very few of these patients displayed clinical symptoms due to insignificant hemodynamic stenosis or ample perfusion via anastomotic circulation by other mesenteric vessels.^[6] The incidence of Dunbar syndrome is estimated to be 2 per 100,000 population, and it typically manifests in women between the ages of 40 and 60 with a thin build.^[2] When present, it most frequently involves postprandial abdominal pain (94%), weight loss (50%), and an abdominal bruit (35%).^[7,8] However, the lack of uniqueness of these clinical features earns Dunbar syndrome a low rank on the list of differential diagnoses for chronic episodic abdominal pain.

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2. CASE PRESENTATION

Earlier this year at an emergency room visit, a well-appearing and well-nourished 24-year-old Caucasian female showed 1+ epigastric tenderness, vomiting, and diarrhea. Because of her work in a health-care facility, she suspected exposure to an infectious agent as the root of her problems. Aside from her past medical history of meningitis, migraines, and mononucleosis, other aspects of her visit were unremarkable. To no avail, she was treated with a GI cocktail, hydrocodone-acetaminophen, lorazepam, ondansetron, hyoscyamine, and pantoprazole.

others, were ruled out.

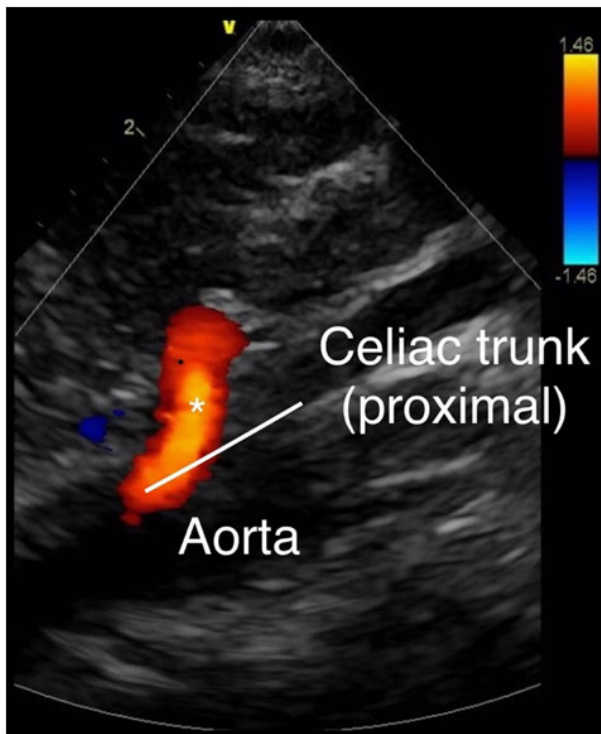


Figure 1. Duplex ultrasound of the proximal celiac trunk. Note the bright yellow intensification, designated by the “*.” This region is flanked by a more red-appearing segment, representative of slower flow

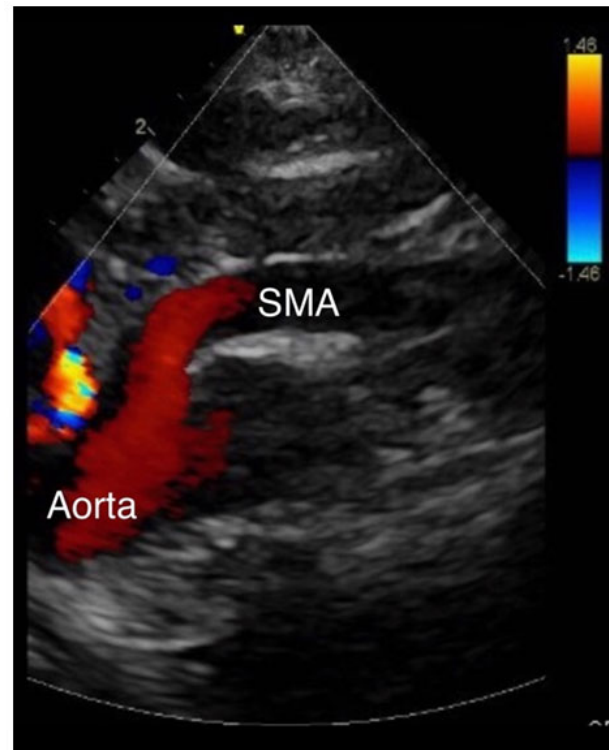


Figure 2. Duplex ultrasound of the SMA. Flow velocity in the SMA matches that in the aorta. Also, flow is uniform throughout the SMA.

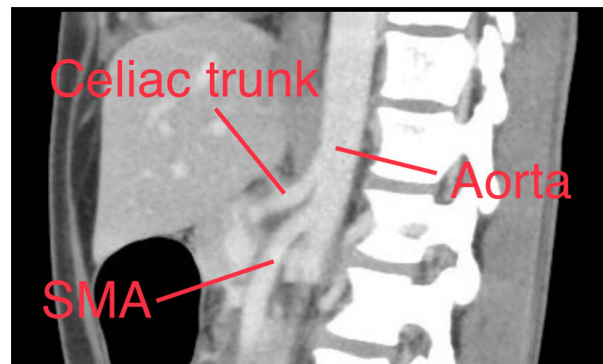


Figure 3. CTA of celiac trunk and SMA. The celiac trunk is narrowed near its origin while the SMA is patent.

