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Celiac artery compression syndrome. Mini-review

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Abstract

Celiac artery compression syndrome is a rare vascular disease, with incidence estimated at 0.4%. However, asymptomatic but hemodynamically significant celiac artery compression is found incidentally in 2.4–8% of the population on examination. The disease is caused by compression of the median arcuate ligament and celiac axis nerve fibers on the celiac artery, usually during expiration, and can cause symptoms of "abdominal claudication". These symptoms include post-prandial epigastric pain, nausea, vomiting, mild weight loss and, less frequently, diarrhea, heartburn, abdominal bloating, constipation, arrhythmias and syncope. Ultrasound, computed tomography (CT), magnetic resonance (MR) and angiography are employed to diagnose celiac artery compression syndrome. Treatment of this disease is based on median arcuate ligament lysis and celiac ganglionectomy either by laparoscopic or open method, with both techniques having similar outcomes.

Key words: celiac artery compression syndrome, celiac trunk, median arcuate ligament, celiac plexus

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Introduction

Celiac artery compression syndrome (CACS) is a rare disease occurring in 0.4% of the population [1], although incidental finding of hemodynamically significant yet asymptomatic celiac artery compression occurs in 2.4–8% of the population [1–3]. The disease is caused by overgrowth of inferiorly placed median arcuate ligament of the diaphragm, causing celiac artery compression during the expiration phase. It can cause symptoms of "abdominal claudication" [4, 5]. An intrinsic component of the disease's pathogenesis is the irritation of the celiac plexus fibers by the overgrown ligament. Common symptoms include post-prandial epigastric pain, nausea, vomiting, and mild weight loss. Less frequently patients experience diarrhea, heartburn, abdominal bloating, constipation, arrhythmias and syncope [6–9].

The anatomical description of celiac artery compression was first published by Lipshutz in 1917 [10]. Celiac artery compression syndrome was later identified by Harjola in 1963 [11] and Dunbar in 1965 [12].

In the contemporary literature, singular cases of diagnosis and treatment of the disease can be found. There are hospitals where, up until 2013, more than 30 patients diagnosed with the syndrome were operated. In the case of pediatric patients under 21 years, University of Chicago Medicine is the leading institution, where 46 patients underwent laparoscopic surgery for the condition between 2008 and 2012 [13]. In University Hospital Dusseldorf, 18 patients with CACS were operated with open method from 2000 to 2009 [7]. In turn, 51 CACS-diagnosed patients were operated in San Francisco hospital until 1984 [14]. Singular cases of typical diagnosis and treatment of CACS can be found in medical literature from 2013, e.g. a paper by Sang Jin Kim from Korea [15].

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Anatomy

The celiac trunk is a single celiac branch of the abdominal aorta. It usually originates between the levels of 12th thoracic and first lumbar vertebrae. The end of the celiac artery lies above the upper edge of pancreas and is 15–20 mm long. Typically, it then divides into three arteries: left gastric artery, splenic artery and hepatic artery, which supply stomach, spleen, pancreas, liver, and duodenum [16].

The diaphragm is divided into lumbar, costal and sternal parts. CACS pathogenesis is related to the lumbar part which originates bilaterally with three arcuate ligaments (median, medial, lateral) and diaphragmatic crura. Median arcuate ligament is a fibrous arch that unites the diaphragmatic crura crossing aorta superior to the celiac axis at the level of the first lumbar vertebra. In addition, another formation surrounding and impinging on the celiac artery is the celiac plexus which comprises of celiac nerves and branches of vagus and phrenic nerves. They are responsible for stimulating the muscular membrane of sphincter muscles in the digestive tract, halting the activity of the other digestive tract muscles, conducting pain stimuli from abdominal organs, and causing abdominal artery stenosis [8, 16].

Diagnosis

The following three conditions need to be satisfied in order to diagnose CACS:

- occurrence of characteristic symptoms of the syndrome;
- exclusion of other causes of the symptoms (ulcerative disease, cancer, etc.);
- confirmation of celiac trunk stenosis without atherosclerosis through additional examination (duplex ultrasound, computer tomography [CT], magnetic resonance [MR], angiography).

Currently, in order to confirm CACS contrast enhanced CT is the gold standard. Other examinations leading to diagnosis are: MR, Doppler ultrasound evaluating blood flow velocity in the celiac artery, and arteriography which is becoming less popular due to its invasive nature. Each of these examinations should be performed both during inspiration and expiration, as the largest compression of the median arcuate ligament occurs during deep expiration but is not always visible during the inspiration phase [17].

Characteristic features on CT angiogram include celiac artery stenosis, significant hooked appearance of the artery in 3D reconstruction in sagittal plane, and post-stenotic dilatation of proximal celiac artery [4].

