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Case Report

Median arcuate ligament syndrome presenting with unintentional weight loss and epigastric pain

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ABSTRACT

Median arcuate ligament syndrome is a rare condition characterized by extrinsic compression of celiac artery and celiac plexus by the median arcuate ligament. Patients typically present with chronic postprandial pain, nausea and occasionally, vomiting and weight loss. Treatment of this syndrome is laparoscopic or open surgical release of median arcuate ligament and gangliectomy. We report a rare case of median arcuate ligament syndrome in a patient who presented with abdominal pain and nausea. The patient was evaluated, investigated and planned for surgical intervention but the patient refused for treatment.

Keywords: Median arcuate ligament syndrome, Epigastric pain, Hooked appearance, Post prandial steal phenomenon

INTRODUCTION

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crurae on either side of the aortic hiatus. The ligament is usually present superior to the origin of the celiac axis. In some people, the ligament inserts low and thus crosses the proximal portion of the celiac axis, causing compression of the celiac artery and celiac ganglion. Symptoms are chronic recurrent postprandial pain with associated nausea and vomiting.[1-3]

CASE REPORT

A 50 year old gentleman was evaluated for 4 month history of complaint of epigastric pain that started 2 hours after taking meal and persisted for the next couple of hours. The pain radiated to his back, sometimes associated with nausea and non-bilious vomiting. Patient also complained of significant weight loss in last 4 months. He had no significant past medical or surgical history, with no history of any addiction.

General physical examination was essentially normal. His abdomen was soft with mild tenderness around the epigastric region; rest of the abdomen was normal. His hematological and biochemical blood parameters were in the normal range. Ultrasonography of the abdomen did not suggest of any abnormality. We advised the patient to maintain a diet record and note his symptoms and frequency of pain after each meal for 15 days. It was observed that the patient experienced same type of postprandial abdominal pain; 3 to 4 times in a day. Contrast enhanced computed tomography (CECT) abdomen with CT angiography was carried out at our Radiology department. CT angiography demonstrated characteristic focal narrowing in the proximal celiac axis, with characteristic hooked appearance due to focal narrowing (Figure 1). Coronal oblique angiogram of same patient reveals prominent collateral vessels between gastroduodenal artery and superior mesenteric artery (Figure 2). All these findings were suggestive of median arcuate ligament syndrome.

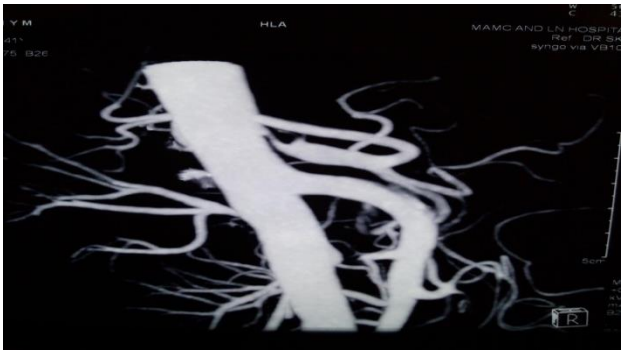

Figure 1: Maximum intensity projection image of abdominal aorta and its branches. Sagittal image showing stenosis of proximal part of coeliac trunk with smooth superior indentation giving characteristic hooked appearance suggestive of external compression by median arcuate ligament.

Figure 2: Maximum intensity projection image of abdominal aorta and its branches. Coronal image showing collaterals connecting gastroduodenal artery with superior mesenteric artery.

We discussed the diagnosis with the patient and presented the surgical option to him, but he refused further treatment.

DISCUSSION

The first case describing celiac artery compression was reported by Lipshutz in an autopsy study in 1917. Harjora, in 1967, first described a clinical case with postprandial pain as a presenting complaint.[1-3] Incidence of celiac artery compression by arcuate ligament is 10-24% in the healthy population. This syndrome typically occurs in the young (20-40 years), thin population and in the females (71%).[4]

Although, the triad of postprandial epigastric pain, unintentional weight loss and/or abdominal bruit is a classical manifestation of celiac artery compression syndrome; the actual clinical presentation of this syndrome is variable and often diagnosed through exclusion.

Pain in median arcuate ligament syndrome is explained by mesenteric ischemia through steal phenomenon, namely postprandial steal via collaterals from superior mesenteric artery. Another theory suggests pain is caused by chronic compression of celiac plexus with subsequent splanchnic vasoconstriction and ischemia.[5]

Diagnosis remains a challenge although CT angiography is considered to be the gold standard. During expiratory phase isolated compression of the celiac axis may be clinically insignificant. Up to 13-50% of healthy population may show the angiographic feature of compression to a variable degree usually during expiration. Vast majority of patients may show only incidental finding without any symptom therefore imaging finding must be correlated with the patient's clinical history.[6,7]

Various surgical modalities have been described to treat median arcuate ligament syndrome. These are the decompression and surgical division of median arcuate ligament, celiac plexus block and resection of stenotic artery. Surgery can be performed minimally invasive (laparascopic or robotic) or traditional open laparotomy approach.

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