Case Report

The Ultrasound Findings in a Rare Case of Nutcracker Syndrome, Wilkie's Syndrome, and Dunbar Syndrome Combination

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Abstract

Vascular compression syndromes represent a group of rare and poorly understood diseases. Dunbar syndrome (DS) is caused by the median arcuate ligament of diaphragm originating lower than normal and causing compression of celiac artery. The Nutcracker is caused by the superior mesenteric artery (SMA) originating from aorta at an acute angle causing a restriction of aortomesenteric space that is traversed by the left renal vein and duodenum; if the compression involves only the left renal vein and becomes symptomatic it is called Nutcracker syndrome; if the symptomatic compression involves only the duodenum it is called Wilkie's syndrome or SMA syndrome. The knowledge of these rare pathologies is essential to reduce the false negatives which still remain very high; it is, therefore, necessary to promote greater knowledge as the lack of diagnosis can be very dangerous for the patient's health. We describe a rare case of a combination of DS, Nutcracker, and SMA or Wilkie's syndrome in a young patient.

Keywords: Cardiovascular abnormalities, color-Doppler ultrasound, Dunbar syndrome, Nutcracker syndrome

Introduction

Vascular compression syndromes represent a group of rare and poorly understood diseases. Dunbar syndrome (DS)[1] also known as Median Arcuate Ligament syndrome,[2] is caused by the median arcuate ligament of diaphragm originating lower than normal and causing celiac artery compression. When the median arcuate ligament insertion is less than normal, during the exhalation phases, with diaphragm elevation, the celiac artery compression occurs [Figure 1a]; these chronic compressions, if they become symptomatic, cause epigastric pain which intensifies with forced exhalation and after meals. In some cases, the stenosis, even if severe, can be asymptomatic thanks to compensation of numerous shunts present between celiac artery and superior mesenteric artery (SMA).[3] There are still controversies about the painful symptoms origin, according to some authors, it is supported by an insufficient postprandial blood flow,[4] while according to others by celiac plexus compression.

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Nutcracker syndrome (NCS)[5] is due to the abnormal course of SMA originating from aorta with an acute angle, <22°, with consequent aortomesenteric space reduction which is crossed by the left renal vein and duodenum [Figure 1b]. In NCS, only the left renal vein is compressed, with consequent flow congestion which manifests itself with increased creatininemia, left lumbar pain, microhematuria, renal insufficiency, varicocele, and, in most severe cases, with thrombosis. Compression can also affect only the duodenum and, if significant, can become symptomatic with nausea, vomiting, and subsequent weight loss, in this case, it is called SMA syndrome (SMAS).[6] We describe a very rare case of NCS and SMAS induced by congenital DS.

Case Report

24-year-old underweight patient suffering from pelvic pain for about 6 months. She underwent pelvic magnetic

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resonance imaging (MRI), ultrasound, and computed tomography (CT). MRI was performed using a 1.5 Tesla device (Signa, GE). The ultrasound examination was performed with a MyLab™ 9 device (Esaote, Genova) with a 3.5 MHz convex probe, by an operator with 20 years of experience. CT was performed with Optima 64 slice device (GE-Healthcare). MRI revealed gonadal plexus congestion [Figure 2a], left renal vein dilatation [Figure 2b], and duodenal compression in aortomesenteric space [Figure 2c]. Gray-scale ultrasound showed significant aortomesenteric distance reduction (3.2 mm) [Figure 3a and b], aortomesenteric angle (AOM) reduction (18°) [Figure 3c], and left renal vein compression with pre and post-stenotic dilatation. Duplex Doppler ultrasound of left renal vein showed: Prestenotic flow congestion with low peak speed velocity (PSV) (5.6 cm/s) [Figure 4a] and poststenotic PSV increase (15.6 cm/s) with flow ratio (FR) of 2.78 (PSV of poststenotic tract/PSV of prestenotic tract); regular resistive index in renal arteries (<0.69); severe stenosis at celiac artery origin with a notable PSV increase in forced expiration (267.8 cm/s) [Figure 4b], less in forced inspiration (238 cm/s) [Figure 4c]; the celiac artery FR, measured in expiratory apnea, was 5.9. Color Doppler ultrasound of gonadal plexus showed varicocele. Results are summarized in Table 1. CT examination after oral contrast medium administration which showed gastric, first and second duodenum dilatation and duodenum stenosis in aortomesenteric space [Figure 5a and b]. After a few days, the patient underwent surgery with laparoscopic ligament release and celiac ganglionectomy which resulted in a rapid regression of epigastric pain. In the following days, she was discharged with a high-calorie diet prescription. After 2 months, the patient had an increase of 6 kg, AOM increase (19°), and PSV reduction in celiac artery (172. 4 cm/s, measured in forced expiration) [Figure 6a and b]. The patient also reported reduction of postprandial vomiting episodes. High-calorie diet continuation and ultrasound periodic follow-up were recommended.

