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Median Arcuate Ligament Syndrome, A Rare Cause Of Post-Prandial Abdominal Pain And An Incidental Finding On Ct Scan--Case Series

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ABSTRACT

Median arcuate ligament syndrome constitutes a constellation of symptoms resulting from the compression of celiac axis by the Median Arcuate Ligament. It is a rare disorder presenting with symptoms like abdominal pain (including exercise induced or post-prandial), nausea, and vomiting and weight loss. According to the literature stenosis can be significant in 7% to 8% of the patients and result in ischemia of the bowel. It is therefore essential to understand the basic presentation and imaging of MAL syndrome. We have presented three such cases diagnosed on CT scan with contrast.

Keywords: *Median arcuate ligament syndrome, celiac Trunk Compression Syndrome, Dunbar Syndrome*

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INTRODUCTION

MAL syndrome is also known as the Dunbar syndrome or celiac artery compression syndrome[1-6].It was first described by Lipshutz in 1917 based on the anatomical variations noted during cadaveric examination. Later in 1963 Harjola also reported one such case which showed resolution of symptoms after surgical intervention[1].Anatomically the lumbar part of diaphragm is formed by the right and left crura.

The right crus of the diaphragm arise from anterior surface of L1 to L4 and the left from L1 to L3 anteriorly also from the intervertebral discs, medial margins of both crus form the aortic hiatus by passing anteriorly and superior to the aorta. Both crus have their insertion at the central tendon. Median arcuate ligament is the fibrous arch that unites both the crura on either side of the aortic hiatus. The median arcuate ligament that normally crosses the aorta at L1 vertebral level hence above the origin of the celiac axis [2]. In approximately 10-24 % individuals the median arcuate ligament is lower and hence causes compression of the celiac axis, so much so that it can result in arterial insufficiency with development of mesenteric ischemia. Due to the chronic nature of insufficiency most cases develop collaterals that prevent ischemia. According to some studies this can also be due to the high origin of the celiac axis. In either case the celiac supply is compromised resulting in symptoms[1,5].Celiac artery compression syndrome is more common among thin lean females about 4 times more than in men between 30 to 50 years of age[1-3]. Most of the patients present with symptoms of abdominal pain (94%), abdominal pain after meals (80%), weight loss (50%), nausea and vomiting (55.6%), bloating (39%) and exercise induced abdominal pain (8%) [1,4]. However some patients may be completely asymptomatic, as per studies about 16% of asymptomatic patients with an abdominal bruit on examination and 12 to 49% of asymptomatic patients have radiological evidence of MAL syndrome[4].

On examination most of the patients have an epigastricbruit that increases on expiration. Patients also have epigastric tenderness that is relieved by bringing the knees close to the chest as it decreases the pressure on the celiac axis[1-5]. The diagnosis of MAL syndrome is one of an exclusion, all the other causes of abdominal pain need to be ruled out by performing ultrasounds, endoscopy, gastric emptying studies, duplex ultrasound and CTA [1-4].Various approaches are now used to relieve the symptoms of MAL syndrome including Open Decompression, Laparoscopic MAL Release, Reconstruction, Endovascular Intervention and the latest technique using robotic-assisted MAL division with neurolysis of the celiac ganglion [1].Most of the patients show complete recovery after the treatment approaches.

CASE REPORT

CASE 1:

35 year old male known case of scleroderma presented to his primary physician with complains of nausea, vomiting and hematochezia. His abdominal examination was unremarkable. He had the typical findings of skin thickening and swelling around the fingers of both hands with joint pain. Since the patient was already a known case of scleroderma, his primary physician advised him a CT angiography to rule out the possibility of vasculitis after which he was referred to our department. The patient underwent CTA which was unremarkable for vasculitis. However an incidental discovery of median arcuate ligament syndrome was noted. His CT scan showed focal narrowing of celiac trunk at its origin giving a characteristic hooked appearance due to indentation on its superior surface as seen figure 1(1A, 1B & 1C).

