Case report

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Double nutcracker syndrome in a patient with circumaortic venous ring: a rare case report

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Abstract

Aim of the study: In this article, we describe a rare case of the nutcracker syndrome caused by combined compression of the left anteroaortic and retroaortic renal veins. **Case description:** A 42-year-old woman presented with microhematuria and left flank pain. The patient underwent computed tomography and Doppler ultrasound which showed the left renal veins with anteroaortic and retroaortic courses, with signs of compression. Compression of the anteroaortic renal vein was caused by a narrowing of the aortomesenteric space, whereas compression of the retroaortic renal vein was caused by a narrowing of the aortovertebral space. **Conclusions:** NCS is a rare disease, poorly understood and difficult to diagnose mainly due to the non-specificity of symptoms. Imaging is essential for diagnosis, and the combination of ultrasound and computed tomography allows for better classification of the disease. Increased disclosure of these cases can significantly contribute to a reduction of false negatives.

Introduction

Vascular compression syndromes are very rare diseases caused by compression of vascular structures by contiguous vascular, musculotendinous, cystic, bone or neoplastic masses. They are very difficult to diagnose because of their rarity, lack of knowledge and non-specificity of symptoms. The nutcracker syndrome is caused by symptomatic compression of the left renal vein (LRV) (Fig. 1). When the LRV has an anteroaortic course, it is known as anterior NCS (ANCS)[1] and can be acquired or, much less commonly, congenital; in the former case, it is caused by rapid weight loss and is,

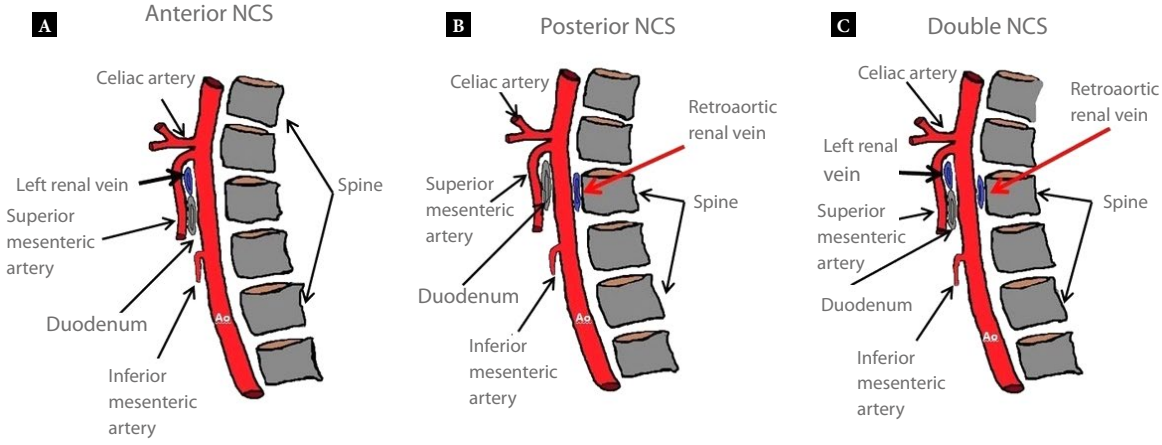

Fig. 1. *Diagram showing the anatomical relationships in a patient with NCS. Ao – abdominal aorta*

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in fact, very frequent in anorexic patients following a significant reduction of perivascular adipose tissue which causes aortomesenteric angle (AMA) with consequent renal vein compression and, in some cases, also duodenal compression (Superior Mesenteric Artery Syndrome)[2] which run in the aortomesenteric space. When the vein has a retroaortic course, the symptomatic compression can also be congenital or acquired and is generally caused by vertebral osteophytosis leading to aortovertebral space narrowing; the condition is referred to as the posterior nutcracker syndrome (PNCS)[3]. In very rare cases, the patients shows double left renal vein with anteroaortic and retroaortic course; this anatomical variant is known as "circumaortic venous ring" or "renal collar"[4]; in these patients, compression of either one or both renal veins may occur. There are, therefore, five variants of NCS: ANCS and PNCS in patients with only one left renal vein, while in patients with double renal vein compression can affect only the anteroaortic vein, the retroaortic vein or both veins. Renal vein compression in asymptomatic patients is mild and often discovered incidentally during ultrasound, CT or MRI examinations done for other reasons; these cases are known as the "nutcracker phenomenon"[5]. The symptomatology of NCS is consequent to the congestion of flow in the left renal vein and manifests itself clinically with low back pain, microhematuria, proteinuria, venous thrombosis, and varicocele. Imaging is essential for diagnosis and includes ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and intravascular ultrasound (ICU). Ultrasonography represents the first-level examination and allows the localization of compressions with great accuracy, also providing an estimate of the degree of stenosis based on the flow ratio (FR) measurement: poststenotic peak systolic velocity (PSV) / prestenotic PSV[6]. Management of these patients should be based on the severity of compressions and symptoms[7]. ANCS therapy can be conservative, with a high-calorie diet[8], vascular stenting[9], or surgery, while PNCS therapy can be endovascular or surgical. We describe a case of double LRV compression in a patient with circumaortic venous ring.

