

Median Arcuate Ligament Syndrome Involving a Celio-Mesenteric Trunk-a Lesson Learnt

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Abstract

Celio-mesenteric trunk (CMT) is a rare vascular variation of the ventral branches of the abdominal aorta that supply blood to the mesentery and the gut. This rare variation is seen in 2/100,000 population. The presence of this anomaly is associated with an increased risk of mesenteric ischemia in the case of proximal occlusion. Median arcuate ligament (MAL) syndrome is a controversial entity characterized by compression of the celiac axis by MAL causing post-prandial pain. We report the fourth case of MAL compression syndrome involving a CMT in the world

Keywords: Celio-mesenteric trunk, computed tomography, median arcuate ligament syndrome

Introduction

Vascular compression syndromes are a group of disorders characterized by external compression of healthy arteries or veins, as well as accompanying nerve structures. These syndromes can severely impact the quality of life in affected individuals, who are typically young and otherwise healthy. The celio-mesenteric trunk (CMT) is a rare anatomical variation where the celiac artery (CA) and the superior mesenteric artery (SMA) arise from a common origin. It occurs in 0.5% to 3.4% of the general population.^{1,2} This variation can complicate the planning and implantation of surgical procedures involving the areas supplied by these vessels. A new classification system for celio-mesenteric trunk based on multi-detector computed

tomography (CT) angiographic (CTA) results and possible embryological mechanisms has been proposed recently.³ CA branch stenosis or occlusion can accompany this anatomical aberrancy, but vascular compression syndrome of CMT by the median arcuate ligament (MAL) is extremely rare.⁴ It is important to be aware of this variation to avoid complications during interventions. Multi-detector row CTA is an excellent tool for detecting the celio-mesenteric trunk before surgical and interventional procedures. The author clarifies that written informed consent was obtained from the parents and the anonymity of the patient was ensured. The study submitted to this journal has been conducted in accordance with the Declaration of Helsinki and according to the requirements of all applicable local and international standards.



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Case Report

An 8-year-old male child was brought to the emergency reception of North Bengal Medical College and Hospital, West Bengal, India with complaints of intermittent abdominal pain that worsened with food intake. The pain was described by the parents as diffuse and intermittent and increasing in intensity within an hour of food intake due to which the child had barely eaten for the last year and had lost around 10 kg body weight. The child rated the pain as 7 on a scale of 0 to 10. The child was admitted to a local private hospital a few days back where after a lot of investigations including endoscopy a diagnosis of SMA syndrome was done on sonography and the patient was treated with a gastrojejunostomy. The parents could not show the imaging reports but the diagnosis was documented on the discharge certificate of the child. However, even after the surgery, the complaint persisted due to which the child was brought to our institute.

On examination, the child appeared malnourished and his weight for his age fell below the 3rd percentile as per the Indian Association of Pediatrics growth charts. Pallor was present but the rest of the general survey was unremarkable. An abdominal examination could not be done as the child was resisting it due to pain from the recent midline incision site. For the same reason, an ultrasound examination could not be performed after routine blood tests the patient was posted for a contrast enhanced CT (CECT) of the abdomen.

CECT abdomen revealed unremarkable solid organs and normal bowel loops. There was no evidence of intramural gas or bowel wall edema. A gastrojejunostomy was seen in place (**Figure 1**). A common CMT was identified on CECT opposite to the 1st lumbar vertebrae (**Figure 2**). It was classified as a Morita type 2 CMT. The left gastric artery was seen to arise as a thin branch below the CMT and was seen to anastomose with the left hepatic artery (**Figure 3**). The common trunk measured 8 mm and was seen to be compressed in its proximal part by the MAL with post-stenotic dilation (**Figures 4 and 5**). A 58% stenosis was identified. The patient was planned for a conventional angiography of the vessels but the parents declined due to their

concerns about radiation exposure. The patient was offered a laparoscopic release of the MAL to which they consented and was referred to Nil Ratan Sarkar Medical College, the apex center of pediatric surgery in the state where he underwent the procedure. Three months post-operative there is near total resolution of symptoms and the patient is eating well. He is under regular follow-up under the department of general surgery and awaiting a reversal of the gastrojejunostomy.