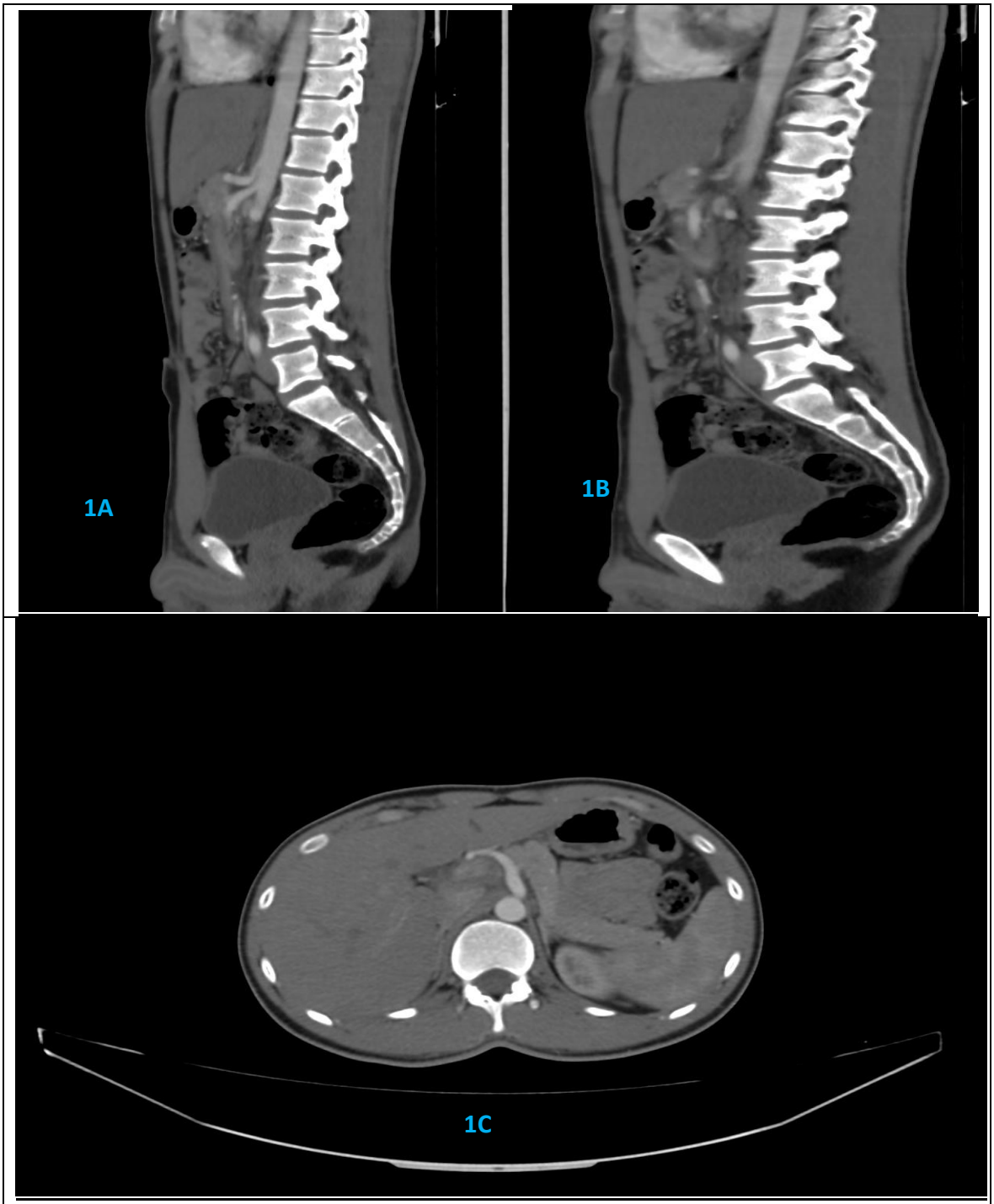

Figure 1 CT abdomen demonstrating acute angulation with indentation upon thre superior aspect of celiac axis

His 3D re-constructed images also show the characteristic appearance of celiac trunk compression (figure 2). Since the patients CT angiography was negative for vasculitis and he did not have any symptoms related to median arcuate ligament syndrome, he was managed conservatively and was advised follow-up by his treating physician.

