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Nutcracker Syndrome ultrasonography imaging features: a case report

Características ultrassonográficas da Síndrome de Quebra-Nozes: um relato de caso

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Abstract

The Nutcracker Syndrome (NCS), also known as left renal vein (LRV) entrapment syndrome, is caused by LRV stenosis due to the angle narrowing between the superior mesenteric artery (SMA) and the aorta. The stenosis leads to dilatation of the LRV distal portion. Patients may be asymptomatic or experience hematuria, proteinuria, flank pain, pelvic congestion in women, and varicocele in men. Doppler ultrasonography (US) is an excellent diagnostic tool for NCS as it facilitates assessing LRV anatomy and physiology. This case report aimed to describe the imaging features of NCS in Doppler US and to discuss the diverse features of NCS in various imaging studies.

Keywords: Renal Nutcracker Syndrome; Renal vein entrapment syndrome; Renal veins; Ultrasonography; Doppler ultrasonography.

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Resumo

A Síndrome de Quebra-Nozes (NS), também conhecida como síndrome de aprisionamento da veia renal esquerda (LRV), é causada pela estenose da LRV em consequência do estreitamento do ângulo entre a artéria mesentérica superior (SMA) e a aorta. Tal estenose resulta na dilatação da porção distal da LRV. Os pacientes podem ser assintomáticos ou apresentar hematúria, proteinúria, e dor no flanco, além de congestão pélvica nas mulheres ou varicocele nos homens. A ultrassonografia com Doppler é uma excelente ferramenta para nortear o diagnóstico, pois permite avaliar a anatomia e a fisiologia da LRV. Este relato tem como objetivo descrever as características de imagem de NS em ultrassonografia e discutir características de imagem em outras modalidades de exames de imagem.

Palavras-chave: Síndrome do Quebra-Nozes renal; Veias renais; Ultrassonografia; Diagnóstico por ultrassom.

INTRODUCTION

The Nutcracker Syndrome (NCS), also known as left renal vein (LRV) entrapment syndrome, was initially documented by Schepper in 1972 while elucidating the context of the LRV stenosis resulting from a narrow-angle (< 35 to 39 degrees) between the superior mesenteric artery (SMA) and the aorta.[1] This stenosis dilates the LRV distal portion.[2] The LRV entrapment syndrome has two categories: anterior and posterior NCS. Anterior NCS involves the compression of a typically positioned LRV by the SMA and aorta, constituting most NCS cases.[3] Posterior NCS, related to a retroaortic LRV, is typically associated with limited space between the aorta and the vertebral column.[3]

Patients with NCS may either present no symptoms or experience hematuria, proteinuria, flank pain, pelvic congestion in women, and varicocele in men.[4] NCS prevalence remains uncertain due to the variability in symptomatic presentation.[2] This syndrome can affect patients of any age, ranging from childhood to middle age, with a peak prevalence observed in the second or third decades. Also, NCS prevalence is not associated with sex.[3]

Several methods have been employed for diagnosing NCS, including Doppler ultrasonography (US), computed tomography (CT) angiography, magnetic resonance imaging (MRI) angiography, and venography.[3] Real-time Doppler US is recommended as the initial diagnostic test for suspected NCS due to its elevated sensitivity and specificity.[5] Consequently, this case report aimed to delineate the imaging features of NCS in Doppler US and elaborate on the varied NCS features observed in different imaging studies. The objective was to provide physicians with a comprehensive understanding of this uncommon syndrome.

CASE REPORT

A 23-year-old Caucasian male reported lumbar pain and pelvic discomfort with painful sensitivity to touch and edema in the left testicle. The patient was later subjected to Doppler US and urinalysis for approximately eight months, with signs and symptoms suggesting varicocele. The previous medical history revealed no significant findings.

The initial imaging study was an abdominal Doppler US, which revealed in the cross-sectional view that the LRV was compressed by the SMA and aorta, associated with increased venous flow within the LRV varicocele region (Figures 1 and 2). Consequently, the hypothesis was that the varicocele resulted from elevated venous pressure in the LRV, constrained between the SMA and aorta.

