SSR Institute of International Journal of Life Sciences
ISSN (O): 2581-8740 | ISSN (P): 2581-8732
Pal and Ray, 2024
DOI: 10.21276/SSR-IIJLS.2024.10.5.2

Case Report

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Median Arcuate Ligament Syndrome-A Rare Case of Abdominal Pain

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Received: 28 May 2024/ Revised: 29 Jun 2024/ Accepted: 30 Aug 2024

ABSTRACT

Background: Median arcuate ligament syndrome, a rare one in clinical entities, has clinical features like post-prandial abdominal pain, nausea, and vomiting with associated slow-onset weight loss. Due to this nature, a patient may present late, taking several years to get detected, and usually, it's a diagnosis of exclusion. Here we present a case of a 23-year-old male suffering from post-prandial abdominal pain, weight loss, and sitophobia for the last eight years, not relieved by any medical management.

Methods: A general physical examination was done, followed by a systemic examination, routine blood investigations, and radiological Imaging. After establishing the diagnosis, the patient had undergone laparoscopic release of the median arcuate ligament.

Results: He had a thin build and pallor on general physical examination. On systemic examination, no abnormal findings were noted. All routine blood investigations, Chest X-ray, ECG, and Echocardiography reports were found normal. On CT, the whole abdomen (plain and contrast) was noted with a thick right diaphragmatic crus with short segment stenosis of the coeliac trunk. The esophagogastroduodenoscopy study came out as a normal one. After that CT Mesenteric angiogram revealed a normal angiogram with focal luminal narrowing in the celiac trunk just distal to the ostium.

Conclusion: Median arcuate ligament syndrome is a very rare disorder, but every clinician should keep it in mind as a diagnosis of exclusion when all common etiologies of abdominal pain cannot be established. It can also present as an incidental diagnosis and also in a complicated status like abdominal bleeding as a chief complaint or PDAA.

Key-words: Median arcuate ligament, Pancreaticoduodenal artery aneurysm, Post-prandial abdominal pain, Retroperitoneal hemorrhage

INTRODUCTION

Median arcuate ligament syndrome (MALS) is an infrequent clinical entity. Multiple theories evolved over the ages to establish their cause. MALS ranges from second to sixth-decade age groups, having a prevalence of 2 in 100000 [1]. Male: Female ratio varies from 2:1 to 3:1 [2]. Lipshutz offered the first anatomic description of the celiac artery and Harjola performed the first surgical release of the ligament in 2019 [3].

How to cite this article
Pal R, Ray U. Median Arcuate Ligament Syndrome-A Rare Case of Abdominal Pain. SSR Inst Int J Life Sci., 2024; 10(5): 6123-6126.

Access this article online
https://iijls.com/

In 2019, Santos *et al.* reported a case series where the operative division of MAL resulted in clinical improvement in 13 of 15 patients [3]. After his name, it's also known as Dunbar syndrome [3]. Compression of the celiac artery by the median arcuate ligament is well documented.

This statement was published by Stanley and Fry in 1971 after reviewing "provocative xylose absorption tests" in ten female and five male patients over 3.5 years, transecting median arcuate ligament of those, they concluded intestinal ischemia was an underlying disorder for this syndrome [4]. Etiology of MALS has shifted from vascular to neurogenic cause with compression of the surrounding celiac plexus and ganglia. Clinically, patients may present commonly with post-prandial pain, nausea,

DOI: 10.21276/SSR-IIJLS.2024.10.5.2

vomiting, unintentional weight loss, exercise-induced pain, and sometimes with autonomic dysfunction, fibromyalgia, and postural orthostatic tachycardia syndrome [6]. But these are like the tip of the iceberg and a large group of patients remain asymptomatic.

CASE PRESENTATION

Our patient is a 23-year-old young male who presented in the Emergency with complaints of 3-4 episodes of non-bilious vomiting for the last 2-3 days with nausea and severe post-prandial pain. He also complained he has been facing vague abdominal pain and nausea of frequent episodes over the previous eight years. The pain was frequent and chronic, occurring most of the time after having large meals with nausea, for which he has developed a fear of taking food gradually. Pain was not associated with fever or melaena. He had undergone an open appendicectomy in childhood. He was allergic to brinjal, smoking 1-2 cigarettes occasionally for the last two years, and had normal bladder and bowel habits.

On admission, he had mild pallor, tachycardia (HR–118/min), BP-110/70 mm Hg, RR- 20/min, Temp 98.5°F, SpO_2 99% in RA. Higher functional were WNL. The chest was bilateral clear. The cardiovascular system was regular. On P/A examination, the abdomen was scaphoid, in the normal position of the umbilicus, having one scar post past appendicectomy over RIF, soft, nontender.

