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Laparoscopic management of celiac artery compression syndrome: A case report

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ABSTRACT

INTRODUCTION: Celiac artery compression syndrome is a rare disorder found mainly in young females. It many a times goes undiagnosed as the symptoms are non-specific and varied.

PRESENTATION OF THE CASE: We present a case of celiac artery compression syndrome in a young female where laparoscopic median arcuate ligament release with celiac ganglionectomy was effective in relieving the symptoms.

DISCUSSION: Commonly they may present with abdominal pain, vomiting and diarrhea but there is absence of significant clinical signs. High level of suspicion and the right imaging techniques like the lateral aortogram, helps us to clinch the diagnosis.

CONCLUSION: Laparoscopic release of celiac artery compression is a safe and effective method to treat this uncommon disease.

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1. Introduction

Celiac Artery Compression Syndrome¹ (CACS) is a rare disorder of the celiac artery where there is an eccentric compression of the celiac trunk. This disease, as the name suggests is a result of the compression of the celiac artery at its origin by the diaphragmatic fibers, or the celiac plexus fibers and ganglia. This was first described by Harjola in 1963 [1].

Here we discuss the case of a young female who was diagnosed as CACS at a tertiary care hospital and subsequently operated laparoscopically for median arcuate ligament² (MAL) release with celiac ganglionectomy which resulted in a satisfactory outcome.

This case has been reported in line with the SCARE criteria 2018 [2].

2. Case presentation

A 21-year-old female who weighed 41 kg and had a BMI of 17.7 kg/m² presented in the emergency room with the history of epigastric post-prandial pain and vomiting for 2 years. The pain was cramp-like and would start 15–20 min after the meal. The pain

would also radiate to the lower chest. It would reduce after a bout of vomiting. The symptoms were more after a solid meal, she was relatively comfortable consuming liquid meals. There were no other co-morbidities. Over the period of 2 years she has been treated with PPI, anti-spasmodic and antibiotics but did not get much relief. The physical examination was unremarkable except that she was thin built and short. Abdominal examination did not reveal any findings.

Hemogram, liver function test, serum amylase and lipase were normal. X-rays of the abdomen and chest were unremarkable. Patient was started on antibiotics, PPIs, and analgesics but the post prandial pain continued. The abdominal ultrasound did not reveal any liver, biliary, pancreatic, or renal pathology. An upper endoscopy was performed on day 3 and found normal. The physician, to rule out an ischemic cause, on day 4 referred her to the interventional radiologist to perform an abdominal vascular DSA. The Findings of the aortogram images in the sagittal plane revealed narrowing and hook shape of the celiac main trunk [Fig. 1]. This narrowing was accentuated in end expiratory phase [Fig. 2]. There was post-stenotic dilatation secondary to significant narrowing.

A diagnosis of 'Celiac Artery Compression Syndrome' was reached, and the patient was referred to the surgeon on day 5. On the 7th day after admission the patient was posted for a laparoscopic division of the median arcuate ligament and celiac plexus fibers to achieve a complete release of the compression by the author who is trained in upper GI and laparoscopic surgery.

The patient was placed supine with a leg-split and in a steep anti-Trendelenburg position. Five port were placed in the upper abdomen [Fig. 3]. The dissection was started by opening the gastro-hepatic omentum. The phreno-esophageal ligament was divided and the right and left diaphragmatic crura were exposed. Retro-

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¹ CACS – Celiac artery compression syndrome.

² MAL – Median arcuate ligament.



Fig. 1. CT scan showing hook or J shape of the C (indicated by the white arrow).

gastric dissection was performed, and the stomach was looped and retracted anteriorly to expose the diaphragmatic fibers overlying the aorta. Care was taken to avoid mobilizing the thoracic esophagus. Both the anterior and posterior vagi were identified and preserved. The left gastric artery, splenic artery, and the common hepatic artery were isolated, and their confluence was identified. All the fibro-fatty tissue including the lymph nodes at station 8A and 9; superior to the common hepatic artery and celiac artery were removed to give clearer access to the median arcuate ligament. The diaphragmatic fibers then were carefully divided from the hiatus downwards exposing the aorta [Fig. 4A]. This was continued caudally till all the diaphragmatic muscle fibers overlying the aorta were divided to expose the celiac artery. The celiac plexus and the ganglia were also found tightly arching over the celiac artery hence were divided individually till the celiac artery was

