



Median Arcuate Ligament Syndrome: A Rare Cause to be Considered in Cases of Non-Specific Upper Abdominal Pain

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: The Median arcuate ligament syndrome (MALS) is a cause of chronic abdominal pain affecting both children and adults. MALS also known as celiac artery compression syndrome (CACS) can be symptomatic, presenting as chronic abdominal pain, nausea, vomiting, weight loss and diarrhoea.

Case Report: 49 years old male presented with complaints of recurrent abdominal pain aggravated after meals. CT angiography of abdomen revealed focal narrowing of the celiac artery at its origin with a thin filling defect, characteristics hooked appearance and linear hypodense band suggestive of MALS. Laparotomy was done and division of median arcuate ligament (MAL) and celiac plexus destruction was done.

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Discussion: MAL is a fibrous arch that unites the diaphragmatic crura on either side of aortic hiatus, with the normal anatomic location of the ligament being superior to the origin of the celiac axis. The cause of abdominal pain is thought to be due to either poor blood flow from CACS, nerve irritation or combination of both. Compression of the celiac artery may result in the blood flow coming from another abdominal vessel (the superior mesenteric artery) and going to the stomach and liver when a patient eats, known as 'steal phenomenon'. The treatment options for MALS include surgical laparoscopic or robotic division of MAL, celiac ganglion destruction and bypass surgery.

Conclusion: MALS is diagnosis of exclusion. CT angiography is an excellent modality to rule out various pathologies and diagnose MALS.

Keywords: Median arcuate ligament syndrome; celiac artery compression syndrome; upper abdominal pain; CT angiography.

1. INTRODUCTION

The Median arcuate ligament syndrome (MALS) is a cause of chronic abdominal pain affecting both children and adults. MALS was initially described by Harjola [1] in 1963 and later by Dunbar et al in 1965. MALS also known as celiac artery compression syndrome (CACS) can be symptomatic, presenting as postprandial intestinal angina due to insufficient blood supply to the gastro intestinal organs. Besides chronic abdominal pain, other symptoms include nausea, vomiting, weight loss and diarrhoea. In some patients the symptoms can be devastating and can lead to erroneous diagnosis of an eating disorder, psychiatric condition [2,3] or functional abdominal pain (like in irritable bowel syndrome abdominal migraine). The diagnosis is made based on a combination of the clinical symptoms and radiology imaging and intraoperative anatomy [4]. Some degree of radiographic compression is observed in 10% - 24 % of asymptomatic patients. We report a case of MALS diagnosed on abdominal CT angiography and successfully treated surgically by the release of median arcuate ligament and destruction of celiac nerve plexus.

2. CASE REPORT

49 years old male presented with complaints of recurrent abdominal pain aggravated after meals. He belonged to labour class and was unable to do heavy work as it increased his pain. He had been taking antacids, proton pump inhibitors and analgesics for last two years. He had weight loss of about 6 kgs in this duration. His present weight was 45 kgs. He had vomiting off & on and sometimes diarrhoea. The previous treating physicians suspected chronic gastritis and abdominal tuberculosis but upper gastrointestinal endoscopy was normal. Routine blood counts and ESR was normal. TB gamma interferon and covid RTPCR were negative. Our area is

endemic for sickle cell anaemia which can lead to chronic abdominal pain but Hb electrophoresis was normal adult haemoglobin pattern. USG report showed gaseous distension of bowels. Plain CT scan showed an enlarged lymph node in right iliac fossa. Patient had occasional intake of alcohol. Serum amylase and lipase were normal ruling out possibility of chronic pancreatitis.

CT angiography of abdomen revealed focal narrowing of the celiac artery at its origin with a thin filling defect, characteristic hooked appearance and linear hypodense band suggestive of MALS. (Figs. 1, 2A,2B). Splenic, hepatic and left gastric arteries showed normal course and calibre. (Fig. 3A). There were no collateral vessels, but accessory left renal artery was noted (Fig. 3B).

The patient was thoroughly evaluated by team of physician, surgeons and psychiatrist and decision for laparotomy was taken. Abdomen was opened by midline incision. Celiac trunk identified by tracing common hepatic and left gastric artery to the root. Root of celiac trunk was compressed by diaphragmatic fibres and narrow just after origin from aorta (Fig. 4A). All the fibres over celiac trunk divided carefully and celiac trunk skeletonized. Narrowing of celiac trunk disappeared after complete release of fibres (Fig. 4B). All the nerve plexus around celiac trunk carefully excised. Intra and post-operative period remained uneventful. Patient has completed 3 months follow-up with complete relief in symptoms, increase in appetite and weight gain of 3 kgs.

