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Case Report

Should Dunbar Syndrome be Operated on? (Clinical Case and Literature Review)

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Abstract

Currently, the syndrome of compression of the abdominal trunk is commonly understood as a symptom complex that occurs due to extravasal compression of the abdominal trunk by the median arched ligament of the diaphragm, the legs of the diaphragm, nerve fibers and/or neurofibrous tissue of the solar plexus.

This nosological form belongs to a group of vascular diseases that lead to a disorder of visceral circulation. The symptom complex caused by a stenosing or occlusive lesion of the unpaired visceral arteries has been called "angina abdominalis", since this pathology clinically proceeds with the appearance of attacks of abdominal pain at the time of increasing functional load on the digestive organs. Other names of this condition caused by the lesion of unpaired visceral arteries: chronic intestinal ischemia, "visceral angina", chronic intestinal ischemia, chronic abdominal ischemia syndrome, etc.

Keywords: Malnutrition; abdominal trunk;

Reports on the clinical picture of this disease were more descriptive until the XIX century.

In 1965, the American radiologist J. Dunbar, in collaboration with surgeons F. Beman and S. Marble, described the result of observations of 27 patients with the clinic of chronic abdominal ischemia. In 15 people, the cause of ischemia was extravasal compression of the abdominal trunk by the median arched ligament, confirmed by angiography and during surgery. Ventral trunk decompression was performed in 13 patients with a positive clinical effect in 12 of them. During the control angiography, the normal course of the vessel was observed. In the English-language literature, this disease has entered under the name "ligamentum arcuatum syndrome" (syndrome of the median arched ligament) or "Dunbar syndrome".

The first operation on the abdominal trunk in the USSR was performed by the outstanding Russian surgeon academician A.V. Pokrovsky on May 25, 1962 in a patient with a clinical picture of abdominal angina. The ventral trunk and the common hepatic artery were decompressed. In 1962, for the first time in the world, A.V. Pokrovsky used retroperitoneal access through the chest and diaphragm to perform manipulations on the thoracoabdominal aorta and visceral arteries. This technique quickly gained popularity among domestic surgeons and became the main method of performing reconstructive interventions on the aorta and its branches, receiving the name "Russian access" abroad. Techniques developed by A.V. Pokrovsky for a long time were used when performing surgical interventions for decompression of the abdominal trunk.

Most often, young female patients suffer from this syndrome, abdominal pain (80%), weight loss (48%) are the most frequent symptoms of varying severity. Auscultatively, in 83% of cases, patients listen to systolic noise in the epigastrium, especially on exhalation. The main manifestations of the disease are abdominal pain after eating, dyspeptic disorders. Dyspeptic disorders, unlike abdominal pain, bother patients not only after eating. Often they become permanent and do not respond well to drug therapy.

Because of the pain and nausea after eating, patients are often afraid to eat and gradually begin to lose weight. Malnutrition and poor blood supply cause weakness and fatigue. Sometimes the weight does not decrease, but it is impossible to gain it with stenosis. In laboratory indicators, there is a decrease in the level of serum iron, hemoglobin.

It usually takes more than one year from the appearance of the first symptoms of stenosis to the correct diagnosis. Very often, patients with this pathology come to an appointment with a surgeon, gastroenterologist, therapist or are urgently admitted to the hospital with suspicion of other acute diseases, such as pancreatitis, biliary colic, acute appendicitis, gastric or duodenal ulcer. They are operated on for suspected diseases, but the symptoms do not disappear.

The most popular and practically significant is the surgical classification of A. A. Spiridonov. It helps to understand whether the patient needs surgery, predict and evaluate the results of treatment.