Discussion

In vascular compression syndromes, it is important to know the severity of venous or arterial stenosis, and Doppler ultrasound thanks to the FR measurement provides us with a very accurate estimate of stenosis degree. In the veins, the FR is obtained

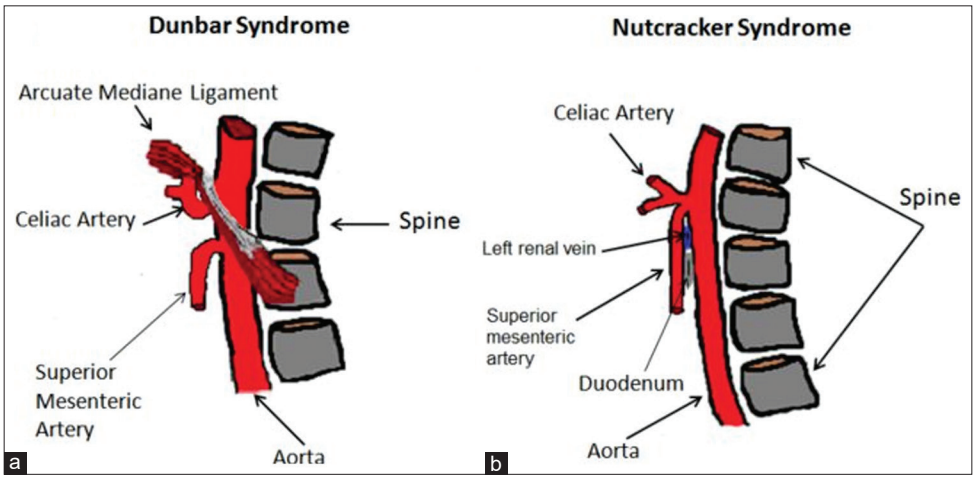

Figure 1: Schematic summary of main anatomical structures involved in Dunbar syndrome and Nutcracker syndrome (NCS). (a) Diagram according to a sagittal plane, showing relationship between celiac artery and aorta in expiratory apnea. (b) Diagram according to a sagittal plane showing relationships between superior mesenteric artery and aorta in NCS

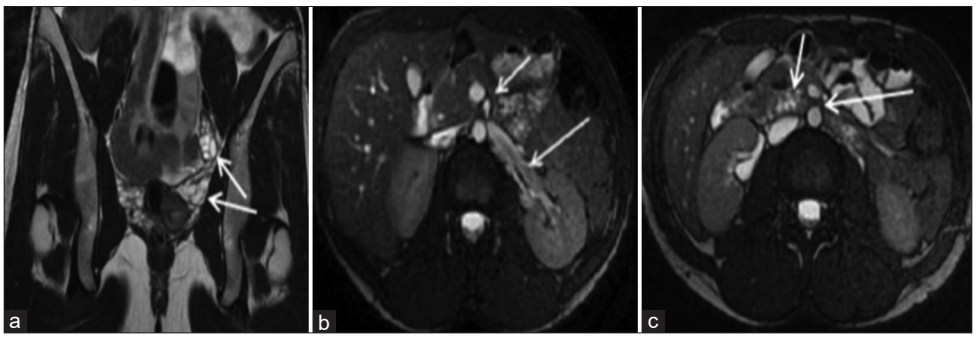

Figure 2: Magnetic resonance imaging. (a) This coronal reconstruction shows varicocele (arrows). (b) In this axial reconstruction we can observe left renal vein congestion (long arrow). superior mesenteric artery (short arrow). (c) This axial reconstruction shows duodenum compression in aortomesenteric space (long arrow). Prestenotic duodenum tract (short arrow)