3. DISCUSSION

The median arcuate ligament (MAL) is a fibrous arch that unites the diaphragmatic crura on either side of aortic hiatus, with the normal anatomic

location of the ligament being superior to the origin of the celiac axis [5]. Lipshutz [6], an anatomist, first described compression of the celiac axis by extraluminal structures. Selme Patrella et al [7] demonstrated the relationship of celiac trunk with MAL on a cadaveric study. The distance between the origin of the celiac trunk and the ligament is approximately 0.94 cm, with a minimum distance of 2.32 cm. MALS results from an aberrant position of the origin of the celiac artery, which is normally positioned at T12 or L1 or abnormal location of MAL. In a study of 75 autopsies, the MAL crossed celiac artery or origin entirely (33%) or partially (48%) resulting in significant CACS [8].

Position of the celiac trunk varies with respiration. During expiration, the aorta and

celiac artery move cephalad and hence the anomalous positioned ligament produces characteristic symptoms due to compression. The reverse happens during inspiration, where in the celiac trunk descends downwards into the abdominal cavity resulting in more vertical orientation, relieving the compression effect thereby alleviating the symptoms. Doppler USG is a non-invasive technique to measure the rate of blood flow through the celiac artery on inspiration and expiration and may show a variation in the peak systolic velocity of greater than 200cm/s and end diastolic velocity greater than 55 cm/sec [9,10].

The cause of abdominal pain is thought to be due to either poor blood flow from CACS, nerve irritation or combination of both. Compression of