Figure 2. The first ventral branch from the abdominal aorta is seen to arise as a CMT (cursor over the origin of the CMT)

CMT: Celio-mesenteric trunk

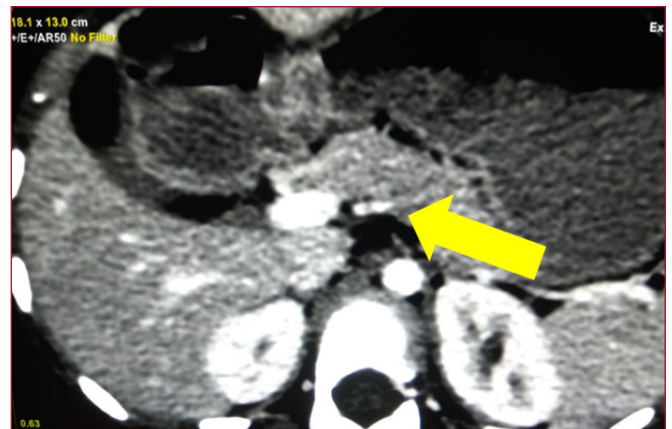


Figure 3. Left gastric artery (Yellow arrow) arising as a separate ventral branch of the abdominal aorta below the CMT

CMT: Celio-mesenteric trunk

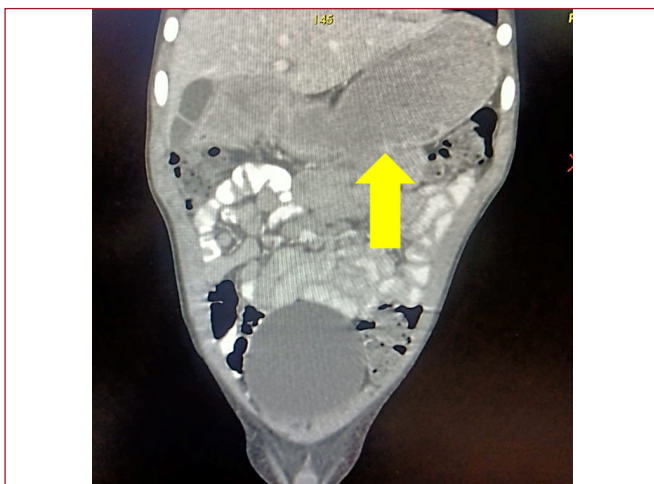


Figure 1. CT scan of abdomen was done and a gastro-jejunostomy was seen in place (Yellow arrow)

CT: Computed tomography

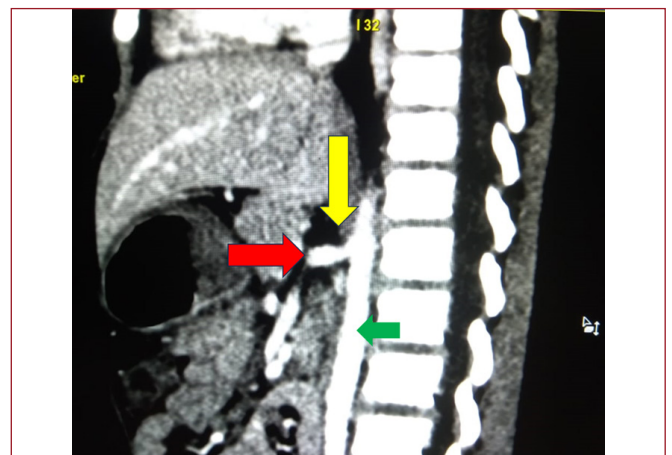


Figure 4. Compression of the proximal portion of CMT by the arcuate ligament (Yellow arrow) with post-stenotic dilation (Red arrow). Abdominal aorta (Green arrow)

CMT: Celio-mesenteric trunk

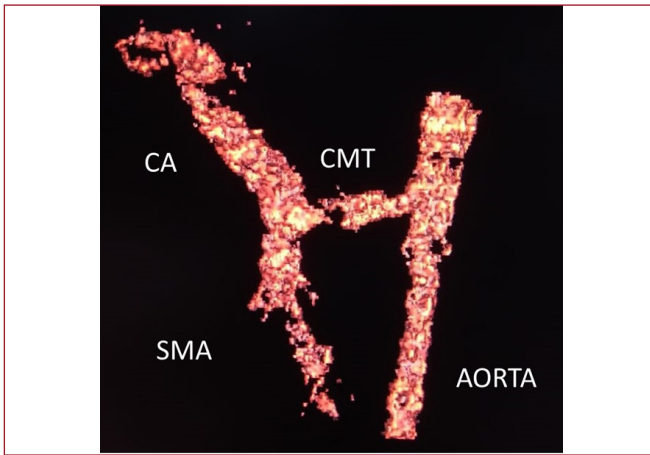


Figure 5. Angiographic anatomy after volume rendering of the vessels showing the aorta, CMT with focal compression, CA, and the SMA

