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Case Report

Dunbar syndrome: a rare presentation of abdominal angina treated by revascularization of the celiac artery by endovascular stenting

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ABSTRACT

Median arcuate ligament syndrome (MALS) is a rare entity characterized by extrinsic compression of the celiac artery and symptoms of postprandial epigastric pain, nausea, vomiting, and weight loss mimicking mesenteric ischemia. The following case illustrates a rare cause of abdominal pain, where this young woman was found to have celiac trunk stenosis , secondary to compression of the trunk by the median arcuate ligament. She underwent a successful stenting to the ostial celiac trunk, thus reliving her symptomatically. Decompression of the celiac artery is the general approach. Usually post PTA, once revascularisation is achieved, 75% of the patients remain asymptomatic at follow up.

Keywords: Celiac artery compression syndrome, Celiac axis syndrome, Dunbar syndrome, Median arcuate ligament syndrome

INTRODUCTION

The median arcuate ligament (MAL) is located at the T12-L1 level and bridges the crura of the diaphragm, just anterior to the aorta. Dunbar's syndrome, median arcuate ligament syndrome (MALS) or celiac artery compression syndrome (CACS) is caused by external compression of the celiac trunk by the MAL, and is characterized by postprandial abdominal pain, nausea, vomiting, and weight loss.[1] MALS is a rare entity, which is diagnosed in only 2 of 100,000 patients with ambiguous upper abdominal pain.[2]

The diagnosis of MALS is one of exclusion, as many healthy patients demonstrate some degree of celiac artery compression in the absence of symptoms. Consequently, a diagnosis of MALS is typically only entertained after more common conditions have been ruled out. Once suspected, screening for MALS can be done with ultrasonography and confirmed with computed tomography (CT) or magnetic resonance (MR) angiography.

CASE REPORT

A 38-year-old, female, presented with recurrent episodes of abdominal pain, aggravated on consumption of food since 4 years. Pain was continuous in nature and with severity of 6/10 on a pain scale of 0-10. Patient also reported exacerbation of pain with eating but with no relieving factors. Patient also reported anorexia and 10kgs weight loss over six months period. She had consulted several doctors for the same but was not relived of her symptoms and put on antacids and proton pump inhibitors. Her general physical examination was unremarkable except that her BMI was 19kg/m^2, just at the lower limit of normal. Her respiratory system and cardiovascular system examination were normal, but on

per abdomen examination there was a faint bruit detected between the xiphisternum and umbilicus. Her blood investigations were within normal limits, ECG and 2D Echo both normal. Esophagogastroduodenal endoscopy revealed gastric and duodenal distension with slow peristalsis. Colonoscopy was normal. Her CECT Abdomen revealed a near total occlusion of the celiac axis due to extrinsic compression by the median arcuate ligament (Figure 1).

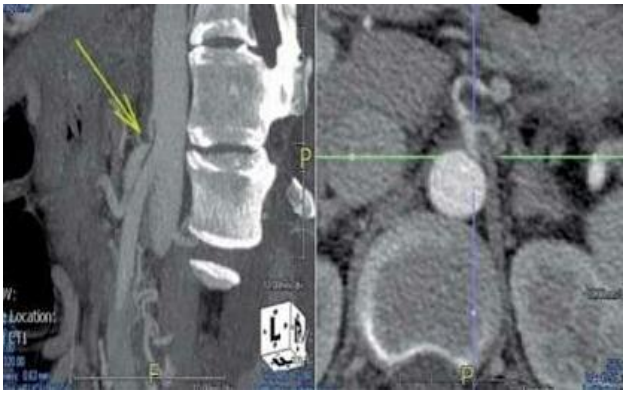

Figure 1: CECT abdomen revealed a near total occlusion of the celiac axis as depicted by the arrow mark.

Her peripheral angiogram reveled 80% ostial celiac trunk stenosis and she was taken up for PTA stent to the celiac trunk. Her stenting was successful, post which the patient was symptomatically better and relieved of her abdominal pain (Figure 2). The post procedure period was unremarkable, and the patient was discharged on day 3. The patient was symptom-free at 12 months after endovascular stenting.

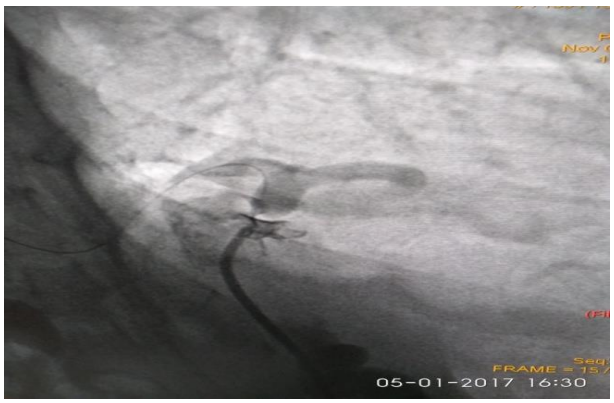

Figure 2: Peripheral angiogram reveled 80% ostial celiac trunk stenosis which was treated by revascularization of the celiac artery by endovascular stenting.