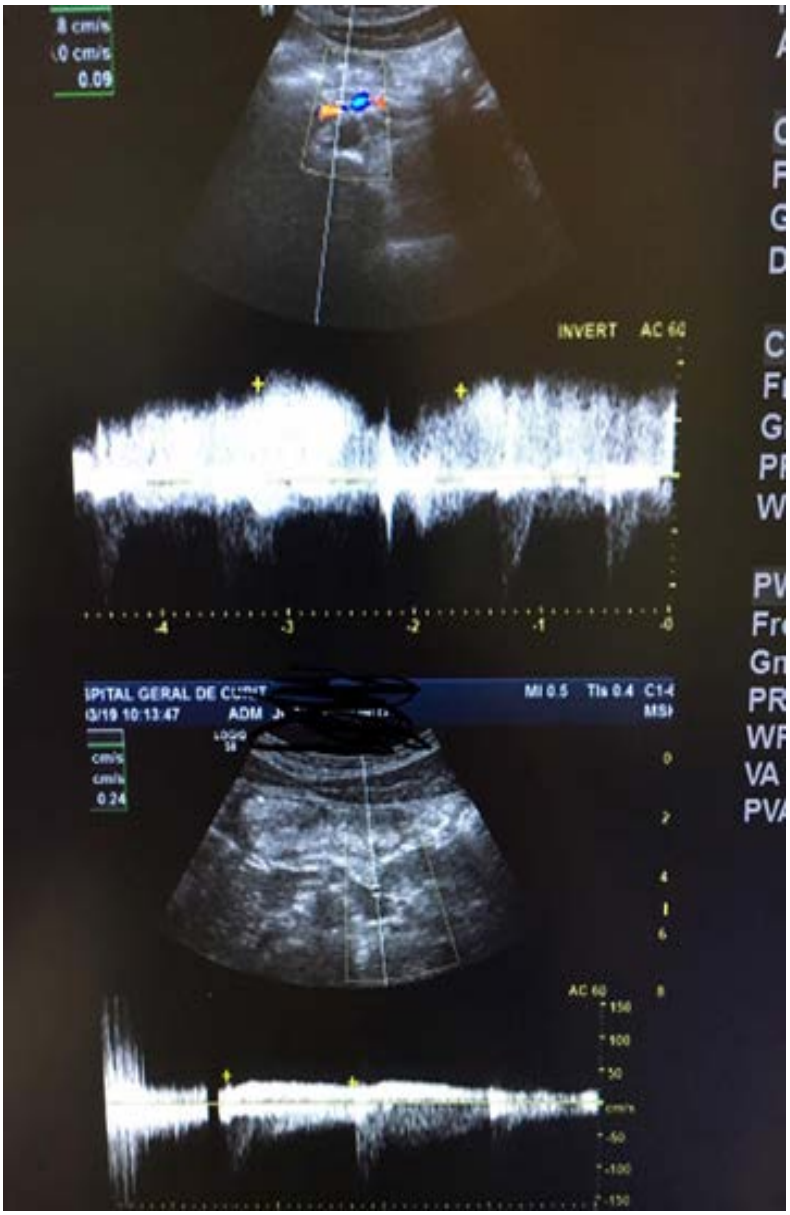

Figure 1. Doppler ultrasonography in cross-sectional view, showing the left renal vein compressed by the superior mesenteric artery and the aorta, with an increased venous flow in the left renal vein.

The Doppler US examination exhibited an increase in the caliber of the pampiniform plexus in the scrotum, characterizing a grade 3 varicocele. Moreover, a fivefold increase in the velocity of venous flow between the inferior vena cava was noticed, suggesting NCS. Additionally, this case was classified as anterior NCS, since the compression was positioned in LRV by the SMA and aorta.

