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Celiac artery compression syndrome in children

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Abstract---The manifestation of celiac artery compression syndrome in childhood is rarely described. At the same time, celiac artery compression syndrome remains one of the most discussed vascular diseases due to an unclear pathophysiological mechanism, difficulties in differential diagnosis, and the lack of a generally accepted algorithm for treating patients. In connection with the above, it seems relevant to analyze the features of managing children with celiac artery compression syndrome. As a result of the analysis of the scientific literature, we have come to the following conclusions: 1. Typical clinical characteristics of celiac artery compression syndrome include abdominal pain, epigastric murmur and weight loss associated with compression of the celiac artery and possibly celiac ganglia by the median arcuate ligament. 2. CT angiography and conventional catheter angiography are the gold standard for diagnosis. However, imaging findings should not be interpreted in isolation from clinical manifestations. 3. Surgical decompression of the celiac artery by resection of the median arcuate ligament is the treatment of choice for this syndrome. Both open surgery and laparoscopic techniques are safely performed with minimal morbidity and mortality. Endovascular treatment of compression celiac stenosis may have limited success.

Robotic surgery is a promising trend in the treatment of celiac artery compression syndrome in children.

Keywords---celiac artery compression syndrome, Dunbar syndrome, celiac trunk, stenosis, children.

Introduction

Celiac artery compression syndrome (CACS), also called as a median arcuate ligament syndrome or Dunbar syndrome, is defined as a chronic recurrent abdominal pain associated with compression of the celiac trunk (CT). Benjamin Lipschutz noticed that the celiac artery was sometimes narrowed by excess fibers extending from the diaphragm during a postmortem dissection in 1917. Later, a median arcuate ligament syndrome was described by Pekka-Tapani Harjola in 1963. Harjola was the first to observe a celiac trunk stenosis due to fibrosis of the celiac ganglia and cured the symptoms via resection of the compressive tissue. In subsequent years, the “median arcuate ligament syndrome” term has become frequently cited as “celiac trunk compression syndrome,” as if emphasizing the hemodynamic aspect of its cause. The first clinical study and surgical treatment of abdominal pain via dissection of the arcuate ligament was fulfilled by J. David Dunbar and Samuel Marable in 1965. Therefore, CACS is also found in foreign literature under the name of Harjola-Marable syndrome and Marable syndrome (Chantip, 2020).

The manifestation of CACS at an early age is rarely described (González C. P. et al., 2021). However, R. A. Zainulabidov et al. (Zainulabidov et al., 2021). report that 10–15% of children and adolescents suffering from chronic abdominal pain have CACS. Nowadays, CACS remains one of the most discussed peripheral vascular diseases due to an unclear pathophysiological mechanism, difficulties in differential diagnosis, and the lack of a generally accepted algorithm for treating patients. Based on the aforesaid, it seems relevant to analyze the features of managing children having CACS. The objective of this paper is to analyze the etiopathogenesis, clinical pattern features, diagnosis and treatment of patients having CACS.

Materials and Methods

A search was carried out among scientific papers in the Science Direct, CochraneLibrary, PubMed data bases by keywords (celiac artery compression syndrome, Dunbar syndrome, celiac trunk, stenosis, and children) for 2020–2022.

Results and Discussion

Celiac trunk compression syndrome is a clinical condition caused by compression of the CT by surrounding tissues, usually the median arcuate ligament (MAL). MAL is a fibrous arch connecting the crura of the diaphragm on either side of the aortic ostium. The MAL usually contacts the CT and the aorta at a level between T₁₁ and L₁, at the origin of the celiac artery, which prones to compress during

expiration. Due to the closer anatomical location of MAL to the celiac artery, this syndrome is more common in women than in men. Various hypotheses of the etiology of this disease have been proposed: the first of them is the abnormal location of the proximal CT spine, leading to its compression by the normally located MAL. According to another hypothesis, an abnormally longer MAL compresses the CT proximal part. Besides, etiology suggests compression by surrounding tissues, such as periaortic ganglion tissue, fibrous cords, or stenosis caused by atherosclerotic plaques (Güngörer et al., 2020.).

The young age of patients may indicate the intrinsic character of this syndrome. Thus, myotonic dystrophy, Marfan's syndrome can occasionally include stenosis of the celiac trunk in its various manifestations. However, there are theories of the acquired nature of this disease. It is assumed that the sclerogenous process (inflammatory or degenerative) causes cicatricial retraction of the diaphragm fibers followed by lowering of the aortic ostium lower edge. This hypothesis is supported by the frequent association of the sclerogenous process of the retroperitoneal space connective tissue around the aorta among persons older than 50 years. Prolonged friction of fibrous-sclerotic structures along the celiac trunk can cause changes in the myoelastic tone of the vessel, and further lead to the formation of an atherosclerotic lesion of this vessel or aorta.