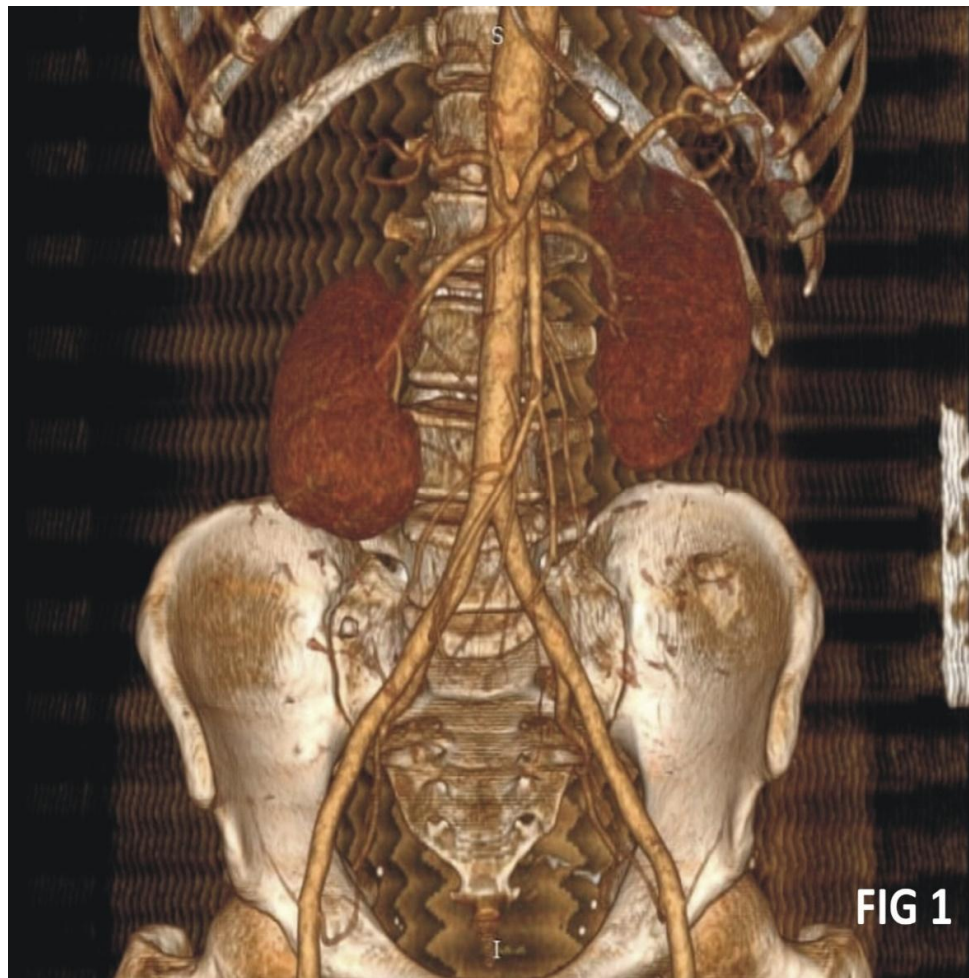


Fig. 1. Narrowing of celiac artery at its origin with