CASE STUDY

DUNBAR SYNDROME: CLINICAL MANIFESTATION IN ADULTS, DIAGNOSTIC PROBLEMS (CASE REPORT)

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ABSTRACT

In this case we used CBC and biochemical tests, ECG, ultrasound of the abdominal cavity and heart, CT scan with and without IV contrast. Women with complains on recurrent severe abdominal pain in epigastric region and right part of the abdomen radiating to the back, nausea, vomiting. Making laboratory and instrumental tests for confirmation of the diagnosis. Surgical treatment of DS was performed and after one year of the follow up there were no complications. DS may mimic other medical conditions such as gallbladder diseases, gastritis/peptic ulcer, appendicitis, colorectal malignancy, hepatitis, atherosclerotic diseases etc. That is why DS is a diagnosis of exclusion. This case illustrates pathway to find correct diagnosis and improve management tactic.

KEY WORDS: Dunbar syndrome, median arcuate ligament, case report

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INTRODUCTION

The compression syndrome of the celiac artery is a complex of symptoms, accompanying the outcast compression of the celiac artery by anatomical elements (sickle bunch and / or medial legs of the diaphragm, solar plexus, tumors, etc.) and due to narrowing disruption (stenosis) His lumen with the formation of chronic mesenteric syndrome ischemia. Typically, the ligament runs across the largest blood vessel in the body (aorta) and sits above the celiac artery without causing problems. But sometimes the ligament or artery may be out of place, causing syndrome. The ligament may also put pressure on the network of nerves surrounding the celiac artery (celiac plexus) [1].

This syndrome also known as MALS (Median Arcuate Ligament Syndrome), CACS (celiac artery compression syndrome), Dunbar syndrome (DS), Harjola-Marable syndrome.

DS is different than median arcuate ligament compression. Median arcuate ligament compression occurs in about 10% to 25% of the population and doesn't cause any symptoms. Chronic abdominal pain is a very common condition that can have significant negative, long-term psychosocial consequences, including increased risk for anxiety, school and work absences, poor functional capacity, and a poor quality of life [2].

The cause is not fully understood; however, it is suspected that there could be a combination of vascular (blood supply) and neurogenic (neurological) components involved [3].

The abdominal pain after eating is the most common symptom, found to be present in approximately 80% of individuals, while weight loss was found in approximately 48% and abdominal bruit was appreciated in approximately 35% [4].

Other symptoms include: weight loss (usually >20 pounds), and abdominal bruit (abnormal sound of a blood vessel when blocked or narrowed), nausea, diarrhea, vomiting, and delayed gastric emptying [3, 5].

Diagnostic methods include different variants of imaging, such as CT scan, MRI, ultrasound, and arteriography. But in clinical practice sometimes it is hard to say when it is necessary to use this methods, especially looking on rareness of DS.

CASE REPORT

Women M., 37 yo was admitted to the City Clinical Hospital (Vinnitsa) with complains on severe abdominal pain in epigastric region and right part of the abdomen radiating to the back, nausea, vomiting. History: these complains appeared rapidly day ago; used NSAIDs to relief pain without any effect. She noticed epigastric abdominal pain sometimes, but NSAID usually helped.

CBC: Hb – 140 g/L, RBC – 4,3 T/L, platelets – 299 G/L, WBC – 7,5 G/L, stabs – 6%, segmental – 52 %, eosinophils – 1%, lymphocytes – 38%, monocytes – 3%, ESR – 10 mm/h.

Glucose level 3,7 mmol/l

General protein 63g/L, ALT 0,15 U/L, bilirubin: general 13,1 μmol/l, conjugated 4,5 μmol/l, unconjugated 8,6 μmol/l.

Prothrombin index 84 %, serum fibrin 2,0 g/L

Serum C-reactive protein 30 mg/L (Normal range 4-10 mg/L).