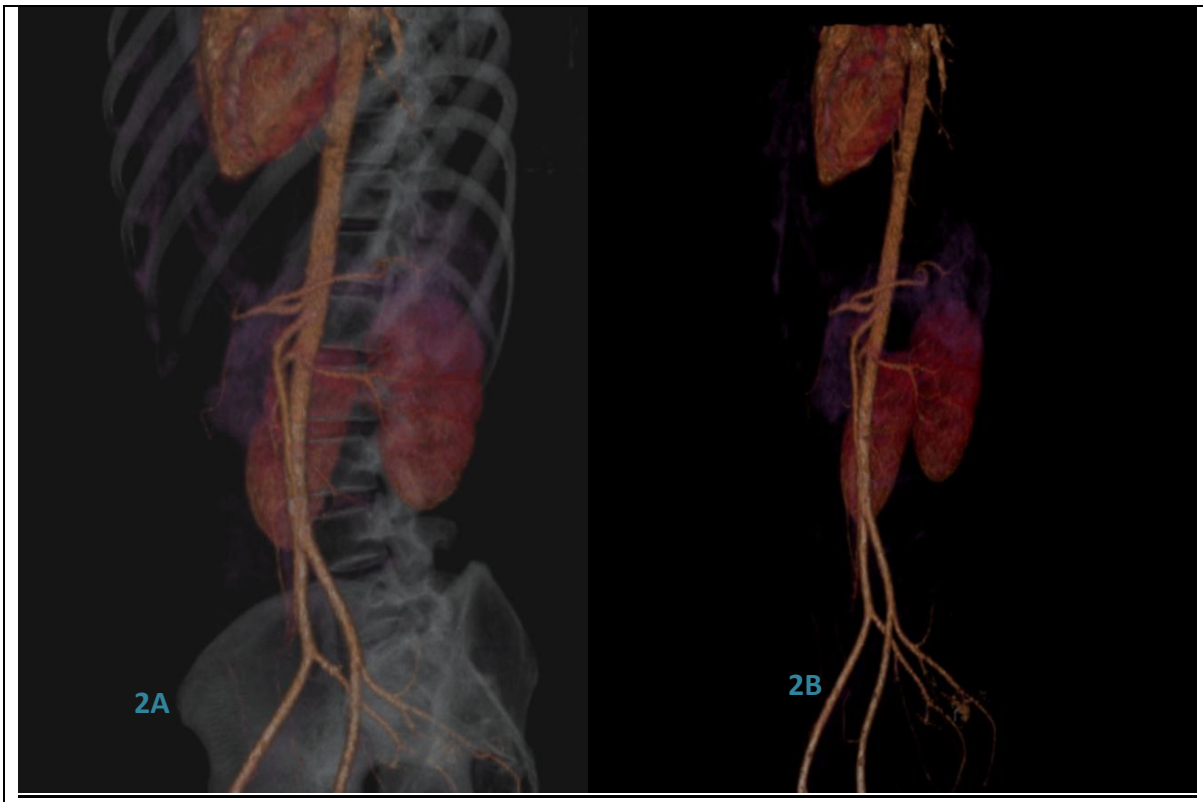

Figure 2 3D reconstruction images show the characteristic appearance of celiac trunk compression

CASE 2

50 years old male known case of hypertension with history cigarette smoking for more than 10 years presented with decreased sensation with both lower limbs with skin discoloration and ulceration. However, he had no abdominal symptoms. He primary physician referred him to our department for CT angiography of bilateral lower limbs to rule out lower limb ischemia. His CT angiography showed moderate to severe arterial insufficiency in bilateral lower limbs. However an incidental note through the lower cuts of his abdomen showed compression over celiac trunk superiorly near its origin giving a hooked appearance (figure 3) representing MALS. The patient was then referred to vascular surgery and interventional radiology department for management of bilateral lower limb findings and the general surgery department for management of MALS. Since the patient was asymptomatic for MALS, he was managed conservatively.