linear hypodense band

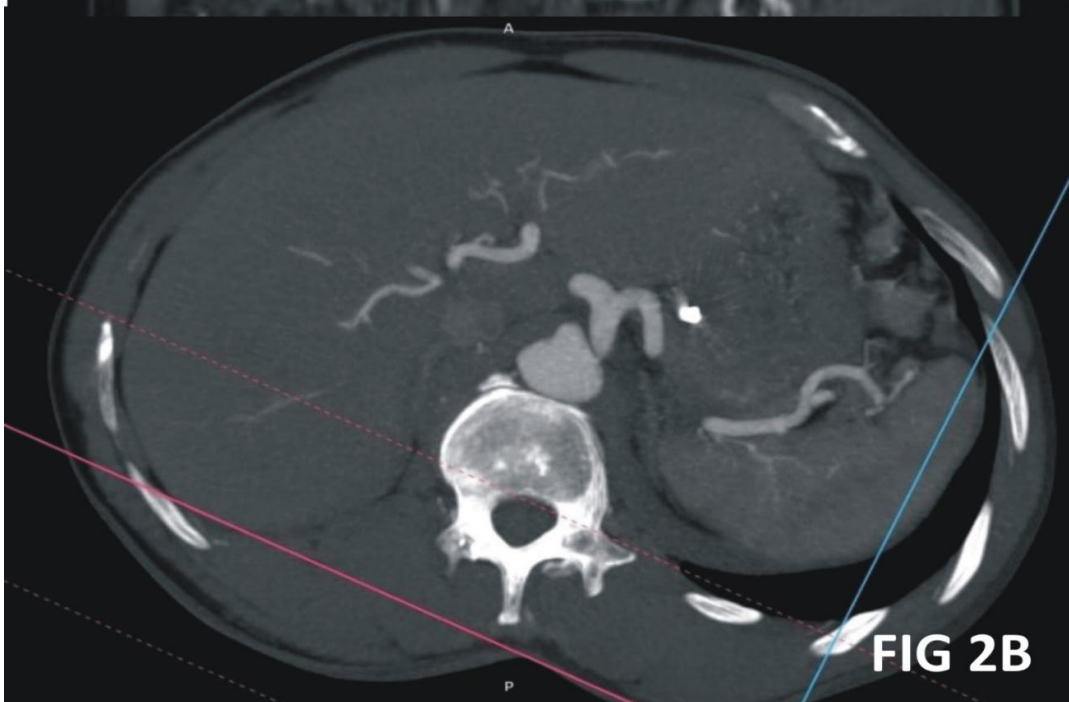
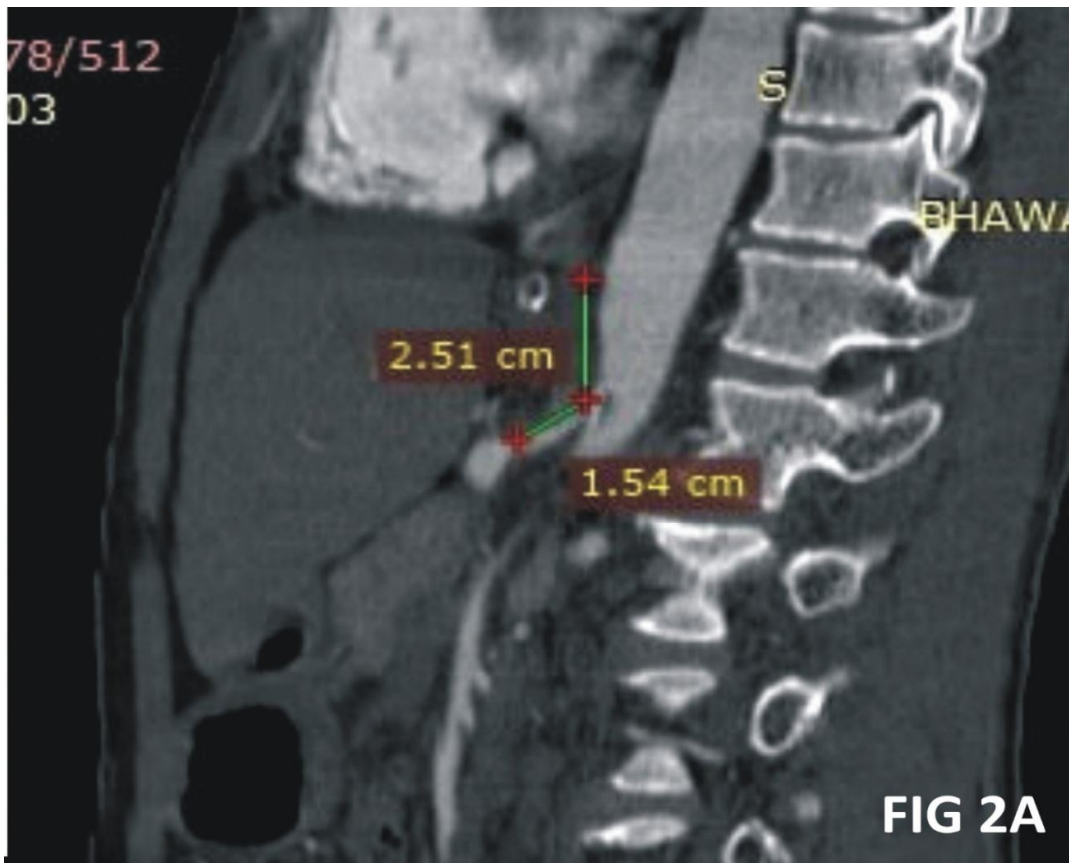