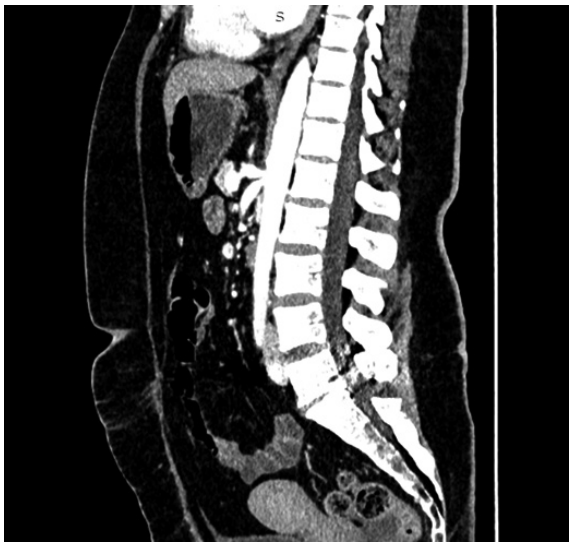

Fig. 1. CT of the abdomen, sagittal view

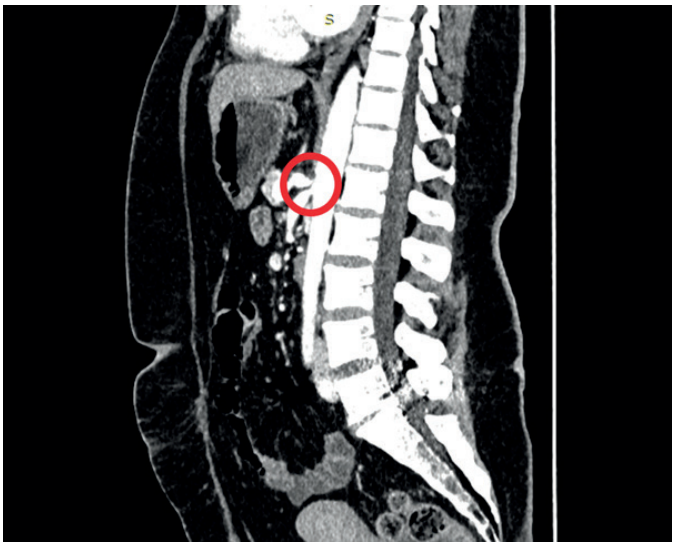

Fig. 2. CT of the abdomen, sagittal view

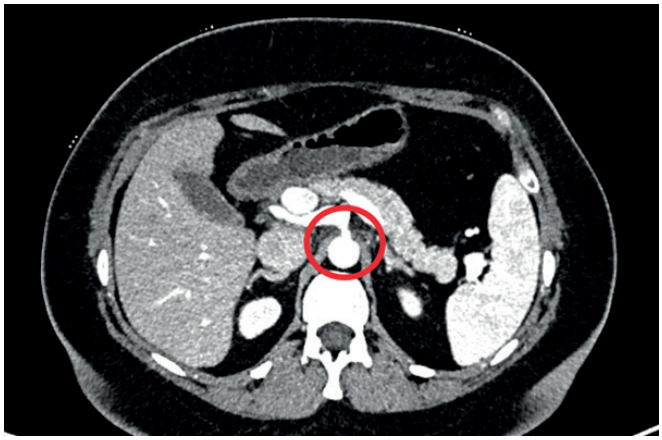

Fig. 3. CT of the abdomen, axial view

Serum lipase 165 U/L (Normal range 10-140 U/L), L-amilase 220 U/L

Procalcitonin 1,8 ng/mL (Normal range 0.15 and 2.0 ng/mL)

Ultrasound of the abdominal cavity: changes in the pancreas, no signs of biliary tract disoders.

EchoCG: left ventricle diastolic dysfunction, EF 68%, dilatation of left atrium and ventricle.

Chest X-ray: normal

Diagnosis: Acute pancreatitis, mild severity

She was treated with rehydration therapy and pain control for a week. A regular diet was recommended on admission.

She was admitted to the hospital one more time after 3 weeks with the same complains.

CBC: Hb – 145 g/L, RBC – 4,2 T/L, platelets – 305 G/L, WBC – 7,2 G/L, stabs – 1%, segmental – 60 %, eosinophils – 1%, lymphocytes – 33%, monocytes – 5%, ESR – 10 mm/h.

Glucose level 4,3 mmol/l

Prothrombin index 87 %, serum fibrin 2,2 g/L

L-amilase 328 U/L

To differentiate the diagnosis CT scan of abdominal cavity (without IV contrast) was performed: diffuse changes of the pancreas; concernments in both kidneys. As a result, therapy of acute pancreatitis was repeat with IV fluids and pain control used epidural analgesia.

Third episode of the postprandial abdominal pain occurred after 4 weeks she was discharged from the surgical department.

CBC: Hb – 149 g/L, RBC – 4,5 T/L, platelets – 262 G/L, WBC – 7,55 G/L, stabs – 8%, segmental – 54 %, eosinophils – 1%, lymphocytes – 30%, monocytes – 7%, ESR – 15 mm/h.

Glucose level 3,5 mmol/l

General protein 71 g/L, urea 5,8 mmol/L, creatinine 105 μmol/l, bilirubin: general 9,1 μmol/l, conjugated 2,8 μmol/l, unconjugated 6,3 μmol/l.

Prothrombin index 81 %, serum fibrin 2,9 g/L

Heart ultrasound: left ventricle diastolic dysfunction, EF 68%, dilatation of left atrium and ventricle, aortic insufficiency I degree.