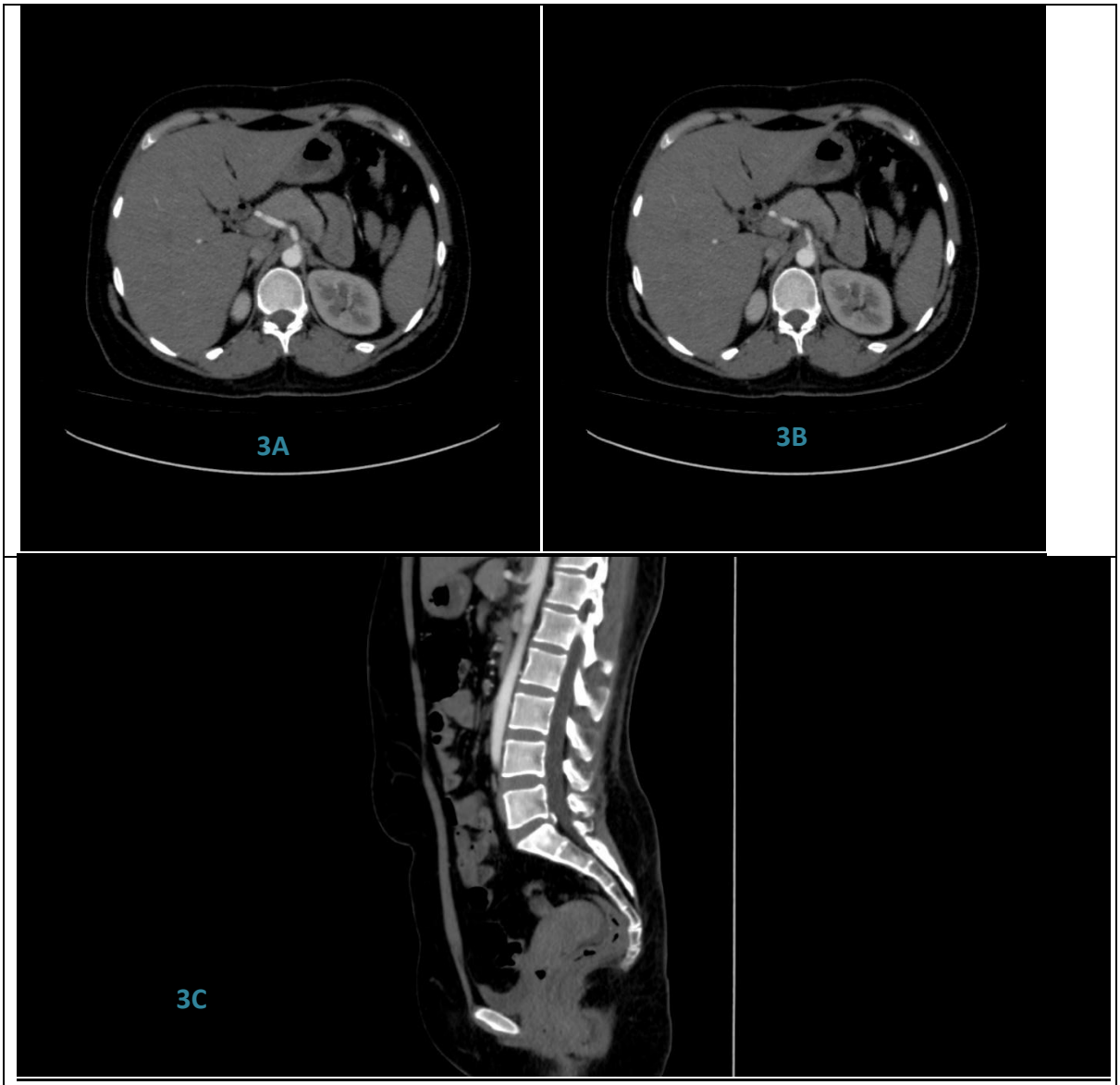

Figure 3 CT scan cuts of lower abdomen showing focal narrowing at celiac trunk origin due to indentation along its superior aspect