Materials and methods

A 42-year-old woman presented with microhematuria and pain in the left flank. The patient underwent Computed Tomography (CT) and Doppler US (DU). CT examination was performed with Optima 64 slice (GE-Healthcare) device, and ultrasound examination with MyLab Nine (Esaote Biomedica) device, using 3.5 MhZ and 7.5 MhZ probes. CT showed retroaortic (Fig. 2A) and anteroaortic (Fig. 2B) LRV compressions and pelvic varicocele (Fig. 2C, D). The sagittal plane view revealed aortomesenteric distance (AMD) of 4.2 mm and aortomesenteric angle (AMA) of 18 degrees. Subsequently, the patient underwent an ultrasound examination which showed: AMD reduction (4.2 mm) (Fig. 3A); AMA reduction (18 degrees) (Fig. 3B); anterior LRV and retro LRV compressions, and pelvic varicocele (Fig. 3C) (Video 1, Video 2). Color Doppler US showed the prestenotic and poststenotic tracts of the retroaortic renal vein (Fig. 3D) and duplex Doppler US revealed low PSV (8 cm/s) in the prestenotic tract (Fig. 3E) and PSV increase (22 cm/s) in the poststenotic tract (Fig. 3F); low PSV in the prestenotic tract of anterior LRV (18.8 cm/s) and PSV increase in the poststenotic tract (50.6 cm/sec); LRV caliber reduction in the stenotic tract (3.5 mm), prestenotic (8 mm) and poststenotic tract (7 mm) of anterior LRV; LRV caliber reduction in the stenotic tract (3.5 mm), prestenotic (8 mm) and poststenotic tract (7 mm) of posterior LRV. The flow ratio (FR) was 2.69 in anterior LRV and 2.75 in retroaortic LRV (the results are summarized in Tab. 1). Ultrasound was performed by an operator with twenty years of experience. We decided to recommend a high-calorie liquid diet for perivascular adipose tissue restoration, low-dose anticoagulant therapy, and monitoring by US. At the first follow-up evaluation after one month, ultrasound showed AMA (20°) and AMD increase (9 mm), FR reduction (2.4) in anterior LRV, and resolution of pain in the left side. We observed no changes in the retroaortic renal vein. The ultrasound was performed by an operator with twenty years of experience. Ethical approval: "All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards." "For this type of study formal consent is not required." Written informed consent was obtained from the patients for publication of this Case Report and any accompanying images and clips. A copy of the written consent is available for review by the Editor of this journal.

