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An unusual cause of epigastric pain and diaphoresis

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ABSTRACT

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. The case reported here refers to a middle aged male patient who presented with intermittent epigastric pain and diaphoresis after a long drive. Diagnosis was made radiologically, during computed tomography scan angiography, which revealed a hook-like appearance of the celiac artery partially loosened during inspiration. Careful history and cautious reviewing of the imaging may refrain from further, unnecessary, diagnostic investigations.

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 due to ischemia caused by compression of the celiac artery, nausea, diarrhea and weight loss [1]. 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.

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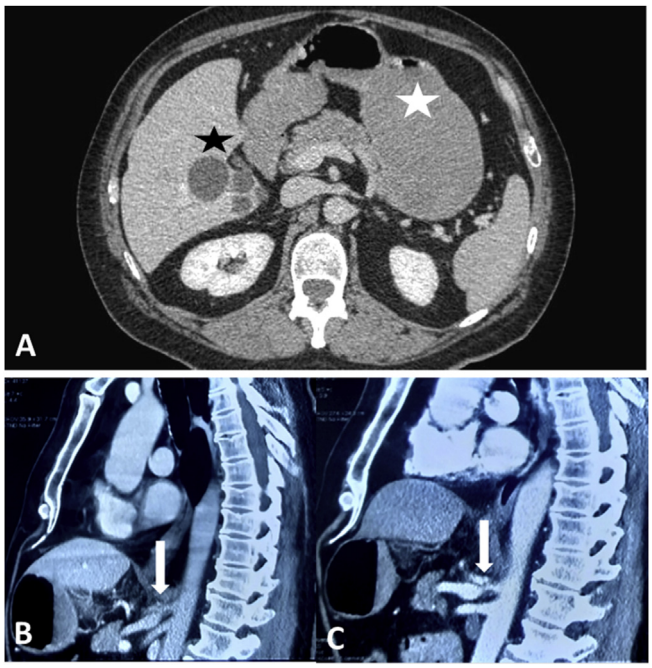

Figure 1. Computed tomography scan images of the thorax and abdomen. A: Axial image of the upper abdomen, showing dilatation of the stomach (white star); black star points to plane cysts of the liver. B,C: Sagittal images of the abdominal aorta and the celiac artery in the inspiration (B) and expiration (C) phase of the respiratory cycle. Arrow shows partial decompression and compression of the celiac artery respectively.

3. Discussion

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. Sympathectomy performed 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 surgical preparations, though symptomatology regressed without removal of the ganglion respectivey [4]. Diagnosis is made by careful reviewing of imaging performed, which deters from other, unnecessary, interventions. Sagittal reformation of computed tomography scan images reveals a hook-like or hook-fish appearance, caused by compression of the proximal celiac trunk by fibrous attachments of the diaphragmatic crura, the median arcuate ligament [5, 6]. Post-stenotic dilatation is observed in about half of the patients. Median arcuate ligament syndrome may be symptomatic or asymptomatic. In fact, celiac trunk compression by the median arcuate ligament was already observed during post-mortem dissections performed by Lipshutz et al in 1917 [7]. Its description, based on clinical symptomatology, followed much later, in the mid 60s. When present, symptoms usually manifest 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 has 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. Gouridou 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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References

[1] Y.C.1 Tseng, C.K. Tseng, J.W. Chou, H.C. Lai, C.H. Hsu, K.S. Cheng, C.Y. Peng, Y.F. Chen, A rare cause of mesenteric ischemia: celiac axis compression syndrome, Intern. Med. 46 (15) (2007) 1187–1190.

[2] M. Koç, H. Artaş, S. Serhatlıoğlu, The investigation of incidence and multidetector computed tomography findings of median arcuate ligament syndrome, Turk. J. Med. Sci. 48 (6) (2018 Dec 12) 1214–1218.

[3] M. Rubinkiewicz, P.K. Ramakrishnan, B.M.1 Henry, J. Roy, A. Budzynski, Laparoscopic decompression as treatment for median arcuate ligament syndrome, Ann. R. Coll. Surg. Engl. 97 (6) (2015 Sep) e96–e99.

[4] Roseborough GS1, Laparoscopic management of celiac artery compression syndrome, J. Vasc. Surg. 50 (1) (2009 Jul) 124–133.

[5] H. Xiang, J. Han, W.E. Ridley, L.J. Ridley, Fish hook coeliac: median arcuate ligament syndrome, J. Med. Imaging. Radiat. Oncol. 62 (Suppl 1) (2018 Oct) 74.

[6] E.N. Kim, K. Lamb, D. Relles, N. Moudgill, P.J. DiMuzio, J.A. Eisenberg, Median arcuate ligament syndrome-review of this rare disease, JAMA Surg. 151 (5) (2016 May 1) 471–477.

[7] B. Lipshutz, A composite study of the coeliac axis artery, Ann. Surg. 65 (2) (1917 Feb) 159–169.

[8] N. Camacho, G. Alves, F. Bastos Gonçalves, R. Abreu, R. Ferreira, J. Catarino, R. Correia, R. Bento, M.E. Ferreira, Median arcuate ligament syndrome - literature review and case report, Rev. Port. Cir. Cardiotorac. Vasc. 24 (3-4) (2017 Jul-Dec) 111.

[9] K.K.F. Ho, P. Walker, B.M. Smithers, W. Foster, L. Nathanson, N. O'Rourke, I. Shaw, T. McGahan, Outcome predictors in median arcuate ligament syndrome, J Vasc Surg 65 (6) (2017 Jun) 1745–1752.

[10] M.V. Patel, L. Dalag, A. Weiner, C. Skelly, J. Lorenz, Inability of conventional imaging findings to predict response to laparoscopic release of the median arcuate ligament in patients with celiac artery compression, J. Vasc. Surg. 69 (2) (2019 Feb) 462–469.

[11] J.C. Jimenez, M. Harlander-Locke, E.P. Dutson, Open and laparoscopic treatment ofmedian arcuate ligament syndrome, J. Vasc. Surg. 56 (3) (2012 Sep) 869–873.

[12] M. Duran, F. Simon, N. Ertas, H. Schelzig, N. Floros, Open vascular treatment of median arcuate ligament syndrome, BMC Surg. 17 (1) (2017 Aug 29) 95.

[13] K.T.1 Delis, P. Gloviczki, M. Altuwaijri, M.A. McKusick, Median arcuate ligament syndrome: open celiac artery reconstruction and ligament division after endovascular failure, J. Vasc. Surg. 46 (4) (2007 Oct) 799–802.