CASE 3

45 year old male with no known co-morbidities presented to his primary physician with complaints of post prandial epigastric pain and weight loss for more than 1 year. He had also previously presented with the same complains for which the workup which was unremarkable. His blood workup and ultrasound abdomen was normal. He was referred to gastroenterology department who advised esophagogastroduodenoscopy. Patient underwent this procedure with biopsies taken which also unremarkable and only showed non-specific gastritis for which he was given medication. However, the patient did not feel any relief in symptoms. He was then referred to the general surgery department. His clinical examination revealed an abdominal bruit and impression of MALS was made and he was advised a CT scan abdomen with contrast. His CT scan revealed a hooked appearance of celiac artery with compression over the superior aspect near its origin with post-stenotic dilatation (figure 4). His findings were consistent MALS. The patient then underwent laparoscopic surgery which was successful. He experienced complete alleviation of symptoms post procedure. The patient was discharged on 3rdpost operative day and was advised follow-up on OPD.

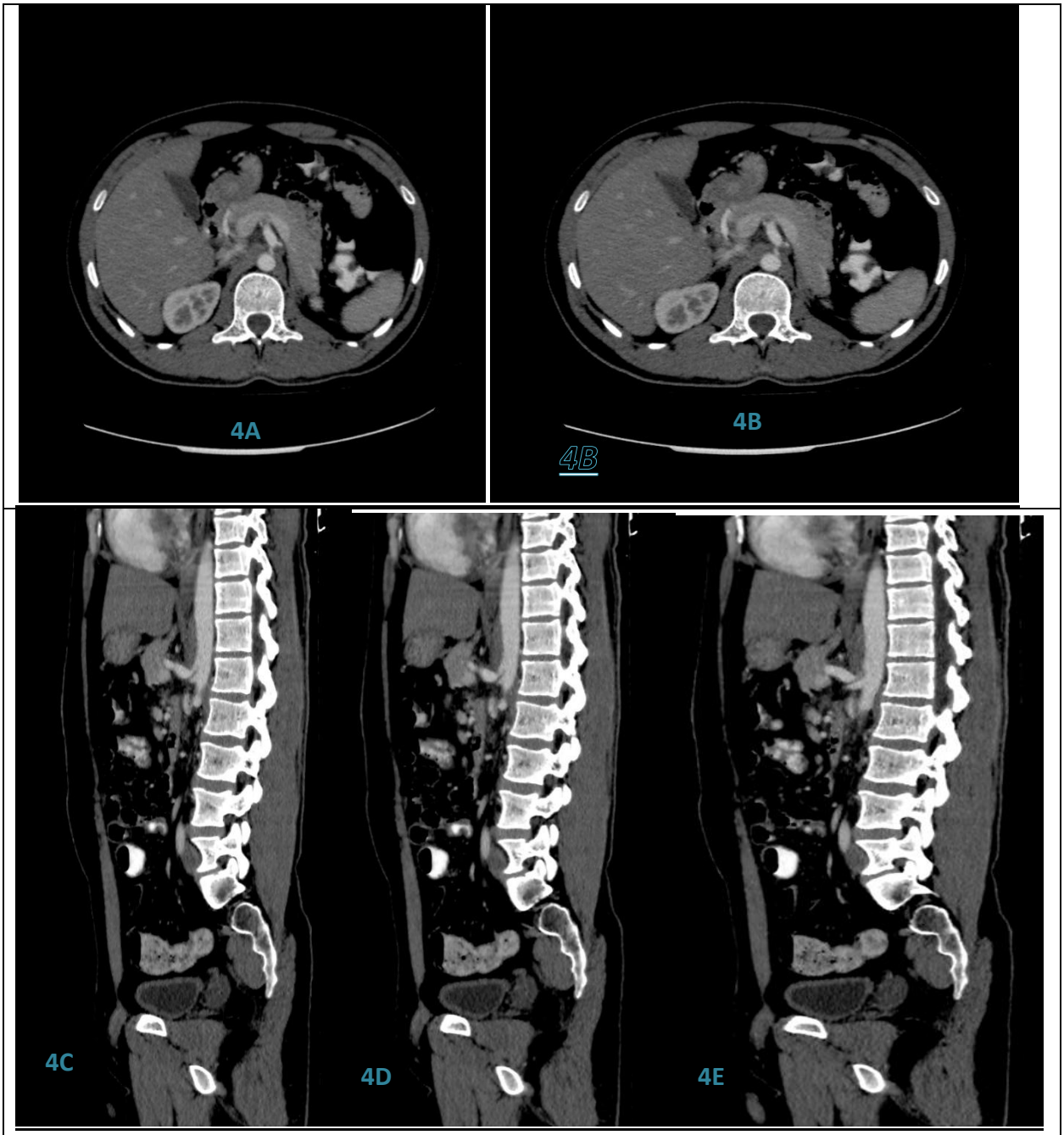

Figure 4 CT abdomen demonstrating the characteristic "hooked" appearance of celiac artery near its origin representing median arcuate ligament syndrome

DISCUSSION

As previously stated, the exact etiology of median arcuate ligament is unknown mostly owing to the fact that it is a rare condition. The most common presenting complaints include post-prandial epigastric pain, nausea and weight loss. Another less common presentation may be exercise induced abdominal pain. The patients may also be asymptomatic with MALS found as an incidental finding in such patients[4]. Similarly in two of the patients in our case series, MALS was an incidental finding. However, in one of the cases the patient presented with classic symptoms of MALS. It is more commonly reported in females between the ages of 30-50 years[1]. However, a rarity noted in our case series was that all patients were male. One of the most widely accepted theory about MALS is that due to the compression over the celiac trunk, there is an increase in demand of blood supply to foregut that may lead to ischemia which eventually presents clinically as epigastric pain and on examination may present with a bruit. Another possibility is that the pain caused by MALS has a neuropathic component and is brought on by a concomitant overstimulation and prolonged compression of the celiac ganglion. This may irritate and stimulate the sympathetic chain fibers leading to vasoconstriction and ischemia[7].