Discussion

The ultrasound diagnosis of ANCS is based on the aortomesenteric angle measurement and aortomesenteric distance measured at 1 cm from the origin. There are cut-off values below which the syndrome manifests itself, i.e. 22 degrees for the aortomesenteric angle and 8 mm for the aortomesenteric distance. On CT examination, the narrowing of the aortomesenteric angle assumes a characteristic appearance known as a "beak sign" typical of ANCS, evident in sagittal images with the superior mesenteric artery originating at an acute angle to the aorta. Asymptomatic patients with the "nutcracker phenomenon" are not treated, but it is advisable to perform an ultrasound check annually to prevent the aggregation of stenosis. Intravascular ultrasound (IVUS)[10] should be considered, as it has demonstrated superior characterization of intraluminal content to angiography and is increasingly used by experts, but limited by equipment availability. In our opinion, significant asymptomatic stenosis, greater than 50%, also requires long-term prophylaxis with low-dose anticoagulant drugs. PNCS is almost always caused by osteophytosis of the spine or, more uncommonly, by aneurysms of the abdominal aorta which cause narrowing of the aortomesenteric space; the management of these patients is more complex because the aortovertebral space is often too narrow to allow endovascular stenting that guarantees a good long-term seal; therefore, anticoagulant pharmacological treatment and surgical therapy are often the only possible approaches. The "circumaortic venous ring" or "renal collar" is a rare anatomical variant, and compression of both renal veins is very rare indeed; in such cases, the treatment is the same as for ANCS and PNCS. Choosing the most appropriate imaging method is not difficult because both ultrasound and CT or MRI make it possible to reach the diagnosis easily. However, the combination of ultrasound and CT offers greater guarantees for the correct stenosis measurement and disease management. The diagnostic suspicion is induced in most cases by renal symptoms, but in some cases it is also suggested by the presence of pelvic varicocele due to flow congestion in the left gonadal vein. In our case, the patient underwent CT for suspected ureteral lithiasis, during which the varicocele and compressions of the two left renal veins were discovered. Subsequently, the ultrasound, in addition to confirming the findings obtained by CT, made it possible to measure the degree of stenosis of renal veins, providing information that proved to be im-

