

Celiac Artery Compression Syndrome – A Unique Presentation

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Introduction

Celiac Artery Compression Syndrome (also known as Median Arcuate Ligament Syndrome, Celiac Axis Syndrome, and Dunbar Syndrome), is an anatomical physiologic disorder that results from compression of the celiac artery by the median arcuate ligament, a fibrous arch that forms at the base of the diaphragm where the right and left diaphragmatic crura join medially to form the anterior portion of the aortic hiatus. The following is a case that depicts an interesting presentation of this disease.

Case Description

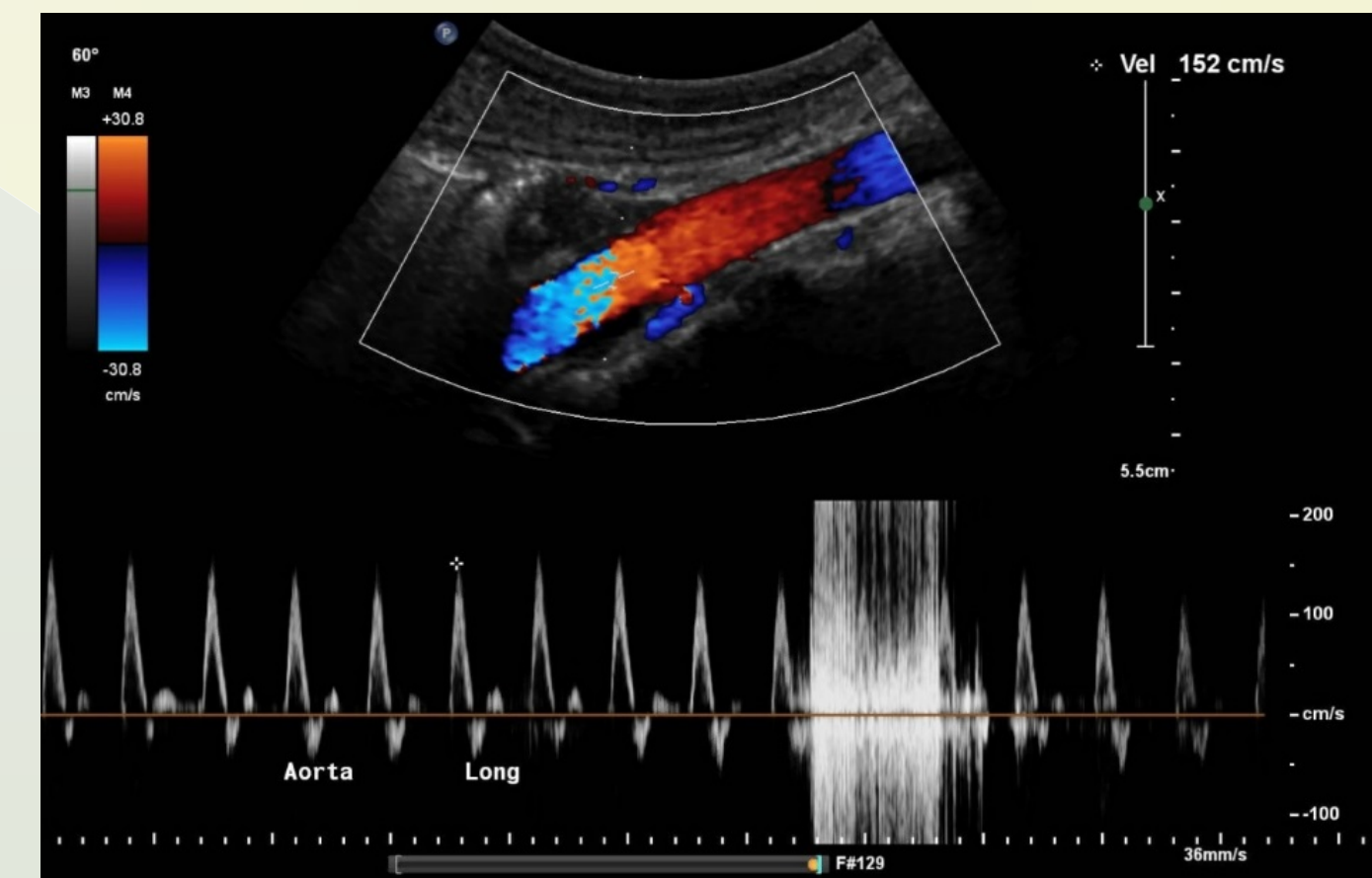
A 44 year old male with past history of cognitive delay was brought in by his family for significant weight loss with associated intermittent abdominal pain, weakness and lethargy over a period of years. Per the family, he used to be overweight 4 years ago. The patient has been living in Haiti his entire life, and recently migrated to the US 1 month ago. Per the patient’s family, he has had chronic abdominal pain for 5 years associated with weight loss of nearly 100 lbs. His family also reports that has poor nutritional intake, and is only able to eat a small amount before he seems to be in pain, then refuses to eat. He also exhibits constipation, passing small hard stool once every few days. He has no other prior medical history, no family history of malignancy, no history of trauma or surgeries, no history of smoking or substance use, and does not take any medications. The gastroenterology team was consulted to investigate the etiology of abdominal pain and chronic weight loss.