On ultrasound Doppler, significant stenosis of celiac arteries is thought to occur when the velocity of the blood flow increases two-fold in comparison to the velocity of the blood flow in aorta. It is commonly agreed that the above-mentioned examination is the most accurate form of screening for people with suspected CACS. This is due to the fact that it is a non-invasive, relatively inexpensive examination without adverse effects such as radiation. In addition, ultrasound Doppler is a very effective tool for post-operative control of patients with CACS. The most objective and comparable indicator is the evaluation of velocity of the blood flow in celiac artery before and after surgery. In a study of 46 pediatric patients under 21 years, preoperative mean peak systolic velocity (PSV) amounted to 381 cm/s, whereas postoperative PSV equaled 235 cm/s. This decrease in pressure corresponded to reduced compression of celiac artery and relief of symptoms in majority of cases [13].

Epidemiology

CACS occurs in 0.4% of the population. However, one has to bear in mind that symptom-free existence of celiac trunk compression is estimated at 2.4–8% [1–3]. Authors from Korea prospectively evaluated the incidence and etiologies of celiac axis stenosis in 400 arteriograms of patients referred to them for chemoembolization of hepatic tumors. All patients were free from symptoms of CACS. It turned out that 7.3% patients had celiac axis stenosis, in 55% the etiology was extrinsic compression by the median arcuate ligament [2]. Another study confirming symptom-free occurrence of CACS is a retrospective analysis of 744 patients who underwent CT of the abdominal aorta for various medical indications. 21 patients (2.8%) were shown to have compression of the celiac artery by the ligament, but only 3 of them had symptoms of CACS such as postprandial epigastric pain, vomiting or diarrhea [1]. Due to the frequent incidental finding of asymptomatic celiac artery stenosis on imaging studies, treatment should be dedicated to patients with symptomatic CACS only.

CACS concerns mainly young people between 30 and 50 years of age, in particular women (75–91%) [7, 13, 18]. There are also reports of familial CACS. In 2010, the case of a 16-year-old girl operated for CACS whose mother had previously been treated for the same disease was reported [19]. In 2011, the case of 61-year-old man with hepatitis B-related cirrhosis and hepatocellular carcinoma awaiting transplantation was described. After computed tomography, celiac artery stenosis was detected. The patient, however, was free from CACS symptoms. His four children also underwent a CT in order to qualify as candidates to be living related liver donors. Each of the children was diagnosed

with celiac artery compression by the median arcuate ligament. None of them had any digestive problems or abdominal pains. According to the authors of the study, this is an unequivocal proof of a familial pattern in CACS [20].

It has to be mentioned that the diagnostic process of CACS is usually long, as most patients are qualified for operative treatment after months, and sometimes even years, of gastroenterological examinations, imaging, and sometimes psychiatric consultations. It seems that the main reason behind the status lies with unspecific symptoms but also doctors' lack of awareness about CACS.

Until present, there have been no risk factors identified predisposing for the disease.

Treatment

In CACS, as the name indicates, compression is the source of pathology and has to be addressed surgically. Preferred treatment involves cutting median arcuate ligament and celiac ganglionectomy which can be performed either by laparoscopic or open method. Both procedures are safe and effective. No death has been reported as a postoperative complication and the percentage of patients with immediate symptomatic relief is 85% [17].

However, even in treated cases, symptoms can persist or recur. In cases of incomplete resolution of symptoms, endovascular treatment can be employed as a further therapeutic method. After re-diagnosis and visualization of celiac artery stenosis, endovascular treatment based on celiac artery PTA with possible stent implantation should be considered. The above-mentioned practice was successful in two reported cases and in the follow-up period of 6 and 15 months, symptoms have not reappeared [5, 21].

Medical literature also mentions cases where primary endovascular treatment failed and required serious reconstructive surgery. The case of a 21-year-old woman with CACS who had celiac artery PTA and stenting has been reported. Two months after the procedure symptoms recurred as a result of restenosis. PTA and stenting were repeated. Unfortunately, after 6 weeks early relapse of the condition was diagnosed on grounds of celiac artery stenosis between the two previous stents. The patient was moved to another treatment center where PTA and stenting were performed for the third time. This time the intervention also did not bring satisfying results. In the end the patient was qualified for an open surgery. During the operation damage of stents was established and was followed by a reconstruction of celiac artery with a side-to-end aortoceliac 8-mm Hemashield Dacron interposition graft. Only after this form of treatment satisfying result was obtained and at a 28-month follow-up the symptoms have not recurred. Description of this particular case indicates that endovascular treatment of CACS should not be the preferred method of treatment [6].

Conclusions

Celiac artery compression syndrome is a rarely diagnosed disease with nonspecific clinical symptoms, and one that should be considered after exclusion of other causes of abdominal symptoms. Preferred treatment method of CACS is median arcuate ligament lysis and celiac ganglionectomy either by laparoscopic or open surgery. Results of both methods are comparable, with a high ratio of cured patients and small number of complications.

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