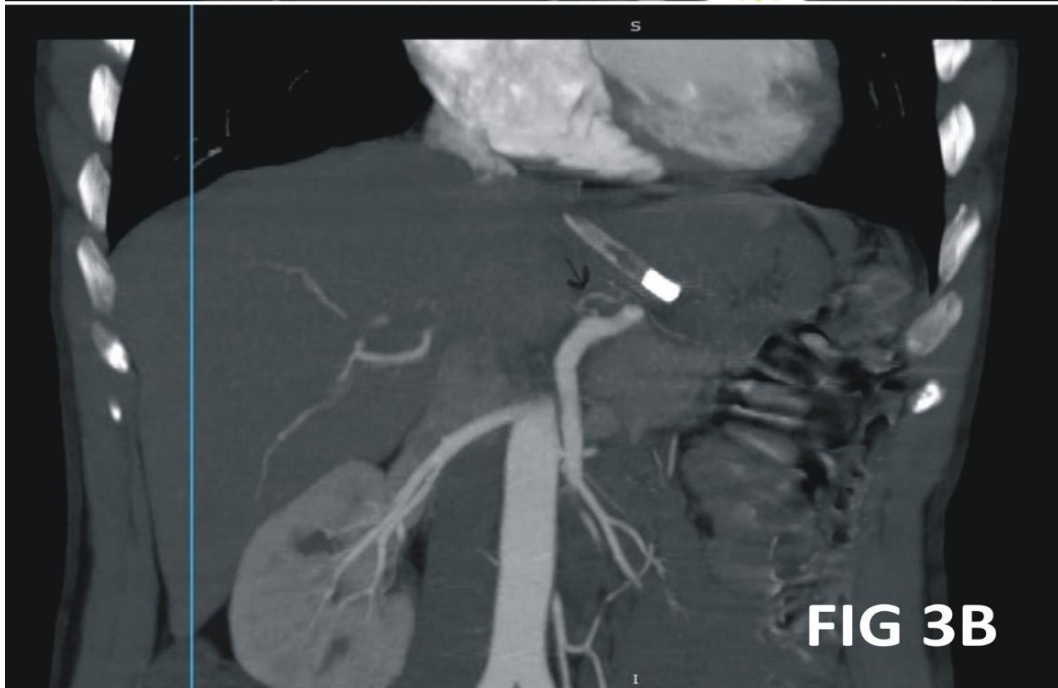
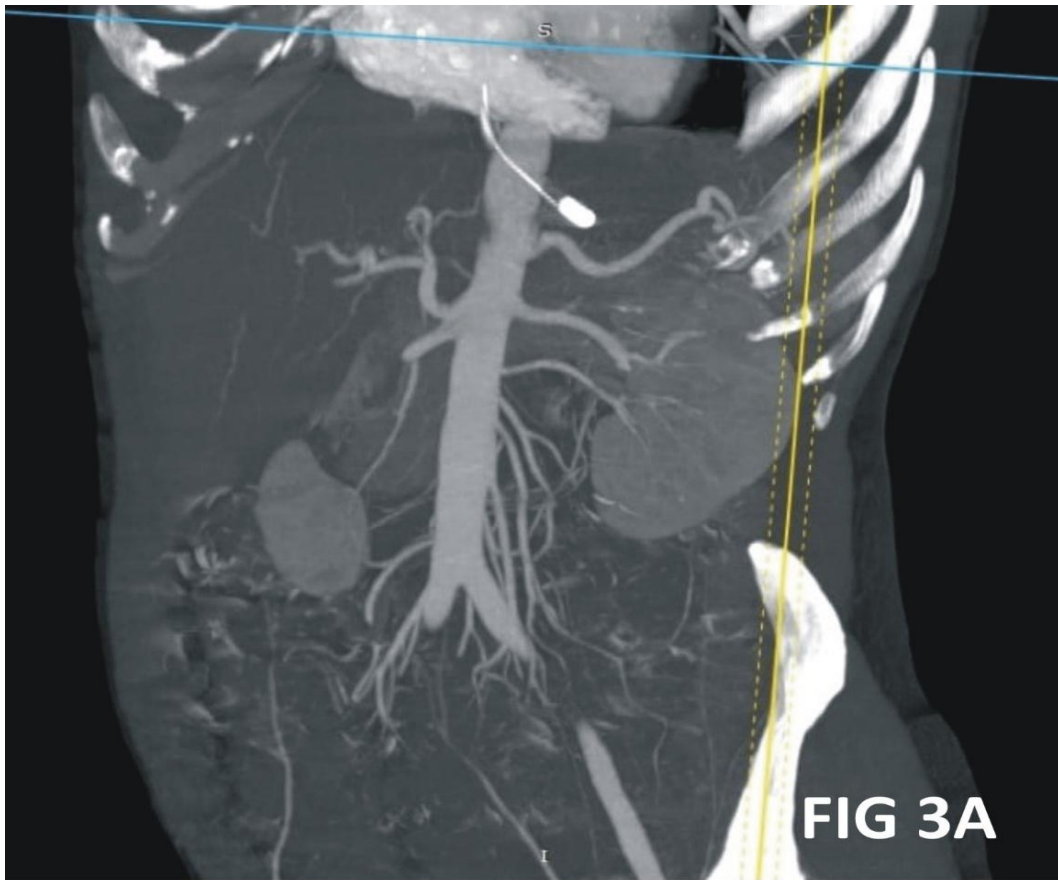