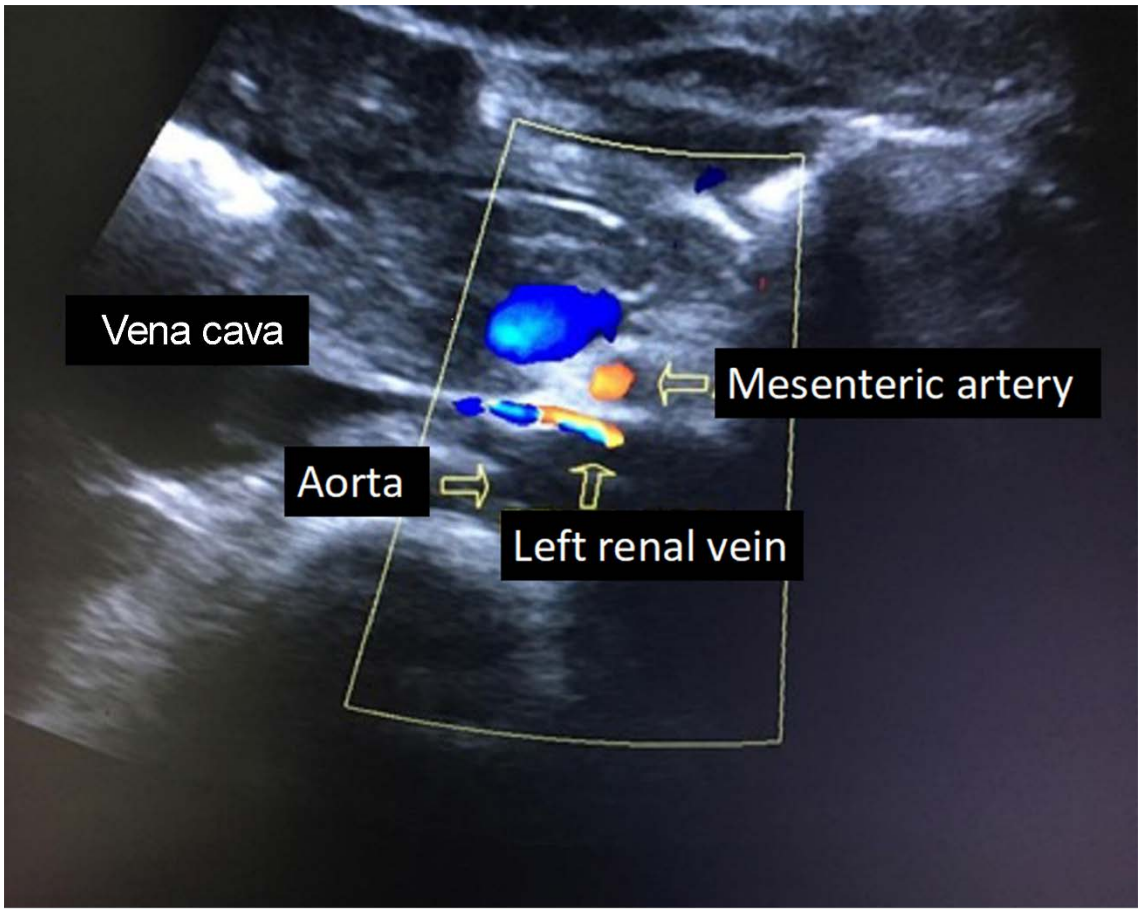

Figure 2. Detail of the Doppler ultrasonography in cross-sectional view, showing the left renal vein compressed by the superior mesenteric artery, aorta, and other adjacent anatomic structures.

The urinalysis results showed increased levels of hematuria and mild proteinuria, leading to the NCS diagnosis. An open surgery approach was chosen for the entrapment correction, and the patient had a great post-surgical recovery without any intercurrence.

The research ethics committee of the Medical University Hospital approved this study (n° 3.691.683).

DISCUSSION

The main symptoms of NCS include pelvic pain, lumbar pain, hematuria, and gonadal varices, such as varicocele,[3] often suggesting pelvic and renal congestion. In this case, a grade 3

varicocele indicating the most severe form of venous dilatation, was also observed and correlated with pain on palpation of the left testicle, as reported by the patient. The elevated blood pressure in the stenosed LRV may result in venous reflux, venous hypertension, and subsequent emergence of varices between the renal pelvis and ureter.[2]

Hematuria was another manifestation observed in this report. When combined with proteinuria, hematuria might indicate NCS. This manifestation originates from the rupture of the thin-walled septum between the small veins and the collecting system of the renal fornix,[6] as the pressure between the LRV and the inferior vena cava can rise to 3 mmHg or more (normal pressure = 1 mmHg).[7] Moreover, mild subclinical immune injury has the potential to induce orthostatic proteinuria, affecting the angiotensin II and norepinephrine release,[8] further sustaining NCS.