The diagnosis of MALS is one of exclusion and as such all the other causes of patient's symptoms should be excluded before considering this diagnosis. Reviewing the patient's symptoms, medical history, and linking imaging findings to the patient's symptoms as well as the patient's symptom resolution following treatment are crucial components in making the diagnosis[8].The initial modality that may aid in its diagnosis is duplex ultrasound. It exhibits an increased peak systemic velocities during expiration, which may return to normal during inspiration or when standing upright[5]. In our case series, the patient that was symptomatic had a normal ultrasound after which he was advised CT scan abdomen on which diagnosis of MALS was made.Previously, the gold standard technique for the diagnosis of MALS was conventional angiography. However, with the advancement made in the field of radiology especially with MDCT, it has become easier to diagnose it. On MDCT, it appears and an area of focal narrowing of celiac trunk giving the so called "hook" appearance especially on sagittal view[2,4].

The surgical management is considered the mainstay of treatment in symptomatic patients. It may be performed open or laparoscopically. Currently available data demonstrates that roughly 80% of individuals undergoing this operation experience alleviation of symptoms. In the majority of cases, pain relief is immediate; however, it may take up to 6 weeks to assess whether the treatment was effective because postoperative pain can resemble preoperative symptoms. In such cases adjunct angioplasty can help alleviate the symptoms[3].

CONCLUSION

MAL syndrome although a controversial diagnosis can be a cause of discomfort to the patients. It may remain silent in some patients. Patients might undergo several diagnostic tests due to the pain caused by the syndrome, it however can be treated and needs to be kept as a differential in young thin patients with no other identifiable cause of abdominal pain.

CONFLICT OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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