Together with co - authors A. A. Spiridonov divided the development of the disease into four stages:

- Compensation stage:
- Ia — asymptomatic stage;
- Ib — pain in the liver and spleen occurs when the digestive organs are overloaded, i.e. after overeating or excessive physical exertion.
- Subcompensation stage — symptoms occur after a normal meal and/or moderate physical activity.
- Decompensation stage — symptoms occur regardless of the load and food intake, the pain becomes constant.
- Stage of ulcerative-necrotic changes — ulcers appear in the stomach and duodenum, enteritis and colitis develop. All these changes relate to complications of abdominal trunk stenosis. But they can occur both at the II and III stages of the disease.

Conservative treatment does not affect the underlying cause of the disease. It is purely symptomatic or aimed at concomitant diseases.

Some clinics include psychotherapy and acupuncture in complex treatment. The latter method of treatment does not have an unconditionally evidence base, but many practitioners note its effectiveness in complex therapy.

Compression stenosis of the abdominal trunk does not threaten life, but significantly affects its quality. Many patients complain that they are afraid to eat and it is difficult to work because of pain or general malaise.

With effective conservative treatment of stenosis, the prognosis is favorable: the patient can safely work and eat. But if the disease goes into the stage of decompensation, the symptoms progress, and conservative treatment does not bring relief, the cause can only be eliminated by surgery. Without it, the prognosis will be unfavorable. Therefore, patients are advised to consult a doctor as early as possible and undergo all prescribed examinations. This will help to identify the disease in time and determine the tactics of treatment.

It is impossible to prevent the development of stenosis, since this is an anatomical feature. To reduce the risk of developing symptoms, it is recommended to give up smoking, eat properly, eat in a balanced way and not physically overload yourself.

The pathophysiological mechanisms of the abdominal trunk compression syndrome are caused by a violation of hemodynamics in the abdominal trunk and associated arteries of the abdominal cavity. The abdominal trunk supplies blood to the organs of the upper floor of the abdominal cavity. Its branches are anastomosed with the branches of the superior mesenteric artery, which supplies blood to the colon and small intestine. A decrease in blood flow in at least one of the unpaired visceral arteries leads to a shortage of blood supply to the entire gastrointestinal tract and the development of abdominal ischemic disease.

Clinical observation

Patient M, 18 years old., was admitted on 12/26/2020 with complaints of: general weakness, feelings of "lack of air", dizziness, periodic dull aching pains in the upper half of the abdomen, increasing after eating, nausea, periodic vomiting after eating.

Anamnesis: syncopal conditions since October 2019, the first attacks were accompanied by headache with chills, abdominal pain, relieved by the adoption of the knee-elbow position. The onset of seizures after a viral infection. At the most critical moment (2019), she was examined by vascular surgeons at the A.V. Vishnevsky National Medical Research Center for Surgery and the A.N. Bakulev National Medical Research Center for Cardiovascular Surgery of the Ministry of Health of the Russian Federation diagnosed with abdominal stenosis.

According to the mother, they are categorically against the operation, because the result is unfavorable. When examined at the age of 8 at the Research Clinical Institute of Pediatrics and pediatric surgery named after Academician Yu.E. Veltischev of the Federal State Educational Institution of the Russian National Medical University named after N. I. Pirogov of the Ministry of Health of the Russian Federation revealed: Ehlers-Danlos syndrome, Secondary disorders of mineral metabolism (osteoporosis compression fracture of the vertebra mitral valve prolapse, dilation of the left ventricular cavity, right-sided nephroptosis, hyperactivity syndrome. The pedigree notes: father of 58 years — bronchial asthma from the age of 3, mother of 57 years - autoimmune thyroiditis, brother of 33 years - autoimmune thyroiditis, atopic bronchial asthma, paternal cousin, 40 years and her son — celiac disease.

I have lost 3 kg in the last 2 months.

Objective status: The patient's condition is satisfactory. Low nutrition, asthenic physique. Hemodynamics is stable, with a tendency to hypotension. The abdomen is soft, sensitive to palpation in the epigastrium, right and left hypochondrium. Systolic murmur is heard over the abdominal aorta.

No deviations from normal indicators were detected in laboratory studies.