DISCUSSION

Dunbar syndrome is a rare and controversial vascular compression syndrome, characterized by postprandial intestinal angina caused by insufficient blood supply from the celiac artery to the gastrointestinal tract. The median arcuate ligament is a fibrous arch located anterior to the aorta and formed by a connection of the diaphragmatic crura. The celiac plexus is located between the arcuate ligament and the celiac trunk in up to 25% of normal individuals. Among other causes, the compression of the celiac trunk is due to a descensus of the diaphragm after period of accelerated growth in adolescents. The female to male ratio is 3:1 and the classic patient is a female aged between 18 and 30 years.[3-5]

The etiology of MALS remains unknown but a case report of monozygotic twins suggests a congenital origin.[6] Whether the pathophysiology is primarily vascular or neuropathic origin remains undetermined.[7] Isolated vascular compression of the celiac artery as the sole etiologic factor seems unlikely. First, in 10%-24% of the population the MAL can cause asymptomatic compression.[8] Second, collateral circulation by the superior mesenteric artery provides adequate blood supply; therefore, postprandial abdominal pain should not be expected with celiac artery compression alone.

The cause of MALS is likely multifactorial, including compressive effects on the celiac artery and surrounding neurogenic structures. In celiac artery compression, it has been noted that either the celiac artery is located slightly higher or the MAL is located lower than expected.[9] In a large series, significantly higher symptomatic relief was achieved through combined release of the MAL and revascularization.[10] If celiac artery compression alone is corrected, evidence suggests up to 53% will be asymptomatic on long-term follow up.[10] Combined release and revascularization however increases the long-term success to 79%.[10] Therefore some patients may require additional revascularization procedures such as mesenteric artery stenting or bypass to provide long-term symptomatic relief. Since delay of revascularization is unlikely to adversely affect outcome, most choose ligament release first, followed by revascularization via stenting or bypass if symptoms persist.[11]

Diagnosis can be made through Doppler ultrasound, spiral CTA, selective catheter angiography and magnetic resonance angiography. Doppler US has been cited as the best initial test for Dunbar Syndrome, whereas selective angiography and CTA are the gold standard diagnostic methods and can be used to create three-dimensional reconstructions [11]. When trying to differentiate between MALS and atherosclerosis, it is important to look for arterial calcifications and a characteristic hook like appearance. Presence of hook like appearance with absence of arterial calcifications is characteristic of MALS.

While historically MALS was treated primarily by vascular surgeons, the shift towards less invasive procedures has allowed general surgeons with training in minimally invasive surgery to operate more. Nonetheless,

injury to the celiac artery and bleeding is a serious risk and the operating team must be prepared to intervene.[12]

There is currently no medical treatment for MALS. In more extreme cases, those with post-stenotic dilatation or collateral vessels, surgery is recommended although it is not without controversy. Surgeons perform both laparoscopic and open surgical techniques to relieve the compression. More recently, the laproscopic approach has gained greater traction. Treatment of MALS include performing celiac angioplasty or endovascular stenting. Endovascular procedures include percutaneous transluminal angioplasty and stent implantation.[13]

CONCLUSION

In conclusion, Dunbar syndrome is a rare and uncommon cause of post prandial abdominal pain and should be kept in mind while evaluating the cause of unexplained gastrointestinal symptoms after eliminating all the other commonly encountered causes. Surgery is the is an effective approach to release the compression by the median arcuate ligament, although it is not without controversy. Hence endovascular procedures like percutaneous transluminal angioplasty and stent implantation as done in our patient is a low risk procedure with complete relief of symptoms and especially indicated in high risk patients unfit for surgery. This procedure can be performed uneventful in order to restore quality of life of the patient.

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REFERENCES

1. Loukas M, Pinyard J, Vaid S, Kinsella C, Tariq A, Tubbs RS. Clinical anatomy of celiac artery compression syndrome: a review. Clinical Anatomy. 2007 Aug 1;20(6):612-7.
2. Foertsch T, Koch A, Singer H, Lang W. Celiac trunk compression syndrome requiring surgery in 3 adolescent patients. J pediatric surgery. 2007 Apr 1;42(4):709-13.
3. Dunbar JD, Molnar W, Beman FF, Marable SA. Compression of the celiac trunk and abdominal angina: preliminary report of 15 cases. American J Roentgenology. 1965 Nov;95(3):731-44.
4. Gander S, Mulder DJ, Jones S, Ricketts JD, Soboleski DA, Justinich CJ. Recurrent abdominal pain and weight loss in an adolescent: celiac artery compression syndrome. Can J Gastroenterol. 2010;24:91-3.
5. Bobadilla JL. Mesenteric ischemia. Surg Clin North Am. 2013;93:925-40.
6. Bech F, Loesberg A, Rosenblum J, Glagov S, Gewertz BL. Median arcuate ligament compression syndrome in monozygotic twins. J Vasc Surg. 1994;19(5):934-8.
7. Lee V, Alvarez MD, Bhatt S, Dogra VS. Median arcuate ligament compression of the celiomesenteric trunk. J Clinical Imaging Science. 2011;1:8.
8. Williams S, Gillespie P, Little JM. Celiac axis compression syndrome: Factors predicting a favorable outcome. Surgery. 1985;98:879-87.
9. Reilly LM, Ammar AD, Stoney RJ, Ehrenfeld WK. Late results following operative repair for celiac artery compression syndrome. J Vasc Surg. 1985;2:79-91.
10. Vaziri K, Hungness ES, Pearson EG, Soper NJ. Laparoscopic treatment of celiac artery compression syndrome: case series and review of current treatment modalities. J Gastrointest Surg. 2009;13(2):293-8.
11. Karavelioglu Y, Kalcik M, Sarak T. Dunbar Syndrome as an unusual case of exercise-induced retrosternal pain. Arch Turk Soc Cardiol. 2014;43(5):465-76.
12. Roseborough GS. Laparoscopic management of celiac artery compression syndrome. J Vasc Surg. 2009;50(1):124-33.
13. Ng F, Wai O, Wong A, Yu S. Median arcuate ligament syndrome. Hong Kong Med J. 2016;22-184e:3-4.

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