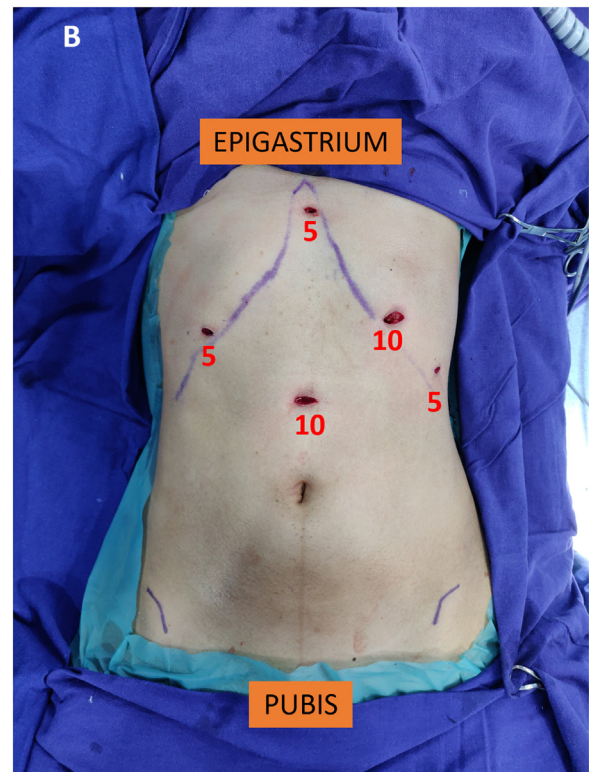


Fig. 3. Laparoscopic port positions.

completely free of all the muscle, nerves and fibro-fatty tissue covering it [Figs. 4B, 5]. A Jackson Pratt drain was inserted to monitor the post-operative bleeding if any. Sequential compression device was used intraoperatively, and postoperatively she was administered injection Enoxaparin 40 mg subcutaneously once daily from the second day till the day of discharge, for DVT prophylaxis.

The surgery time was 180 min and there was minimal blood loss. Postoperatively patient was sent back to her room, mobilized within 4 h and feeding was resumed 6 h after surgery. There was no significant post-operative collection in the drain and hence was removed after 48 h. She was discharged on the 4th post-operative day. The patient could not assess her post-meal pain for a week due

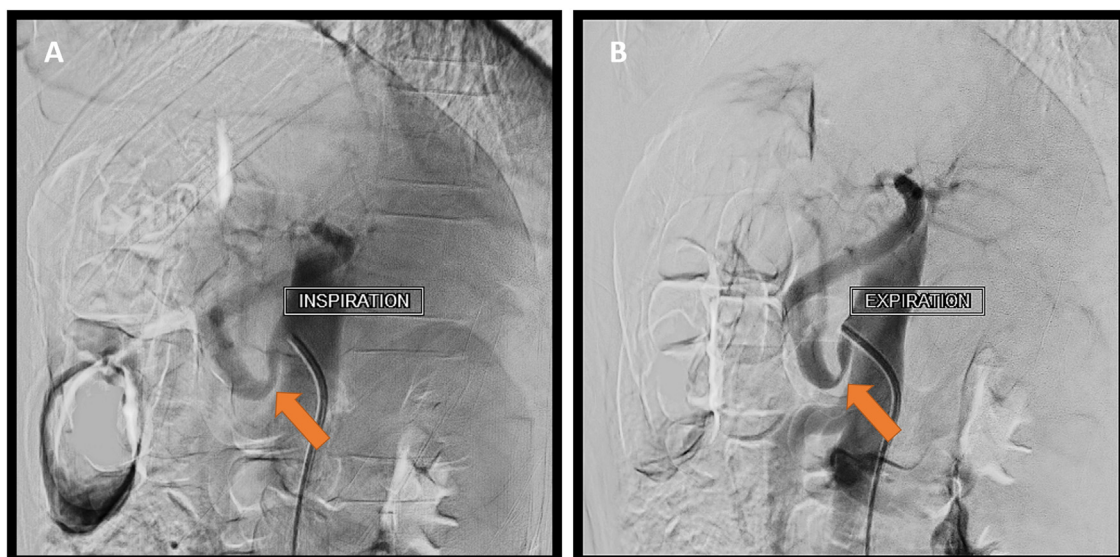


Fig. 2. Lateral inspiratory and expiratory angiogram of celiac trunk with orange arrows pointing to the hook deformity.