He was managed conservatively, and blood was sent for routine investigations, in which HB –11.3 gm/dl, rest WNL.

The esophagogastroduodenoscopy study was normal.

USG W/A -mild hepatomegaly noted.

CT W/A(Plain+ contrast) revealed a thickened right diaphragmatic crus of maximum thickness ~6.4 mm, noted with short segment stenosis of celiac trunk from its origin for a length of 3.6mm with ~45-50% luminal narrowing with post stenotic prominence of celiac trunk; mild hepatomegaly; nonspecific mesenteric lymphadenopathy.

CT Mesenteric Angiogram (Fig. 1) showed that the celiac axis is normal at its origin. Still, there is a focal indentation of the superior wall of the trunk just distal to its ostium with mild focal narrowing. Still, there is no wall irregularity, and branches of the celiac axis are normal.

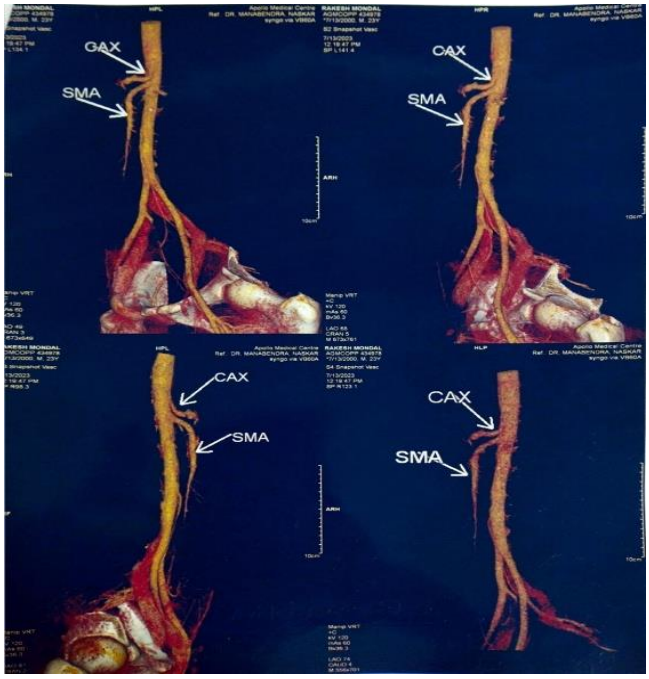

Fig. 1: CT Mesenteric Angiogram showing compression of celiac axis-hook or J sign

Subsequently, Diagnostic Laparoscopy plus Laparoscopic division of median arcuate ligament was done under general anaesthesia (Fig. 2). Standard five ports done àpneumoperitoneum created àliver retracted by liver retractorà dissection started from pars flaccida àleft gastric vessel traced to delineate celiac trunk all branches of celiac trunk noted and preserved àceliac trunk traced up to anterior border of the aorta and median arcuate ligament traced and dividedà Hemostasis secured àports closed with no. 1 vicryl àskin apposed with 3-0 rapid vicryl.

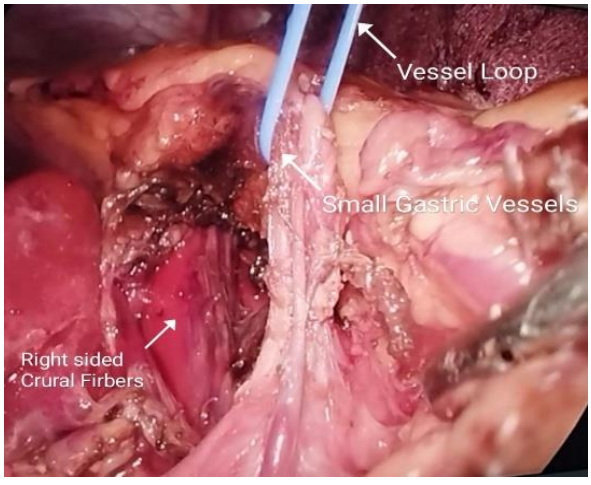

Fig. 2: Retraction of short gastric vessels by vessel loop

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The classical appearance of the coeliac axis at its origin is given in Fig. 3.