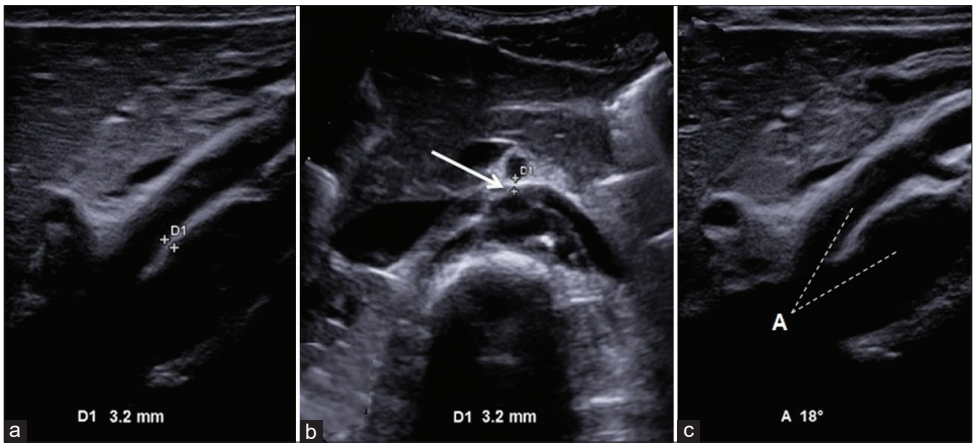

Figure 3: Nutcracker syndrome. Gray-scale ultrasound. (a) In this longitudinal ultrasound scan of abdominal aorta the aortomesenteric distance has been measured. (b) The aortomesenteric distance (arrow) can also be measured with a transversal ultrasound scan of abdominal aorta. (c) This longitudinal scan shows aortomesenteric angle (a) measurement which is obtained by tracing two lines along longitudinal axis of aorta and superior mesenteric artery

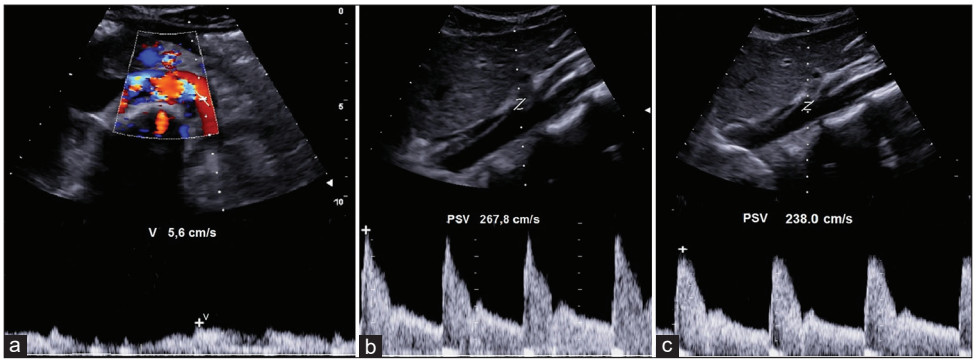

Figure 4: Duplex Doppler ultrasound of abdominal aorta. (a) Left renal vein peak speed velocity (PSV) measurement performed in prestenotic tract shows a severe flow slowing (5.6 cm/s). (b) PSV measurement of celiac artery, performed in expiratory apnea, shows much higher values than in inspiratory apnea phase in stenotic tract (c)

Table 1: Summary of results obtained by ultrasound

	Prestenotic PSV	Poststenotic PSV	FR	CA PSV in expiratory apnea	CA PSV in inspiratory apnea	CA diameter in expiratory apnea	CA diameter in inspiratory apnea
Left renal vein	5.6 cm/s	15.6 cm/s	2.78				
CA			5.9	267.8 cm/s	238 cm/s	1.4 mm	2.2 mm

PSV: Peak speed velocity, CA: Celiac artery, FR: Flow ratio

from the difference between the PSV measured in poststenotic tract and the PSV measured in prestenotic tract: A FR of 2.5 corresponds to a stenosis of 50%,[7] and in most severe cases it requires treatment. In celiac artery, the FR is obtained from difference between PSV measured in stenotic tract and PSV measured in abdominal aorta: A FR of 3: 1 corresponds to a stenosis >70%.[8] Based on a careful analysis of the patient's clinical history, we learned that the patient had suffered from mild intermittent postprandial painful episodes 2 years earlier for which she had not requested controls. Postprandial epigastric pain is typical of DS not of SMAS which causes postprandial nausea and vomiting only; therefore, the weight loss was probably initially induced by celiac artery stenosis causing a progressive reduction of perivascular adipose tissue that caused NCS and SMAS.