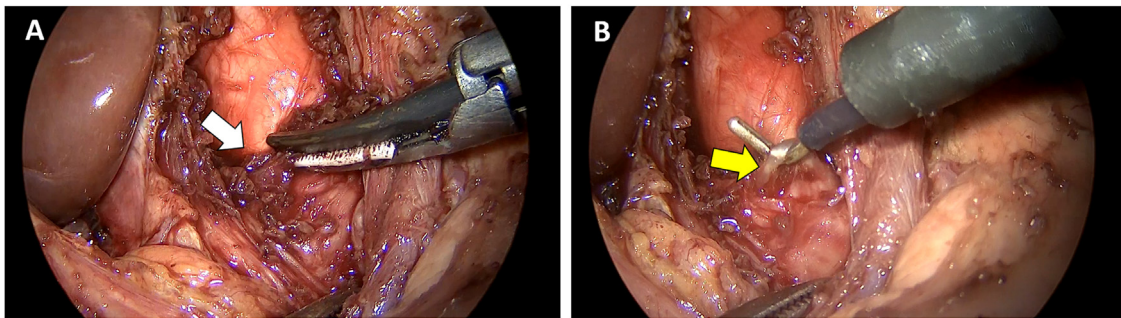


Fig. 4. A - Division of Median arcuate ligament fibres (white arrow), B - Division of Celiac plexus neural tissue (yellow arrow).

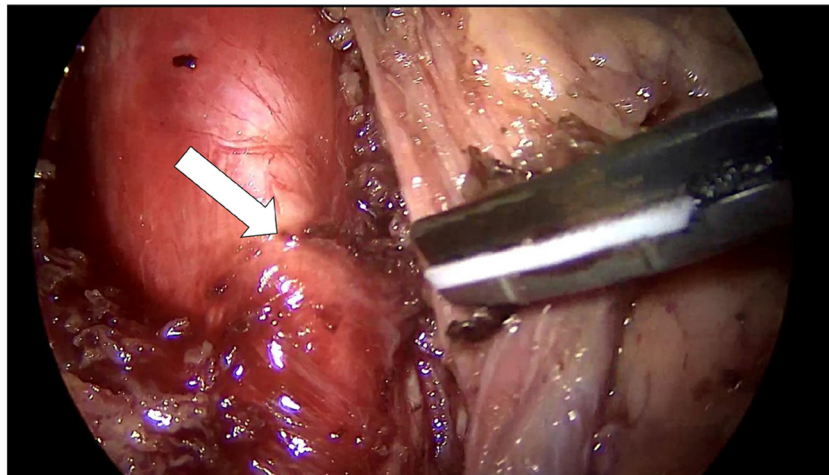


Fig. 5. Completed division of all neuro-muscular and fibro-fatty tissue overlying the Celiac Artery.

to the pain at the port sites. In the second week after surgery she appreciated the fact that she had no post-prandial epigastric pain. The patient was followed up for 11 months postoperatively and has been symptom-free till then. Patient had gained 6 kg of weight and now weighed 47 kg.

3. Discussion

The incidence of 'Celiac artery compression syndrome' is higher in females (81%) and the commonest age group being between 20–40 year [3]. In literature cases have been reported between the age of 22 months to 71 years [4,5].