The patient is non-verbal, but does follow some commands. When asked to point to area of pain, he points to his epigastrium. He is unable to assist much further in answering questions.

His vital signs are normal. His height is 5’ 8”, weight is 90 lbs, and BMI is 13.7 kg/m². Physical exam reveals a thin cachectic male with scaphoid abdomen and mild tenderness in the epigastrium. His abdomen is otherwise soft and non-distended with normal bowel sounds, no abdominal bruit, no guarding or rebound tenderness, and Murphy’s sign could not be elicited. Skin is dry with tenting and mucous membranes are dry. There is no scleral icterus. Cardiopulmonary exam is normal. He is nonverbal, but cooperative and calm, with spontaneous movement observed in all extremities without evidence of focal weakness. There are no signs of skin rash or bruising.

Labs are significant for a mild normocytic anemia with Hgb 13.1 g/dL. Chemistry panel is normal. Hepatic function panel shows a mildly elevated aspartate aminotransferase and alanine aminotransferase at 60 U/L and 74, U/L respectively, mildly reduced albumin at 3.4 g/dL and elevated alkaline phosphatase at 199 U/L.

Abdominal duplex ultrasound and computed tomographic angiography is performed and is shown.



Figures:

For this patient, a mesenteric vascular duplex ultrasound revealed patency of the abdominal aorta with a peak velocity of 152 cm/second (Figure B), whereas examination of the celiac trunk demonstrated severe grade stenosis with pre-stenotic velocity of 152 cm/second, and a remarkably high post-stenotic velocity of 520 cm/second with a ratio of >3:1 (Figure A). Interestingly, the peak velocity did not vary significantly with respiration, which may indicate chronic fibrotic changes in the wall of the celiac artery secondary to compression over time. Subsequently, contrast enhanced abdominal CTA revealed acute angle J-configuration of the takeoff of the celiac axis, with stenosis (red arrow) at its origin and focal post-stenotic dilatation (Figure C). Thus, the diagnosis of celiac artery compression syndrome was made. The patient is currently pending surgical intervention.

Discussion

Celiac Artery Compression Syndrome can occur in the setting of anatomic anomalies such as an abnormally cephalad origin of the celiac trunk, or abnormally caudad insertion of the diaphragm. Physical extrinsic compression of the celiac trunk by the median arcuate ligament, as well as celiac ganglion plexus dysfunction leading to splanchnic vasoconstriction, is thought to cause intermittent mesenteric ischemia in the area of vascular distribution supplied by branches of the celiac trunk. The external compression is thought to be exacerbated during expiration, as cephalad motion of the diaphragm stretches the crura and increases tension at the median arcuate ligament; conversely, compression may be relieved during inspiration, where caudad movement of the diaphragm increases laxity of the crura. Recurrent compression of the celiac artery over time may lead to fibrotic changes in the arterial wall, resulting in chronic stenosis.

Although most patients are asymptomatic, those affected may clinically present with the classic triad of post-prandial abdominal pain and weight loss, and sometimes an abdominal bruit. Symptoms are often associated with nausea, vomiting, dyspepsia and reduced appetite.² Post-prandial pain is thought to result from intestinal angina, or increased intestinal oxygen demand that cannot be met due to celiac artery stenosis.

The diagnosis is one of exclusion, as a majority of patient are asymptomatic, but can be established by a combination of imaging modalities. Contrast enhanced computerized tomography angiography (CTA) or magnetic resonance angiography would demonstrate stenosis of the celiac axis secondary to extrinsic compression by the median arcuate ligament, and often post-stenotic dilatation. Abdominal duplex ultrasound with respiratory maneuvers demonstrating ≥70 percent stenosis of the celiac artery, a peak systolic velocity >200 cm/second, and worsening stenosis with expiration establishes the diagnosis. Less commonly utilized is invasive angiography with respiratory maneuvers. Treatment is achieved primarily through surgical decompression of the celiac trunk via open, laparoscopic or robot-assisted division of the median arcuate ligament, with select patients requiring intravascular procedures to relieve stenosis of the celiac artery.

Conflict of Interest and Financial Disclosure

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