Ultrasound of the abdominal cavity: diffuse changes of the liver and pancreas.

FEGDS: catarrhal esophagitis, catarrhal gastroduodenitis, signs of pancreatic-biliary disorders.

CT scan with IV contrasting: mediastinum lipomatosis. Compression of the celiac trunk by the diaphragmatic crurae, collateral circulation between gastroduodenal artery and superior *mesenteric* artery (fig. 1, 2, 3). Ascites. Concernments in both kidneys. Transpedicular fixing system.

Diagnosis: Median arcuate ligament syndrome (MARS). Chronic rheumatic heart disease, non-active stage, combined mitral heart defect: mitral insufficiency II degree and mitral stenosis I degree, heart failure 0 degree.

Treatment. Endovascular celiac artery stenting with release of the median arcuate ligament were performed. One year follow up didn't find any complications or recurrence of the symptoms.

DS occurs most frequently in females aged 40 to 60 with duration of symptoms ranging from 3 months to 10 years

[6, 7]. There are several debates in the diagnosis and treatment of DS as this is rare clinical situation. Postprandial abdominal pain, vomiting, and weight loss are the typical clinical symptoms for this syndrome. However, these patients are frequently diagnosed and treated as gastritis, peptic ulcer or acute (chronic) pancreatitis before the correct diagnosis is confirmed [8].

The etiology of DS remains unknown but a case report of monozygotic twins suggests a congenital origin [9]. Whether the pathophysiology is primarily vascular or neuropathic origin remains undetermined [10]. Isolated vascular compression of the celiac artery as the sole etiologic factor seems unlikely. DS can cause asymptomatic compression in 10%–24% of the population. Collateral circulation by the superior mesenteric artery provides adequate blood supply; therefore, postprandial abdominal pain should not be expected with celiac artery compression alone [11].

The compression of the celiac artery by the median arcuate ligament is believed to cause intermittent mesenteric ischemia. However, this explanation alone may not completely explain the condition as there is usually a rich collateral network of mesenteric vessels between the celiac artery and the superior mesenteric artery. Therefore, there may be a role for underlying celiac nerve plexus dysfunction as well when considering the etiology of this condition. Nerve dysfunction may lead to abnormal splanchnic vasoconstriction, leading to ischemia [6].

The etiology of DS is likely multifactorial, including compressive effects on the celiac artery and surrounding neurogenic structures. In celiac artery compression, it has been noted that either the celiac artery is located slightly higher or the median arcuate ligament located lower than expected.

Nowadays, surgery is the only treatment option for DS. Several management strategies are instituted for the treatment of DS including interventional angioplasty or stenting and surgical procedures such as releasing of the median arcuate ligament compressing the celiac artery or bypass surgeries [12, 13]. The releasing of the ligament may be done by open, laparoscopic or robotic surgery [14, 15]. Minimally invasive techniques have gained popularity in the surgical management of this condition limiting open surgery to more complicated celiac by-pass procedures. Some patients may require prolonged use of analgesics while some others may necessitate para-spinal celiac ganglion blockage as an additional measure to completely alleviate the symptoms [8].

In a large series, significantly higher symptomatic relief was achieved through combined release of the MAL and revascularization. If celiac artery compression alone is corrected, evidence suggests up to 53% will be asymptomatic on long-term follow up. Combined release and revascularization however increase the long-term success to 79% [16]. Therefore, some patients may require additional revascularization procedures such as mesenteric artery stenting or bypass to provide long-term symptomatic relief as in a presenting case.

Since delay of revascularization is unlikely to adversely affect outcome, most choose ligament release first, followed by revascularization via stenting or bypass if symptoms persist. Endovascular celiac artery stenting alone without release of the ligament is discouraged because of clinical failure and recurrent stenosis [17].

About 60% to 70% of the patients report symptom relief after surgical treatment. Symptom relief has been reported to be immediate in some instances. In others, it may take up to a few months for the resolution of the pain [18]. The presence of postexertional pain has been shown to be correlated with symptomatic improvement after surgery while the presence of emesis and unprovoked pain preoperatively has been shown to be associated with suboptimal surgical outcomes postoperatively. Patients who have a good response to a diagnostic celiac plexus block preoperatively also seem to report better symptom relief following surgical intervention. The presence of atherosclerotic risk factors may predict poorer outcomes postoperatively [19].

CONCLUSIONS

DS may mimic other medical conditions such as gallbladder diseases, gastritis/peptic ulcer, appendicitis, colorectal malignancy, hepatitis, atherosclerotic diseases etc. That is why DS is a diagnosis of exclusion. This case illustrates pathway to find correct diagnosis and improve management tactic.

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The Authors declare no conflict of interest.

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