This compression can possibly result from a low insertion of the diaphragmatic fibers, a high origin of celiac artery or compression due to celiac plexus fibers and ganglion [6,7]. Lipshutz in 1917, after dissecting 24 cadavers published the observation that, celiac artery was frequently found partially covered by the fibers of the diaphragm [8]. The pathophysiology of CACS is poorly understood, and its symptoms have been attributed to both ischemic as well as neurogenic causes. Also, there is conflicting evidence to support both mechanisms. Ischemia from 'Steal syndrome' is described as a mechanism where the superior and inferior mesenteric artery blood supply is diverted to the celiac artery distribution giving rise to ischemic pain [9]. Alternatively, compression of the celiac plexus by the median arcuate ligament may also produce abdominal pain, and may delay gastric emptying leading to the symptoms of CACS [7,9]. The neurogenic theory of pain is also supported by the fact that percutaneous celiac plexus block gives relief in some patients with chronic abdominal pain [10,11]. Unusually, acute CACS has

been reported after kyphosis correction surgery and pancreaticoduodenectomy operations [12,13].

Jimenez in a meta-analysis of 400 cases listed the common symptoms of CACS that were: abdominal pain (80%), weight loss (48%), abdominal bruit (35%), nausea (9.7%), and diarrhea (7.5%) [10].

Radiology plays an important role in the diagnosis of median arcuate ligament compression. On color doppler study, peak systolic velocity of ≥ 200 cm/sec in the celiac artery, and a $\geq 70\%$ narrowing would confirm the diagnosis of CACS [4,14,15]. CT or catheter angiography with images taken in lateral projection would reveal a narrowing in the proximal celiac artery forming a 'J' or hook. The compression is accentuated during deep expiration [10,16,17]. Post-stenotic dilatation and presence of collaterals in the gastro-duodenal arterial circulation when present would suggest long standing narrowing. Smooth aortic wall rules out atherosclerotic or inflammatory arterial disease. If on CT scan, the thickness of the median arcuate ligament fibers is more than 4 mm then this would also be suggestive of MAL compression [15].

Though CACS was conventionally managed by vascular surgeons, general surgeons trained in minimally invasive surgery successfully started treating CACS [18]. Tulloch compared outcomes of laparoscopic versus open surgery for median arcuate ligament division and ganglionectomy, the laparoscopic group resumed feeding faster than the open group which was 1.0 vs 2.8 days respectively ($P \leq .05$), and the hospital stay was significantly reduced in the laparoscopic group as compared to the open group (2.3 vs 7.0 days respectively) [18]. Laparoscopic technique

though advantageous does have a higher rate of complications like vascular injuries and bleed requiring conversion to open [18]. Robot-assisted laparoscopic approach for CACS has also been used successfully and was reported for the first time by Jaik et al. in 2007 [19]. Though laparoscopic surgeon can undertake the procedure successfully, injury to the aorta or the celiac artery are serious complications and one should preferably have a vascular surgeon standby in anticipation [20]. Other complications reported in the laparoscopic approach is injury to the visceral arteries, pneumothorax, and phrenic artery laceration [10].

There is literature supporting additional techniques like celiac revascularization using bypass grafts, or angioplasty and stenting which may be beneficial in reducing the long-term recurrence rate [10,21]. These additional techniques may have a place in case there is a failure to completely divide the median arcuate ligament surgically or if there is a recurrent disease [21].

4. Conclusion

CACS is considered as a rare disorder, it is often misdiagnosed and inadequately treated. Clinically, the non-specific symptoms make it challenging to diagnose and suspicion arises mainly through exclusion. Imaging modalities like the angiogram and color doppler can accurately diagnose and quantify the problem of CACS. Minimally invasive methods like laparoscopic and robotic surgery should be the preferred choice. The surgery if done adequately will give good long-lasting relief. The surgery needs to be done with great care as there is a real danger of vascular injury and significant morbidity. A delayed reappearance of symptoms may be due to inadequate release or other vascular causes, these can be treated with angiography and stenting or bypass graft.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Approval of the institutional head obtained.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

1. Sharad Sharma – Treating surgeon, responsible for concept, data collection and drafting the case report.
2. Vimal Someshwar – Radiologist, responsible for diagnosis, reporting, data collection and drafting certain relevant parts of the case report.
3. Farah Ingale – Treating Physician, responsible for revising the draft.

Registration of research studies

N/A.

Guarantor

Sharad Sharma.

Patient perspective

The patient reports good relief from the symptoms on last follow up 11 months since the operation.

Provenance and peer review

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