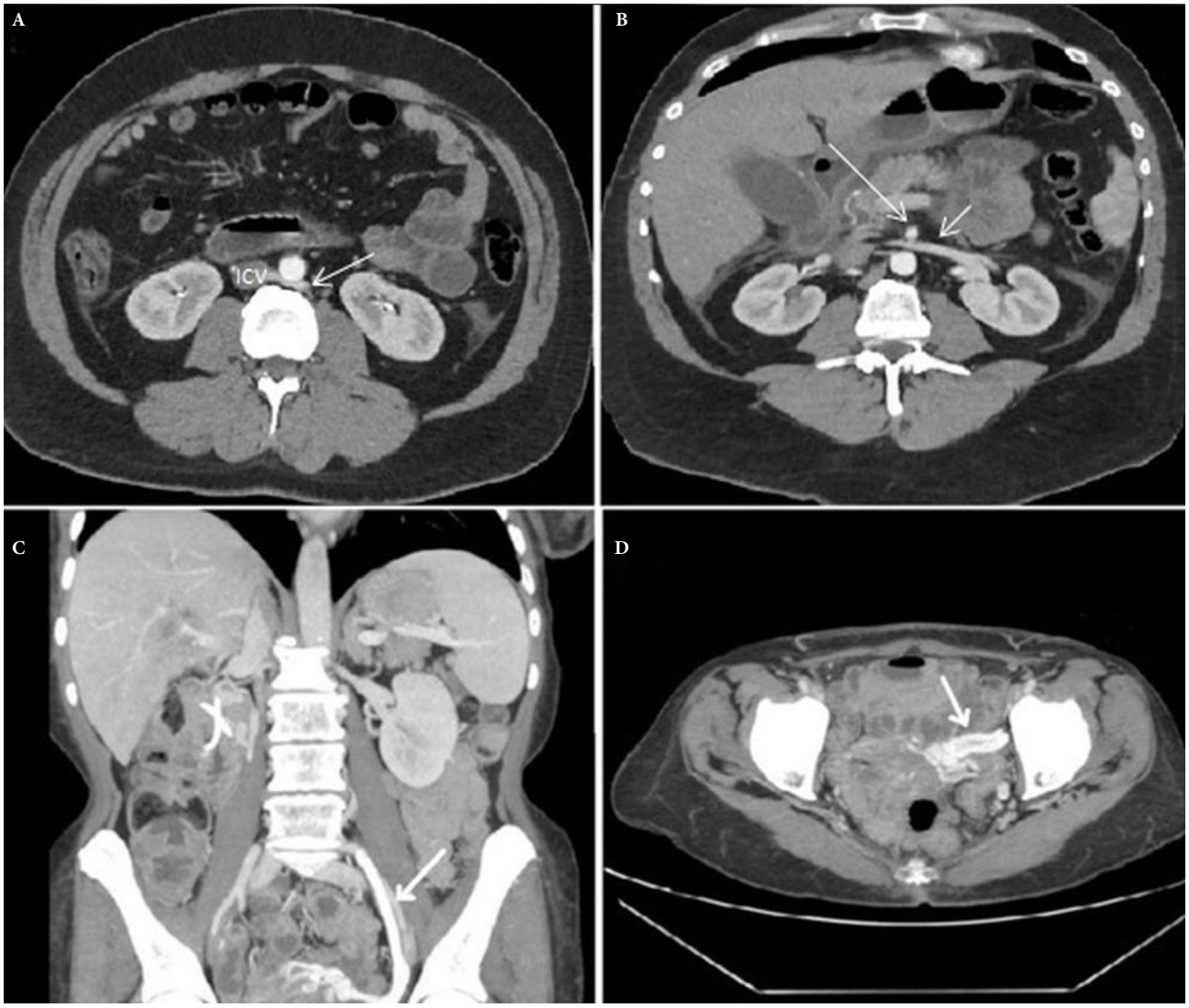

Fig. 2. *CT of double NCS.* **A.** *In this image, the retroaortic renal vein (arrow) appears compressed between the aorta and spine. ICV: Inferior cava vein.* **B.** *This image shows the anteroaortic renal vein (short arrow) which appears compressed in the space between the aorta and superior mesenteric artery (long arrow).* **C.** *This coronal view shows gonadal vein dilatation (arrow). d: This axial view shows varicosities of the gonadal plexus (arrows)*

portant for disease management. In fact, unlike the CT or MRI, the ultrasound is suitable for comparing the pre- and poststenotic flow and, thanks to the FR, provide an estimate of the degree of stenosis. Knowledge of the LRV stenosis degree is very important for the management of patients with NCS, and ultrasound provides a very reliable estimate obtained from the ratio between poststenotic PSV and prestenotic PSV (flow ratio, FR): a stenosis of 50% corresponds to a FR of 2.5 and does not require treatment but only periodic ultrasound follow-up (6 months). When the stenosis is greater than 70%, it requires treatment, and one must choose a modality that guarantees the best long-term outcome. A high-calorie diet aimed at restoring the normal perivascular adipose tissue layer should be the first approach to resolve compression. Where it fails, endovascular stenting of the LRV can be a good alternative to surgery because, in addition to ensuring patency of the renal vein, it causes a significant increase in the aortomesenteric distance, which also helps to prevent the possible onset of the superior mesenteric artery syndrome. Surgical treatment is invasive and should be considered, in our judgment, only after previous treatment modalities have failed. In our case, since the anterior LRV stenosis is less than 70%, a conservative treatment option was chosen, with a high-calorie diet combined with low doses of anticoagulant drugs, periodic laboratory checks (glomerular filtrate, creatinine, urinalysis) and six-monthly ultrasound scans. The aortovertebral distance being very small (2 mm), an intervention on the retroaortic gonadal vein was not an option.

Tab. 1. Summary of results obtained by Doppler US

	Flow ratio	Prestenotic PSV	Poststenotic PSV	Flow ratio after 1 month
Anterior LRV	2.69	18.8 cm/s	50.6 cm/s	2.4
Posterior LRV	2.75	8 cm/s	22 cm/s	2.75
PSV – peak speed velocity; LRV – left renal vein				