Blood test for selective screening from 22.02 2018: revealed changes in the nucleotide sequence: in a heterozygous state. Pathogenicity prediction algorithms regard this variant as probably pathogenic. Depth of reading x59. Spondyloepimetaphyseal dysplasia type of Kozlovsky HELL inheritance.

Echo KG from 01.02.2019: echo signs of mitral valve prolapse, tricuspid valve dysfunction.

MSCT of the abdominal cavity from 07.11.2019: central narrowing of the lumen of the mouth of the ventral trunk to the threadlike for 12 mm before its bifurcation. hemodynamic significant stenosis of the abdominal trunk.

Duplex scanning of the abdominal aorta from December 26, 2019: the blood flow in the abdominal trunk is moderately turbulent, the rate of calm breathing is 126 cm/s, high-speed blood flow on exhalation is 208 cm/s - the presence of hemodynamically significant stenosis, possibly functional, is not excluded.

Duplex scanning of the abdominal aorta from 01.06 2021: dynamic extra vasal compression of the abdominal trunk. It is impossible to exclude the narrowing of the mouth of the ventral trunk. Donbar syndrome.

Consultation of the surgeon on 06/03/2021: hospitalization in the A.N. Bakulev NCSSH is recommended, for further examination and resolution of the issue of treatment tactics.

Literature Review and Discussion of the Problem

The issues of etiology, pathogenesis and choice of treatment for Dunbar syndrome remain largely unresolved until now. A number of authors generally dispute the existence of such a nosology, and the diagnosis of stenosis of the abdominal trunk in many cases is like a diagnosis of exclusion.

The main causes of this disease indicate in favor of an innate nature, however, symptoms in childhood are noted casually rarely and occur mainly in middle age. Perhaps one of the assumptions of later clinical manifestations is associated with the exhaustion of compensatory mechanisms, although the equally developed collateral network in symptomatic and asymptomatic patients with this pathology, detected by angiography, suggests that the occurrence of symptoms is caused not only by insufficiency of the collateral bed.

In our example, the occurrence of symptoms due to critical stenosis of the abdominal trunk is confirmed by genetic determination.

In a patient suffering from connective tissue dysplasia, gene sequencing revealed changes in the nucleotide sequence responsible for bone pathology. In the family history, there are immediate relatives with autoimmune diseases.

In the early stages of the disease, the nonspecific nature of the clinical picture led to the fact that the patient was examined for a long time by doctors of various specialties not only about the pathology of the abdominal organs, but also neuropathologists, cardiologists, therapists, urologists, surgeons diagnosed their pathology and recommended treatment, which was without effect and did not cause proper alertness and was treated for a long time out of connection with vascular pathology.

Despite her young age, observations of family heredity, lack of effect from treatment, increased psychoemotional loads (a student of Bauman Technical University), as well as the appearance of the patient – asthenic physique. There was no reason to doubt that the cause of this could be abdominal ischemia due to compression of the abdominal trunk. In women, the disease occurs much more often, in a ratio of 4:1 to men. If you suspect a stenosis of the abdominal trunk, first of all, you should pay attention to the classic picture of the syndrome characterized by abdominal pain, usually 20-30 minutes after eating, impaired intestinal function (often expressed in repeated diarrhea), weight loss. It was present in her.

With normal laboratory tests characterizing the work of the gastrointestinal tract, the endocrinological system against the background of disorders of the autonomic nervous system with somatophoric vasovagal syncopal states.

In our observation, colleagues drew attention to the auscultation of the abdominal cavity, namely the presence of systolic noise in the epigastrium, which increases on exhalation, which serves as an important differential diagnostic symptom. However, the absence of noise cannot serve as a reason for exclusion, subtotal stenosis or occlusion of the ventral trunk may not give noise symptoms.

Therefore, the next stage of diagnosis was the use of minimally invasive techniques: Dopplerography and duplex scanning, digital subtraction angiography, spiral computed tomography, magnetic resonance angiography. By now, the diagnostic criteria for each of the methods have already been well developed.