Approximately 10 weeks later, she was admitted to the hospital with worsening abdominal pain, a decline decrease in weight of 16 pounds, increased frequency of nausea, and diarrhea with occasional melena. The pain was so debilitating that it prevented her from doing routine daily activities such as working and exercising. Stool studies, gastric emptying assay, blood and urine cultures, STD testing, RUQ ultrasound, CT enterography, EGD, dynamic cholecystography, and abdominal X-ray all failed to reveal any significant findings. Common causes of recurrent epigastric pain, including celiac artery atherosclerosis, gastric mobility diseases, infectious diseases, carcinoid syndrome, pancreatitis, and IBS amid

Exclusion of more common diseases led to suspicion of Dunbar syndrome. Doppler ultrasonography illustrated increased flow velocity through the celiac artery with expiration (356/156 cm/s) compared to inspiration (224/68 cm/s). Flow velocity was normal through the superior mesenteric artery (see Figure 2). CTA and T1 MRI showed downward displacement of the trunk and greater than 50% reduction of vessel diameter at the region of focal stenosis (see Figures 3 and 4). The combination of these two findings is consistent

with Dunbar syndrome.^[9] Robotic-assisted median arcuate ligament release alleviated the compression that the ligament had on the celiac axis. Several days after the operation, the patient appeared to be doing well and denied any abdominal pain. Following a 27-day hospitalization, she was discharged and able to live with much less pain than prior to admission.

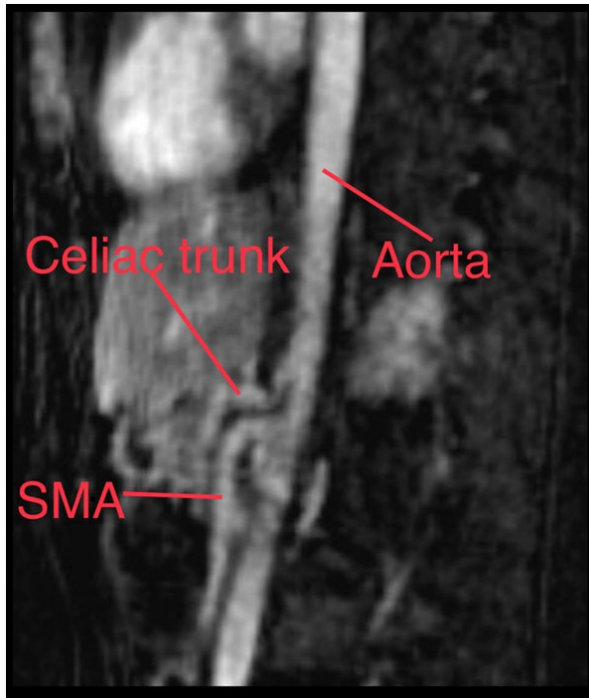


Figure 4. T1 MRI of celiac trunk and SMA. Note the downward displacement and narrowing of the proximal celiac trunk. Post-stenotic dilatation of the distal celiac artery is apparent.

3. DISCUSSION

Dunbar syndrome is rarely found in clinical patients, which limits the ability to for diagnosis. The mean time of diagnosis is 34 months, so earlier identification is crucial to lessening the debilitation that patients experience.^[10] Doppler ultrasound serves as a suitable screening method for Dunbar syndrome because it reliably shows focal narrowing with increased flow velocity through the celiac artery during end-expiration versus end-inspiration.^[11] The diaphragm is depressed during inspiration, which slackens the fibers of the

median arcuate ligament and mitigates the tension on the celiac trunk. Conversely, relaxation of the diaphragm during expiration causes its movement in the cranial direction, tightening the median arcuate ligament and heightening the transient celiac ischemia. Definitive diagnosis is governed by vascular imaging, notably CTA and MRA, that highlights a regional stenosis and inferior drooping of the celiac trunk.^[4] These exact findings in our patient led to Dunbar syndrome as the diagnosis.

Within the past decade, first-line treatment has changed from celiac artery stenting to robotic alterations of the ligament due to minimal invasiveness and immediate relief with very few side-effects.^[12] Although percutaneous revascularization is also possible, the beneficial effects of surgery are longer lasting.^[10] This approach proved successful in the patient discussed in our case presentation, who now has relief of upper abdominal pain.

Surgical treatment of Dunbar syndrome remains controversial today because of limited outcome data.^[2,9,10,12,13] Current predictors of successful surgical outcomes include age between 40 and 60 years, weight loss greater than 20 pounds, and post-prandial abdominal pain, of which our patient possessed only 1 out of 3 characteristics.^[13] Still yet, she was among the 85% of individuals with celiac artery compression who benefited from surgical intervention.^[14] This case study aids in identification of prognostic factors for this type of treatment. Continued monitoring of the patient via subsequent CTA scans is essential to track progression. There are no current medical treatments for Dunbar syndrome, so non-surgical care represents another area of future investigation.

4. CONCLUSION

Symptoms seen in celiac artery compression mimic intestinal angina and foregut ischemia, which can be caused by a myriad of diseases. Because Dunbar syndrome occurs so infrequently, case reports are crucial to augmenting its knowledge base. In a case involving chronic abdominal pain that continues after the exclusion of common etiologies, Dunbar syndrome may be a cause to consider.

CONFLICTS OF INTEREST DISCLOSURE

The authors have declared no conflicts of interest.

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