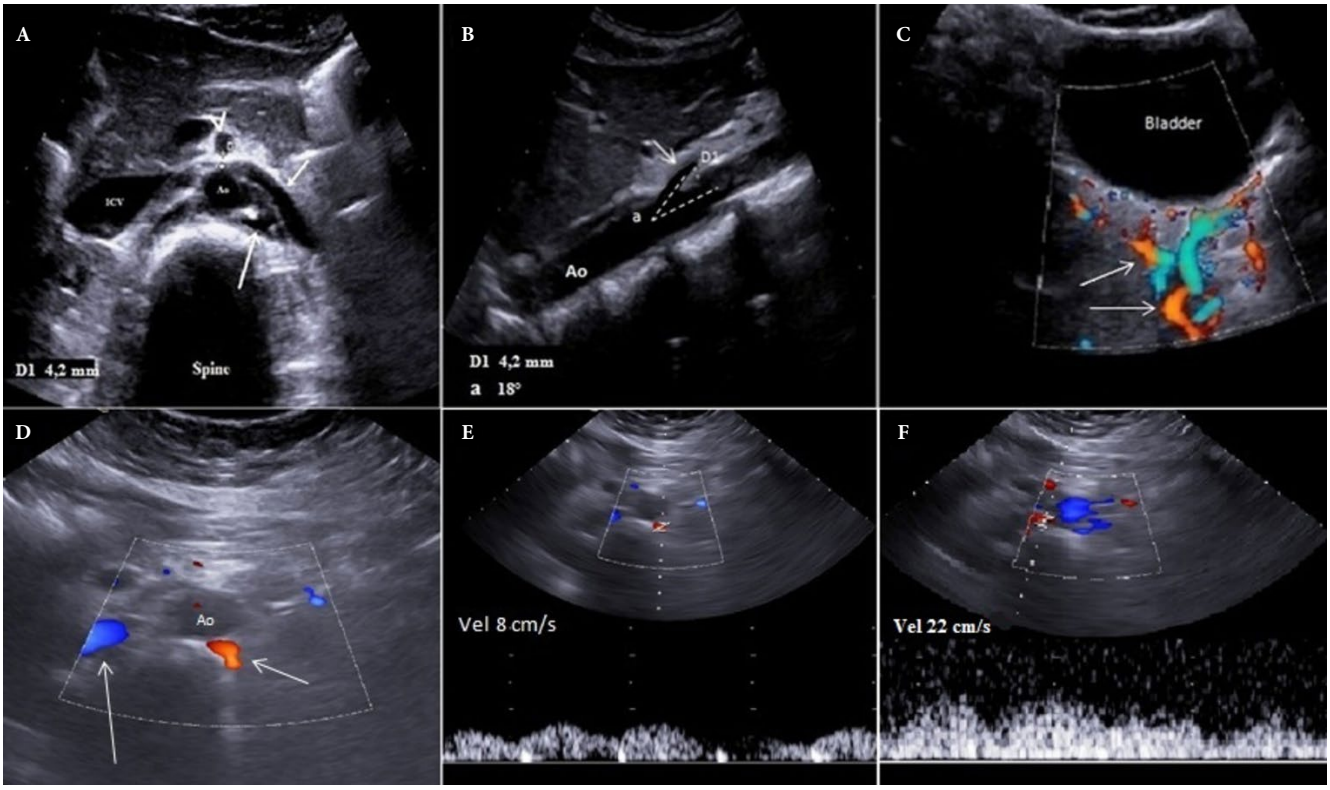

Fig. 3. *Grayscale US of the abdominal aorta. Measurement of aortomesenteric distance and aortomesenteric angle.* **A.** *Transverse scan of the abdominal aorta shows a narrowing of the aortomesenteric space (D1) measuring 4.2 mm. Superior mesenteric artery (arrow head). Left anteroaortic renal vein (short arrow). Left retroaortic renal vein (long arrow). Abdominal aorta (Ao). Inferior cava vein (ICV).* **B.** *Longitudinal scan of the abdominal aorta shows a narrowing of the aortomesenteric angle (18°) and aortomesenteric distance (D1) measuring 4.2 mm.* **C.** *Pelvic ultrasound scans show gonadal plexus varicosities (arrows).* **D.** *Color Doppler US shows prestenotic tract of the retroaortic renal vein (short arrow) and poststenotic tract of the anteroaortic renal vein (long arrow). Ao: Abdominal aorta.* **E.** *Duplex Doppler US measure of the prestenotic flow of retroaortic renal vein.* **F.** *Duplex Doppler US measure of the poststenotic flow of retroaortic renal vein*

Conclusions

NCS is a rare disease, little known and difficult to diagnose, mainly due to the non-specificity of symptoms. Imaging is essential for the diagnosis, and combination of US and CT allows a better classification of the disease. Management of these patients should take into account the severity of symptoms and the degree of stenosis. The best therapeutic approach seems to be conservative, with a high-calorie diet, for anteroaortic renal vein compression; and surgical for retroaortic renal vein compression. Endovascular stenting offers more assurance for anterior compression and should be considered when the high-calorie diet fails. Increased disclosure of this pathology can significantly contribute to a reduction of false negatives. Failure to diagnose may expose patients to serious health risks.

Conflict of interest

The authors declare no conflict of interest. All authors declare that no competing financial interests exist.

Author contributions

Original concept of study: RF. Writing of manuscript: PVF, LN. Analysis and interpretation of data: RF, PVF, LN. Final acceptation of manuscript: RF. Collection, recording and/or compilation of data: RF, PVF, LN. Critical review of manuscript: RF, AB

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