At the present stage, multispiral computed tomography with contrast of abdominal vessels has a high resolution, and if no surgical intervention is planned, this study can serve as the final stage of diagnosis.

But in this clinical case, the remaining clinical manifestations and the surgical treatment proposed by the surgeons were rejected by the patient's mother because of the search for any unusual disease. "Masha, as always, has an atypical manifestation of the disease." (Masha, as always, has an atypical manifestation of the disease, categorically against diagnostic laparoscopy and the operation itself in general. Many people have had this operation more than once, the result is not always good. Now, we are not considering it. We hope for conservative treatment. I'm not going to accept the situation. We have already understood a lot, I will dig further). From 2017 to the present, the patient has been hospitalized 8 times with different diagnoses and has been consulted by various medical specialists more than 10 times.

In the case of a planned surgical intervention, angiography, especially in the lateral projection with respiratory tests, is the most informative in the diagnosis of anatomotopographic features and signs of abdominal trunk stenosis. Indications for surgical treatment are usually determined in the presence of symptoms of abdominal ischemia, proven critical compression of the abdominal trunk and ineffectiveness of conservative therapy. Methods of surgical treatment for abdominal ischemia are divided into palliative, decompression and reconstructive. Decompression operations, i.e. aimed at dissection or removal of various congenital or acquired compression factors (resection of the ligament or legs of the diaphragm, removal of a tumor, aneurysm, etc.) are the most frequent, accounting for up to 55% of the number of operations performed for chronic ischemia of the digestive organs. In the case of compression of the abdominal trunk by an arched ligament or the legs of the diaphragm, most authors give good results, extremely low mortality and the frequency of complications.

Such optimism and the relative simplicity of decompression operations prompted the search for even less invasive techniques.

In recent years, there have been reports in the literature about the successful use of laparoscopic techniques for decompression of the abdominal trunk and robot-assisted operations. However, if, after separation of the structural elements of the diaphragm and/or periarterial nerve tissues involved in the process, the stenosed ventral trunk does not recover to normal caliber and a significant pressure gradient remains at the site of stenosis, reconstruction of this segment is required by resection of the stenosed area with end-to-end anastomosis or prosthetics/bypass surgery or endarterectomy. Completing this stage requires conversion to open intervention.

In the available literature, there are no indications of criteria that make it possible to reliably predict the final volume of intervention at the preoperative stage. The possibilities of interventional radiology – balloon dilation and stenting of ventral trunk stenosis – are evaluated as contradictory and, together with the absence of a significant number of observations and long-term results, do not allow considering this method as a choice. The choice of access is at the discretion of the surgeon.

The final volume of the operation becomes clear only after the elimination of the squeezing factor. Although the exact percentage of favorable surgical results is unknown, in most publications it exceeds 80% [1-8].

Conclusion

The given clinical example and literature review show a number of problems in the diagnosis and treatment of abdominal stenosis. The literature data and a little personal experience allowed us to formulate a preliminary algorithm for the successful solution of these issues.

It is based on the clinical picture - manifestations of abdominal ischemia. Auscultation of the abdominal cavity should be a mandatory study. The detection of pathological vascular noise, its connection with breathing or other functional tests serves as a justification for conducting duplex scanning of the visceral branches of the aorta. And the combination of clinical manifestations - abdominal ischemia and confirmed by ultrasound examination of abdominal trunk stenosis is an indication for the final stage of diagnosis – multispiral computed tomography with contrast or angiography of visceral arteries.

The choice of the intervention method – open, laparoscopic or endovascular surgery depends on a number of circumstances and today should be considered individually. With the accumulation and systematization of experience, it is possible to develop more detailed indications for various types of surgical treatment.

The above clinical observation confirms the complexity of the diagnosis. Awareness of specialists about the features of this disease and timely diagnosis contributes to a faster appointment of treatment. Conflict of interest. The authors declare that there is no conflict of interest.

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