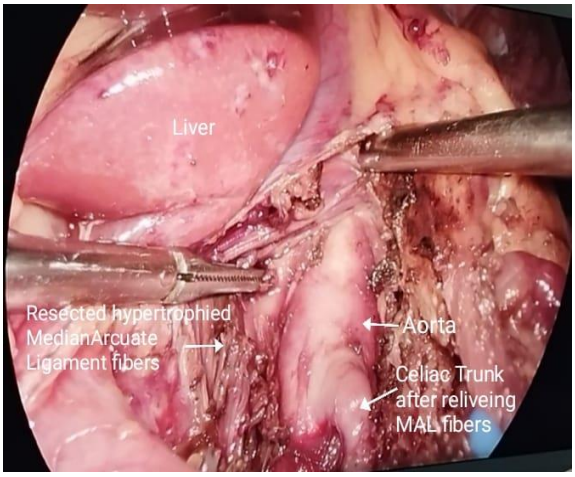

Fig. 4: Resection of hypertrophied Median Arcuate Ligament for relieving compression over celiac trunk

DISCUSSION

Median arcuate ligament (MAL) is a fibrous arch connecting the diaphragm's right and left crus [6]. Embryologically, part of the diaphragm descends from the neck toward the celiac axis between 9-12 weeks and forms MAL, crossing the aorta at the level of the celiac artery. These fibres splay over the aorta and extend laterally towards the suspensory ligament of the duodenum. Normally, these fibres cause no compression over the celiac artery, but in 10 to 24% of cases, there may be compression; however, less than 1% are symptomatic [5]. One theory states that compression of this artery compromises blood supply to the foregut, leading to post-prandial epigastric pain [7]. Another theory states that vascular steal syndrome causes pain by diverting blood from the superior mesenteric artery through collaterals to compensate for the inefficient supply of the celiac artery, causing midgut ischemia [7]. The celiac nerve plexus branches across the abdominal aorta between levels T11 to L1 and these are also thought to cause pain in MALS [8]. So, there may be many pathological ways to cause MALS. Hence, its symptoms overlap and are often nonspecific, creating diagnostic delays. Here also, our patient underwent extensive investigations for his pain over the years with medical treatments, which were all ineffective for his relief.

Due to its mimicking nature, other abdominal pathologies are diagnosed radiologically. Earlier, an invasive lateral aortogram was used to diagnose MALS, which is now replaced by an ultrasonographic colour Doppler, a noninvasive one. Peak systolic velocity of more than 200cm/s, no flow, and reversal of common hepatic artery flow in the expiratory phase indicate significant stenosis. However, the gold standard for MALS is now computed tomography angiography (CTA) with 3D reconstruction or magnetic resonance angiography (MRA), which can show very well Fish hook or J-shaped stenosis of celiac artery during the inspiratory phase [8].

If needed, MALS can be treated by open, laparoscopic and robotic division of MAL and celiac nerve plexus [7]. However, not all patients need surgery. Those asymptomatic patients with non-significant stenosis, whose pain is not post-prandial or associated with exercise, with vasculitis may have poor surgical outcomes. Open surgeries are associated most commonly with thrombophlebitis, gastroesophageal reflux, and cerebrovascular accidents. Robotics are not cost-friendly to most patients. The most common risks associated with laparoscopic surgery here are bleeding and pneumothorax. But, now, the laparoscopic division of MAL has emerged as the most feasible surgical treatment method in this era for MALS[9]. Intraoperative Doppler ultrasonography can show blood flow correction immediately after the MAL division.

Due to long-term celiac artery stenosis, it can form gastroparesis, aneurysm, or pseudoaneurysm of the pancreaticoduodenal artery (PDA) [10]. About 68% of cases of MALS had been seen to be associated with PDA Aneurysm (PDAA) [10]. Sometimes it can present as obstructive jaundice caused by PDAA or retroperitoneal hemorrhage due to a ruptured aneurysm, leading to hemorrhagic shock and death [10,11].

CONCLUSIONS

Median arcuate ligament syndrome though a rare entity, should always be excluded in a patient with postprandial pain, sitophobia, and vomiting where common diseases for abdominal pain could not be diagnosed. Due to better radiological facilities nowadays these cases are also diagnosed incidentally. The disease symptoms may range from an asymptomatic to PDAA and retroperitoneal haemorrhage. It is a rare and treatable cause of postprandial pain abdomen. A high index of suspicion and good imaging techniques clinch the diagnosis. Minimally invasive surgery has become the gold standard treatment. Adequate adhesiolysis and

DOI: 10.21276/SSR-IIJLS.2024.10.5.2

division of the ligament leads to remarkable results. Inadequate division and lateral clearance may lead to recurrence.

CONTRIBUTION OF AUTHORS

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Materials- Dr. Udipta Ray
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