Although venography was the main imaging study for this condition, the Doppler US performed met key diagnostic criteria for NCS.[5] The grade 3 varicocele was characterized by a marked increase in the caliber of the pampiniform plexus within the scrotum. Additionally, a five-fold elevation in the peak velocity of venous flow between the LRV and the inferior vena cava was observed, indicating a venous gradient of 4 ≥ 3 mmHg, which aligns with the recognized diagnostic criterion for NCS. Quantitative assessments also showed a peak post-stenotic velocity in the LRV exceeding 100 cm/s as it passed by the SMA compared to the renal hilum, confirming a diagnosis consistent with NCS.

Venography with pressure gradient measurement is considered the gold standard for NCS diagnosis, providing the most reliable diagnostic confirmation. Doppler US is the preferred imaging modality since it avoids radiation exposure and facilitates assessing LRV both anatomical- and physiologically.[5] This exam requires six to eight hours of fasting and is conducted with the patient in an upright position, which enhances the visibility of the stenosis. Diagnostic evaluation for NCS includes measuring the peak systolic velocity in the compressed LRV and comparing it with the hilar peak systolic velocity, along with the LRV stenosis observation.[5]

Although CT and MRI angiography studies may indicate NCS, these modalities are complementary.[9,10] Contrast-enhanced CT imaging findings for NCS are not definitively established. However, indicators such as the beak sign and angle, LRV diameter ratio, and aortomesenteric angle may suggest NCS.[9] In MRI angiography, an SMA angle lower than 36.8 degrees and other signs are highly indicative of NCS.[10]

Angiography techniques, such as CT and MRI angiography studies, are useful for assessing vascular abnormalities associated with NCS. CT angiography can provide detailed images of the renal veins and surrounding structures, revealing compression or stenosis. On the other hand, MRI angiography offers the advantage of being non-invasive and does not require ionizing radiation, making it particularly useful for patients who need repeated evaluations.

The range of conditions considered in the differential diagnosis for NCS includes those with similar clinical manifestations or with the potential for LRV entrapment diagnosis. Those include retroperitoneal neoplasms, lymphadenopathy, pancreatic tumors, and even varicocele itself[11]. Varicoceles are characterized by the dilatation of pampiniform plexus veins and can lead to symptoms similar to those of NCS. The diagnosis of varicocele involves a physical examination,[11] complemented by imaging studies, such as the Doppler US, which was performed in this report.

NCS can also be caused by aortic dissection with duodenal obstruction attributable to SMA syndrome, which was not considered in this case since the medical history of the patient did not include any previous surgery. Additionally, NCS may also be associated with Henoch-Schönlein purpura, immunoglobulin A and membranous nephropathy, and idiopathic hypercalciuria with nephrolithiasis.[12] When flank pain and hematuria are present, additional considerations should include nephrolithiasis, pyelonephritis, or renal cell carcinoma.[13]

Various treatment modalities for NCS are available, and choosing one depends on the severity of clinical symptoms.[3,14] Conservative treatment is often recommended for patients aged under 18 years, especially those with minimal symptoms. This approach considers the potential for physical development, increased fat, and fibrous tissue at the SMA origin to relieve LRV entrapment. The development of collaterals may also help to relieve hypertension. Additionally, angiotensin-converting enzyme inhibitors may be prescribed, with ongoing follow-up.[15]

In adults, surgical intervention is often necessary, especially when symptoms are severe or persist beyond six months of conservative therapy.[3] Diverse surgical techniques have been described, including open, endovascular, and laparoscopic surgeries.[3,14] An open surgery approach was chosen in this report because the patient was healthy and seeking a treatment option with minimal risk of future complications.

In summary, NCS can manifest as either asymptomatic or symptomatic entrapment of the LRV due to compression from the SMA and aorta. Doppler ultrasound is highlighted as an imaging study for NCS diagnosis, facilitating a comprehensive assessment of the anatomy and physiology of LRV.

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CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

AUTHOR CONTRIBUTIONS

YS: Conceptualization, Definition of intellectual content, Data acquisition, Data analysis,

Writing – review and editing, **MRSA, NOM, AVK, ITR, and PCAW:** Data analysis, Writing – review and editing, **LM:** Conceptualization, design, definition of intellectual content, literature search, manuscript preparation, Writing – review and editing. All authors read and agreed with the final version of the manuscript.

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