Fig. 2A. Distance of stenotic segment and distance from diaphragmatic crus
2B. Hooked appearance of celiac artery



**Fig. 3A. Normal calibre of gastric, hepatic and splenic artery
3B. No collateral vessels and accessory left renal artery**

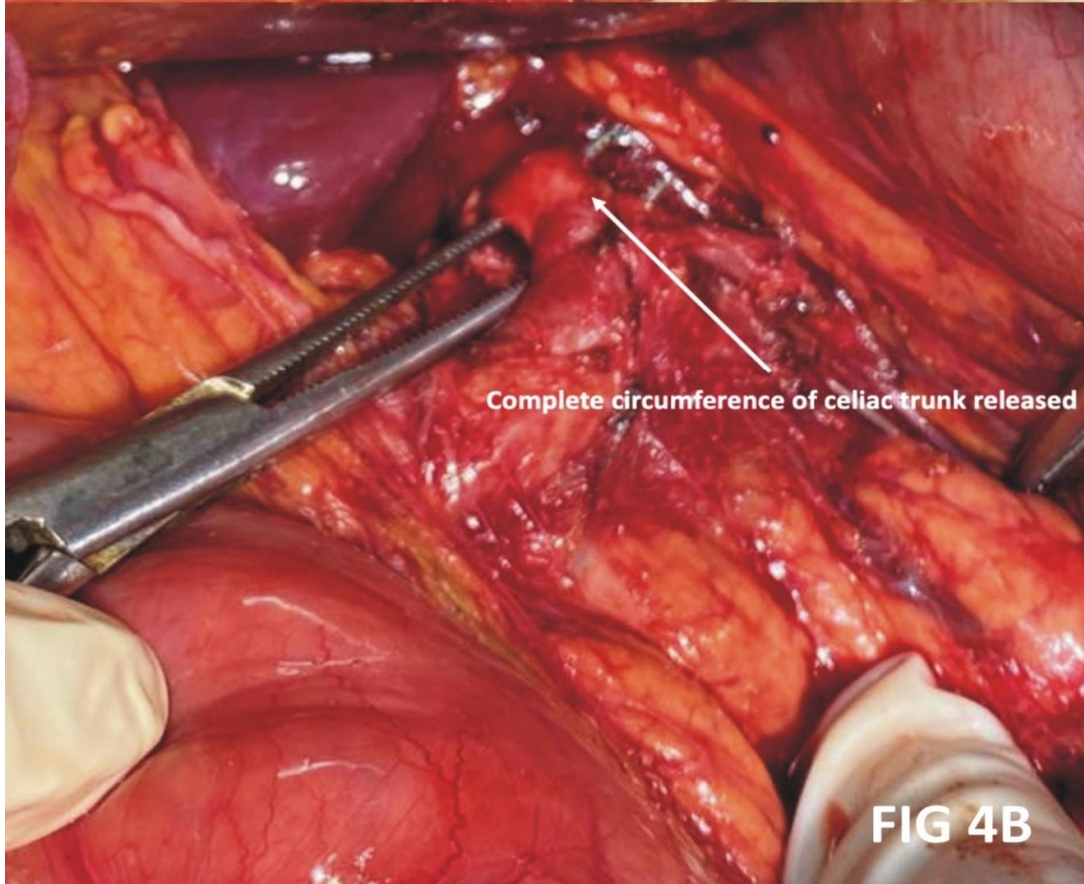


Fig. 4A. Celiac trunk origin
4B. complete circumference of celiac trunk released

the celiac artery may result in the blood flow coming from another abdominal vessel (the superior mesenteric artery) and going to the stomach and liver when a patient eats, known as 'steal phenomenon'. Another theory is that the nerves in the area of the celiac artery (the periaortic ganglia and celiac nerve plexus) get over stimulated leading to spasm (vasoconstriction) of the arteries and resultant symptoms. The compression of the nerves may cause interference of the brain-stomach, neuroenteric pain pathways resulting in hypersensitivity and pain.

Various modalities can aid in the diagnosis of MALS such as Doppler ultrasonography, spiral CT angiography, selective catheter angiography and magnetic resonance angiography. CT angiography reveals a characteristic focal narrowing and hooked appearance in the proximal celiac axis in MALS. and this appearance distinguishes this condition from atherosclerosis. Post stenotic dilatation and collateral vascular vessels can be seen on CT angiography.

Selective catheter angiography is the gold standard diagnostic method but is more invasive. Hence CT angiography has gained superiority over catheter angiography. Superior mesenteric artery, renal arteries and collateral vessels can also be visualized on CT angiography.

The treatment options for MALS include surgical laparoscopic or robotic division of MAL, celiac ganglion destruction and bypass surgery [11].

Preoperatively all patients should be evaluated by a multidisciplinary team consisting of general and vascular surgery, psychiatrist, and pain service. Average success rate of surgical intervention is reported to be 60- 80% [12,13] postoperative complications tend to be minor and self-limited pancreatitis. The major post-operative problem is either persistence or return of the abdominal pain due to anxiety or depression. Our patient is completely relieved of pain till date with weight gain but has been counselled thoroughly for long term follow-up.

4. CONCLUSION

MALS is diagnosis of exclusion. Chronic upper abdominal pain can occur because of many common aetiologies like gastritis, gastric ulcer, pancreatitis, cholecystitis, hepatitis, splenomegaly, portal vein thrombosis, mesenteric ischemia etc. CT angiography is an

excellent modality to rule out various pathologies and diagnose MALS. Psychiatric evaluation of the patient should be emphasised. Surgical treatment with division of the median arcuate ligament and excision of the celiac plexus gave us satisfactory result.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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