CMT: Celio-mesenteric trunk, CA: Celiac artery, SMA: Superior mesenteric artery

Discussion

The MAL is a fibrous arcade that unites both crura of the diaphragm on either side of the aortic. It usually passes over the aorta over the CA origin. In between 10% and 24% of the population, ligaments may pass through the proximal part of the CA and cause indentation.^{1,5} Generally, CA arises as the 1st ventral branch of the abdominal aorta around 1 cm below the MAL however in a small subset of the population in about 10% to 24%, the ligament may cross over the proximal portion of the celiac axis and cause a characteristic indentation that is usually asymptomatic. Most such patients are asymptomatic however a rare controversial entity called MAL syndrome is described where compression of CA by an indenting MAL causes epigastric pain exacerbated by food intake, weight loss, and nausea.⁶ Although discovered several years ago the existence of this entity is still debated. The pathophysiology postulated is a combination of vascular occlusion and neuropathic pain due to compression of celiac ganglion which is increased during inspiration due to downward displacement of the diaphragm.^{5,6} The diagnosis of this condition is even more challenging due to the non-specific nature of the presentation. There is no consensus yet on the diagnostic criteria for this condition. Generally, doppler sonography of the CA in inspiration and expiration shows stenosis and turbulence in the post-stenotic dilated portion.⁶ In our case, the child would not allow a Doppler study due to the pain in his abdomen from the recent surgery,

CTA or conventional angiography is considered the gold standard for documenting compression. Axial sections are often inadequate to fully assess the CT findings typical with MAL compression. The best CTA plane to assess CA focal constriction is the sagittal plane. The distinctive hooked appearance of the focal narrowing can aid in differentiating it from other causes of CA constriction, such as atherosclerosis disease. Additionally, post-stenotic dilatation and large collateral arteries may be seen on CT.^{5,7} The CMT is a rare anatomical variation first described by Lipshutz. He used the term “truncus celiaco-mesenterica” to describe the variant anatomy found in two cadavers where the CA and SMA had a common origin from the abdominal

aorta. Due to the rare nature of the variant anatomy, the exact prevalence is known but it is estimated to be around 0.42% to 2.7%.^{6,8} The identification of this variant anatomy can have serious implications related to patient symptomatology. Dual origin of vessels and multiple mutual anastomoses have a protective effect against mesenteric ischemia if either of the major ventral branches of the aorta supplying the gut is blocked. In the case of a CMT this benefit is lost in case an obstructive or a stenotic lesion occurs at the common origin. Hence such a case would have serious ischemic consequences like acute or chronic mesenteric ischemia which we suspect was the cause of abdominal pain in our patient. To the best of our knowledge, this is the fourth case in the literature reporting a case of MAL syndrome associated with a CMT.^{5,6,9} Treatment is targeted towards the restoration of normal mesenteric vascular perfusion. Both open and laparoscopic management are done in various tertiary care centers. The MAL is divided along with the removal of the periarterial neural ganglionic tissue to relieve the neuropathic pain.^{5,10} SMA syndrome also known as Wilkie’s syndrome, is a condition that can cause duodenal obstruction. It occurs when the SMA compresses the third part of the duodenum due a reduced aorto mesenteric angle, leading to inadequate drainage of duodenal contents into the jejunum. This can result in weight loss and malnutrition. Aortomesenteric angles less than 22 to 28 degrees with an acute otitis media distance between 2-8 mm are strongly suggestive of SMA syndrome. Although sonography can be used as a rapid and effective modality to screen for the condition but definitive diagnosis can be made only on CTA or conventional angiography.¹¹⁻¹³

Conclusion

CMT is a rare vascular variant where the CA and SMA have a common origin. Due to a lack of collateral supply, a proximal occlusion can result in acute or chronic mesenteric ischemia. Clinical diagnosis of this condition is virtually impossible and diagnosis is based on radiological findings of CTA. Although to be considered as a diagnosis of exclusion we highlight the role of radiology in the successful diagnosis of MAL compression syndrome in a CMT along with its successful management. Also, we strongly discourage the use of sonography as a sole modality to diagnose SMA syndrome to guide treatment options and recommend all such cases to be considered for CECT abdomen to clearly define and describe the vascular anatomy.

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Conflict of Interest: The author has no conflicts of interest to declare.

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Informed Consent: The author clarifies that written informed consent was obtained from the parents and the anonymity of the patient was ensured.

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