An unusual cause of epigastric pain and diaphoresis

Vivian Georgopoulou, Evgenia Gouridou, Athina Pyrpasopoulou, Andreana Kozanidou, Christodoulos Papadopoulos, Stergios Tzikas, Maria Sidiropoulou

1. Introduction

The median arcuate ligament, or celiac artery compression syndrome is a rare syndrome, caused by extrinsic compression of the celiac trunk by the median arcuate ligament. Its symptomatology mainly comprises of visceral angina. Differential diagnosis includes cardiovascular angina, other vascular events and causes of vagotonia. Recognition of the syndrome requires clinical suspicion and appropriate radiology and diligent reviewing of the images, as it is basically a radiological diagnosis.

2. Case presentation

A 64-years-old male patient presented to the Outpatients Clinic with intermittent worsening epigastric pain of 10-hours duration. The patient had driven for 6 h straight; due to development of symptomatology he had taken a 500mg tablet of salicylic acid. Prior to presentation the patient had had a small meal which exacerbated his epigastric discomfort. The patient's history was unremarkable but for hypercholesterolemia (LDL 150 mg/dl) and chronic recurrent prostatitis. He mentioned normal bowel movement prior to presentation at the hospital.

At presentation, vital signs were normal (BP 130/85mmHg, 75 pulses/min, sO2 97% on ambient air). Shortly after entering the consultation room, and while in the supine position, the patient developed diaphoresis and nausea, during which BP significantly dropped to 85/60 mmHg. Electrocardiogram was remarkable only for an incomplete right bundle branch block. Arterial blood gas sampling did not reveal any hypocapnea or hypoxemia. Laboratory tests were ordered; hemoglobin levels were normal (15,2 g/dL), D-Dimers were within normal limits (324 ng/ml) and high sensitivity troponin levels, tested three times within 8 h were negative (0.5, 0.9 and 3.6 pg/ml respectively, nv < 15 pg/ml).

An emergency computed tomography scan of the thorax and abdomen was requested, which revealed dilatation of the stomach (gastroparesis) (Figure 1A). Careful reviewing of images taken during the respiratory cycle showed compression (narrowing) and post stenotic dilatation of the celiac trunk. The phenomenon was exacerbated during exhalation (Figure 1C) and was partly relieved on inspiration (Figure 1B). The CT scan was not suggestive of any other pathology of the gastrointestinal tract or the biliary tree/pancreas. The diagnosis of median arcuate ligament or celiac artery compression syndrome was made. A posteriori the patient mentioned experiencing episodes of vagotonia since childhood. Symptomatology was significantly relieved after administration of intravenous metoclopramide.

Consent was gathered from the patient investigated in this study, as well as consent regarding the use of the tomography images presented in this report.

* Corresponding author.
E-mail address: a.pyrpasopoulou@doctors.org.uk (A. Pyrpasopoulou).
Celiac artery compression syndrome is a rare cause of intestinal angina, more commonly encountered in middle-aged women [2, 3]. Interestingly, visceral pain represents the main feature of the syndrome, despite the fact that compression of the celiac artery is exacerbated during exhalation. A neurologic contribution to the disorder cannot be excluded. Symptomatic performance in experimental animals has been shown to increase blood flow in the celiac artery, and celiac ganglion fibers, rather than the diaphragm, have been reported to compress the celiac artery in isolated vascular preparations, though symptomatology regressed without removal of the ganglion respectively [4]. Diagnosis is confirmed by compression of the proximal celiac trunk by the median arcuate ligament syndrome, due to the far more complex pathophysiology than mere symptom relief [10]. Significant symptomatology prior to surgery appears to be associated with better response to treatment [11]. Treatment options include open release of median arcuate ligament, laparoscopic release of median arcuate ligament, robot-assisted release of median arcuate ligament and open vascular treatment (decompression of the celiac artery or revascularization) [12]. Endovascular repair and stenting are options in symptomatic patients, though symptomatology may persist postprandially. In the presence of persistent symptomatology, treatment usually involves surgical release of the median arcuate ligament [8, 9] with minimal mortality, but variable outcome in terms of symptom relief [10]. Significant symptomatology prior to surgery appears to be associated with better response to treatment [11]. Treatment options include open release of median arcuate ligament, laparoscopic release of median arcuate ligament, robot-assisted release of median arcuate ligament and open vascular treatment (decompression of the celiac artery or revascularization) [12]. Endovascular repair and stenting have limited success in the management of the median arcuate ligament syndrome, due to the far more complex pathophysiology than mere atherosclerotic mesenteric occlusive disease [13]. Our patient was given advisory consultation. His symptoms regressed promptly. This episode was the most intense he had experienced and developed after severe strain. He is currently being followed-up to determine whether step-up surgical management will be needed.

Declarations

Author contribution statement

V. Georgopoulou: Conceived and designed the experiments; Performed the experiments; Analyzed and interpreted the data; Contributed reagents, materials, analysis tools or data; Wrote the paper.
A. Pyrpasopoulou, E. Gouriou and A. Kozanidou: Conceived and designed the experiments; Analyzed and interpreted the data; Contributed reagents, materials, analysis tools or data; Wrote the paper.
C. Papadopoulos and S. Tzikas: Conceived and designed the experiments; Analyzed and interpreted the data; Wrote the paper.
M. Sidiropoulou: Analyzed and interpreted the data; Wrote the paper.

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Competing interest statement

The authors declare no conflict of interest.

Additional information

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