For instrumental diagnosis of these syndromes, ultrasound can be used as a first-level examination that allows to localize vascular compressions and to measure the stenosis degree, this method requires integration with CT or alternatively with MRI to exclude other causes of vascular compression (abdominal masses, aneurysms, pseudoaneurysms, etc.) and to verify duodenal compression; in our case, we also used CT due to a patient's claustrophobic crisis that did not allow us to complete the MRI exam. For DS diagnosis it is important to perform CT or MRI scans in expiratory apnea. Measurements of celiac

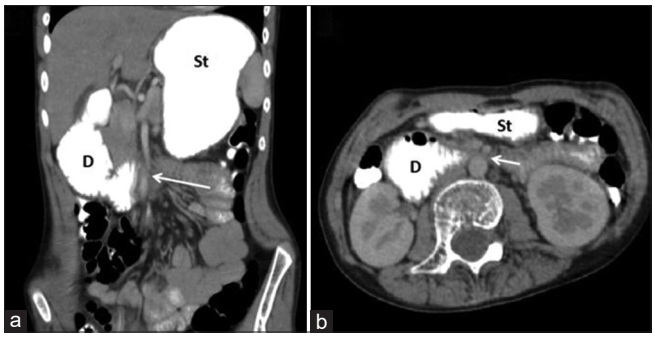

Figure 5: Computed tomography examination performed after oral administration of contrast agent (Gastrografin). (a) This reconstruction according to a coronal plan shows duodenum (d) stomach (St) dilatation due to the narrowing of aortomesenteric space (arrow), as can be better highlighted in axial scan (b), where it is possible to notice the severe distance reduction between superior mesenteric artery and aorta (arrow) in aortomesenteric space. Duodenum (d), Stomach (St)

axis diameter and celiac artery stenosis ratio performed with Doppler ultrasound were in line with CT measurements, in particular, the percentage of celic artery stenosis measured according to the NASCET method was 89%, however, in DS it is preferable, in our opinion, to use the ultrasound method of FR measurement which allows us to highlight the significant differences in flow between the inspiratory and expiratory phases necessary for the diagnosis. CT measurements of the caliber of prestenotic, stenotic, and poststenotic tract of the left renal vein were in agreement with the ultrasound findings. The treatment of NCS and SMAS depends on the symptoms and stenosis severity of renal vein and duodenal and must initially be based on a high-calorie diet aimed at restoring perivascular adipose tissue; if the diet fails, NCS can be treated with left renal vein stenting;[9] allows the restoration of regular flow and AOM increase with consequent duodenal decompression; in cases of stenting failure, the surgery can be used.[10] Asymptomatic patients with mild vascular compression (<50%) (Nutcracker phenomenon),[11] should only undergo periodic ultrasound monitoring. The surgical treatment of SMAS consists in the resection of first duodenal loop and retro-vascular duodenum with anastomosis between duodenum and second duodenal loop which are brought anteriorly; allows excellent results but is more invasive than the previous ones and should be reserved, in our opinion, for cases of high-calorie dietary failure. The elective treatment of DS is surgical and consists of median arcuate ligament resection and gangliotomy.[12] Celiac artery stenting is performed only in case of relapse (7%) for complications related to the stent obstruction and migration.[13] In our case, the epigastric pain episodes quickly ceased after surgery and the patient responded well to the high-calorie diet with significant weight gain after 2 months (+6 kg). Surgical treatment allows the best results in DS, while for NCS and SMAS the best results are guaranteed by high-calorie diet. The number of misdiagnoses in people with vascular compression syndromes still remains very high, it is therefore necessary to promote greater knowledge as the lack of diagnosis can be very dangerous for patients health.

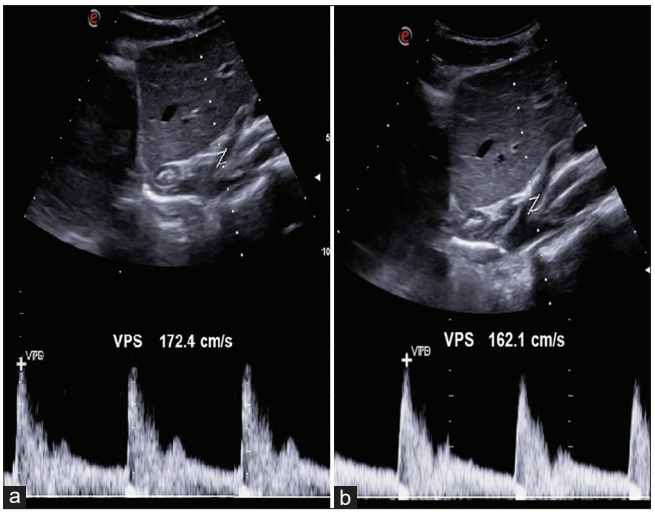

Figure 6: The Doppler ultrasound control performed 2 months after surgery showed a significant reduction in peak speed velocity in the celiac artery in both expiratory apnea (a) and in both inspiratory apnea (b)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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