Two theories have been proposed for explaining the symptoms of CACS. The first theory suggests the development of mesenteric ischemia due to a decrease in blood flow because of compression. The second theory states that the syndrome develops with neurogenic stimulation due to compression of the celiac ganglion or plexus, which can lead to irritation of sympathetic fibers. Celiac artery compression may also cause poststenotic expansion or the formation of a prominent distal aneurysm. The development of the pancreatoduodenal artery aneurysms is also associated with the celiac artery compression.

Let us note that patients with CACS are mostly asymptomatic. In symptomatic cases, postprandial abdominal pain, nausea, vomiting, diarrhea, loss of appetite, and abdominal discomfort are common (Aldahas et al., 2021; Chaum et al. 2021). Therefore, CACS is easily misdiagnosed as dyspepsia or peptic ulcer disease. Let us note that weight loss is detected in about half of patients. Some patients have chronic incessant epigastric pain, which sometimes radiates to the left side or back.

Symptoms increase during expiration, since the pressure on the CT increases even more due to the diaphragm movement, and the symptoms become more noticeable. During inspiration, the celiac artery descends into the abdominal cavity, resulting in a more vertical orientation of the CT, which often reduces compression. An abdominal murmur that is detected during expiration and disappears during inspiration increases the likelihood of a clinical diagnosis. Particular positions, such as forward pitch, can allay or alleviate abdominal pain. In the upright position of the patient, the celiac artery descends further into the abdominal cavity, which further increases the vertical orientation of the CT and additionally reduces the compression caused by MAL.

Symptoms of CACS have similar characteristics to mesenteric ischemia caused by atherosclerotic occlusive lesions of major vessels. However, postprandial discomfort in CACS is less pronounced than in mesenteric ischemia. There are some clinical aspects that allow differentiation of celiac trunk compression from atherosclerotic celiac-mesenteric ischemia, such as the predominance of pains to the level of the abdomen upper quadrant and weight loss due to a deliberate reduction in food intake and not as a result of the malabsorption process. Besides, unlike diarrhea in atherosclerotic diseases of the mesenteric arteries, diarrhea in CACS is more associated with irritable bowel than with malabsorption. Let us note that childhood often makes it possible to exclude the diagnosis of mesenteric arterial insufficiency of atherosclerotic origin.

It is known that the frequency of isolated CT stenosis among asymptomatic individuals ranges from 2.3% to 7.2%. This figure increases to 24% in a series of autopsies (Güngörer et al., 2020). Although celiac artery stenosis is widespread, the fact that CT compression syndrome does not develop in most cases can be explained by the fact that celiac artery stenosis is minimal in most cases or there is a normal blood flow nature due to collaterals from the superior mesenteric artery to the celiac artery or its branches.

Such methods as Doppler ultrasound, computed tomography angiography, selective angiography, and magnetic resonance angiography can be used to diagnose CACS to visualize celiac artery compression. If CACS is suspected, patients should undergo duplex ultrasound. The diagnosis is confirmed by an increase in blood flow velocity in the celiac artery, which normalizes with deep inspiration followed by computed tomography angiography (Skelly and Mak, 2021). Although selective catheter-based angiography is the gold standard for diagnosis, computed tomography angiography is used more frequently due to its non-invasive nature.

Some anomalous aspects of the CT direction and gauge obtained by angiography make it possible to diagnose stenosis of this MAL vessel. One of them is the vertical direction of the proximal celiac trunk spine, becoming parallel and pressing against the anterior wall of the aorta by the lower edge of the arcuate ligament in its first centimeters, which significantly compresses the lumen of the CT and provides the vessel with a vallecule on its anterior surface in the “axe blow” form. Moreover, distal to the “vallecule”, the CT takes on a horizontal or oblique upward deviation in the “elephant trunk” form. Another aspect is reducing the angle between the posterior surface of the celiac trunk and the underlying aorta.

J. Varter made proposals to classify various morphological and radiological characteristics of CT stenosis with due regard for its more or less pronounced degree. Five types of stenosis are distinguished in J. Varter's classification. Type I is characterized by initial narrowing of the celiac trunk and poststenotic dilatation. The narrowed and long part of the celiac trunk appears pressed against the aorta in IA and IC subtypes, while an empty space can be seen between the stenosed segment and the aorta in IB subtype. Type II gives the CT a fusiform shape with or without poststenotic dilatation, and atheromatous isolated stenosis and fibromuscular hyperplasia of the celiac trunk can also be identified

in this type. In type III, it is possible to verify the presence of an upper and lower notch in the CT without poststenotic dilatation, including compression by hypertrophied ganglia of the celiac plexus and periarterial fibrous sheath. The CT gauge is reduced and includes Reiter's and Ohlin's atheroma in type IV. Type V is characterized by complete obliteration of the celiac trunk. If IA type is the most common and occurs mainly among adults, then IB type occurs in young people or thin women.

The objective of treatment of patients with CT compression syndrome is to normalize blood flow in the artery for eliminating symptoms. This objective can be achieved by dissection of the MAL using an open or laparoscopic surgical technique (Güngörer et al., 2020). Let us note that, regardless of the technique, assessment of the blood flow adequacy should be included in any surgery regarding CACS, since further additional vascular reconstruction may be required. The assessment is fulfilled via intraoperative duplex ultrasound.

It is possible that the complete hemodynamic release of the CT depends both on the transection of the arcuate ligament and on the resection of the periarterial neurofibrous tissue. Thus, a certain degree of periarterial fibrosclerosis can be observed during surgical operation among some patients. Therefore, an extensive denervation of the celiac plexus and periarterial sympathectomy are performed in certain cases.

In case of an arcuate incision of the fibers of the diaphragm aortic hiatus, the retraction of the muscle fibers makes it possible to expose the initial segment of the celiac trunk and its origin in the aorta. After dissection of the MAL, it can be seen that the anomaly is quickly resolved with normal expansion of the CT and a decrease in the pressure gradient between the CT and the aorta. A weak pulse on the hepatic, gastroduodenal arteries and the absence of a pulse on the left gastric artery are restored after the ligament dissection.

Nowadays, there has been a similar efficacy in alleviating symptoms using open and laparoscopic methods (Chantip, 2020; Goodall et al., 2020). The traditional surgical approach is supramedian laparotomy and thoracophrenolumbotomy. However, traditional surgical approaches are associated with injury rate and a long postoperative period. The advantages of the laparoscopic approach include early oral alimentation, shorter hospital stay, and less risk of postoperative adhesions. Disadvantages of the laparoscopic approach include difficulties in dissection and bleeding control (Sharma, Someshwar, Ingale, 2020).

Percutaneous transluminal angioplasty and stent placement are also possible, but the recurrence rate is high and the duration of symptomatic relief is short. This may be due to the continuing external pressure of the surrounding tissues. However, angioplasty with or without stenting may be considered in case of residual stenosis or symptomatic relapse after surgical decompression of the CT, if resources are available.

B. Roberts, R. Pevsner, F. Alkhoury (Roberts, Pevsner, Alkhoury, 2020). propose to use more actively robotic assisted surgery among children having CACS. In their paper, two patients aged 12 and 15 years underwent robotic dissection of

the MAL fibers and the scar tissue covering them. After surgical operation, patients remained in the hospital overnight, and the average outpatient follow-up period was 10 months. As a result, both patients had immediate pain relief after surgical operation, and there were no symptomatic relapses after 10 months of follow-up. There were no complications associated with the procedure. Quite often, after dissection of the MAL, minimal residual stenosis in the CT compressed area, distal murmur, and pressure gradient are revealed intraoperatively. However, there is a regression and disappearance of the "vallecula," and a return to the normal gauge of the stenotic CT over the course of time as a rule.

If, after dissection of the MAL and/or periarterial neurofibrous tissue fibers, the CT does not restore its normal gauge and the pressure gradient across the stenotic segment remains significant, reconstruction of the segment with an autograft, shunting, or endarterectomy is necessary. Histologically similar stenoses are associated with intimal fibrosis or hyperplasia, sometimes with associated atheromatous lesions. Moderate intimal changes in the form of fibrosis and hyperplasia are probably inversive among young patients and do not require reconstruction.

An arterial pressure in the hepatic or left gastric artery before and after removal of the CT compression in such cases is a more sensitive and reliable test for assessing the immediate results. If there is no considerable increase, the artery can be quickly reconstructed. Continuous abdominal noise after surgical operation presupposes persistence of poststenotic dilatation, which may be sufficient to cause turbulence. It may disappear after a few months or not disappear completely. The return to normal angiographic images represents the most convincing evidence that improvement in the clinical pattern of ischemia depends on the restoration of normal anatomical relationships at the CT level.

Conclusions

As a result of the performed analysis of scientific literature, we have come to the following conclusions:

1. Typical clinical features of celiac trunk compression stenosis include abdominal pain, epigastric murmur, and weight loss associated with compression of the celiac artery and possibly the celiac ganglia by the median arcuate ligament.
2. Computed tomography angiography and traditional catheter-based angiography are the gold standard for diagnosis. However, imagery findings should not be interpreted regardless of clinical manifestations.
3. Surgical decompression of the celiac artery via resection of the median arcuate ligament is the selection method for this syndrome. Both open surgery and laparoscopic techniques are safely performed with minimal morbidities and mortality. Endovascular treatment of celiac artery compression syndrome may have a limited success. Robotic assisted surgery is a promising trend in the treatment of a celiac artery compression syndrome among children.

Conflict of interest

The authors declare no conflict of interest regarding the research, authorship and publication of this paper.

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