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Nutcracker syndrome: a case report

Síndrome de Quebra-Nozes: relato de caso

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ABSTRACT: Nutcracker syndrome is a rare disease of varied etiology and clinical presentation. It is described that Computed Tomography scan or Magnetic Resonance Imaging, followed by Doppler ultrasonography or venography for confirmation, are the diagnostic methods of choice. We report the case of a 35-year-old female patient who presented with macroscopic hematuria, fever and lumbar pain. She was managed with open surgical approach, the standard of care by many experts, with a double shunt performed: one between the left renal vein and the distal inferior vena cava and another between the left and right gonadal veins.

Keywords: Renal nutcracker syndrome; Hematuria; Renal veins.

RESUMO: A síndrome do Quebra-nozes é uma doença rara, de variada etiologia e apresentação clínica. É descrito que a tomografia computadorizada ou a ressonância magnética, seguidas por ultrassonografia Doppler ou venografia para confirmação, são os métodos diagnósticos de escolha. Relatamos o caso de uma paciente de 35 anos que apresentou hematúria macroscópica, febre e dor lombar. Foi tratada com abordagem cirúrgica aberta, defendida como padrão-ouro por muitos especialistas, com duplo shunt realizado: um entre a veia renal esquerda e a veia cava inferior distal e outro entre as veias gonadais direita e esquerda.

Palavras-chave: Síndrome do Quebra-Nozes; Hematúria; Veias renais.

INTRODUCTION

Nutcracker syndrome (NCS) is a condition caused by compression of the left renal vein (LRV), most often, between abdominal aorta and superior mesenteric artery (SMA)[1,2]. It may cause many symptoms in both children and adults, such as intermittent hematuria, proteinuria, flank pain, pelvic congestion in females, and varicocele in male patients. Diagnosis is challenging and is generally made after exclusion of other more common causes.

It is a rare entity, with few cases described, and no concrete data exist about the real prevalence.

CASE REPORT

A 35-year-old woman presented to the emergency department with a 20-days history of macroscopic hematuria (Figure 1), fever and lumbar pain. She denied any other symptoms. Her medical history included the same symptoms 5-years ago treated with ciprofloxacin and clavulin. Physical examination revealed right flank pain to deep palpation and the Giordano's sign was positive. Hematuria was detected in the urine test with a normal sediment, and urine cultures were negative.

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Figure 1. Macroscopic hematuria

Renal ultrasound (US) showed cortical irregularity in the right kidney with intravesical clot. The patient of spontaneous reflux.

underwent a Computed Tomography (CT) that showed LRV stenosis due to compression at the point where the LRV crossed between the aorta and the SMA, with dilatation of the upstream left renal and gonadal veins, leading to bilateral pelvic venous complex engorgement (Figures 2I, 2II and 3). The US doppler showed a hyperkinetic flow at the origin of the SMA, with a peak systolic velocity = 489 cm/s, a final diastolic velocity of 112 cm/s, indicating stenosis > 70%. The calculated aortomesenteric angle was 24º. The LRV showed narrowing in its aorto-mesenteric transition, with a smaller caliber of 1.3 mm, with a pre-stenotic segment with a larger caliber of 10.6 mm. Diameter ratio = 8.1. The flow velocity at the site of the stenosis was not possible due to the thin gauge; in the ectasied segment it measured 34.4 cm/s. Finally, the left gonadal vein was dilated, measuring 7.5 mm in diameter, with the presence

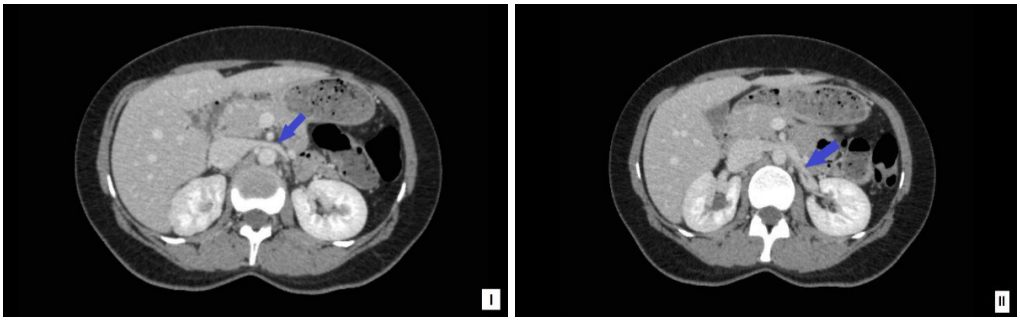

Figure 2 - I. CT showing the left renal vein (arrow) being compressed between the superior mesenteric artery and the aorta in an axial section; **II.** Dilated left renal vein (arrow).

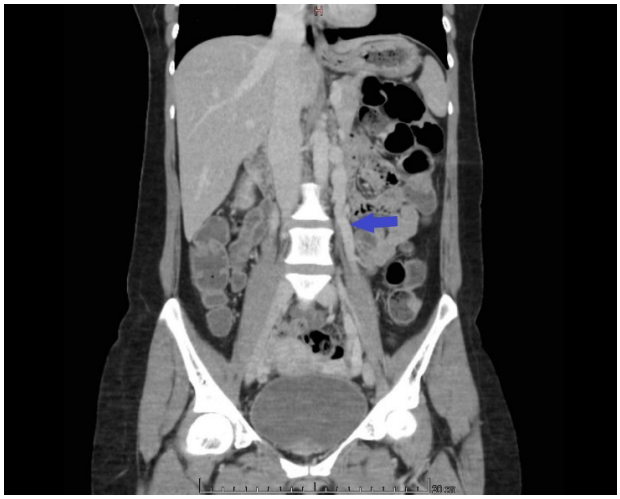

Figure 3. Left gonadal vein dilated on coronal view (arrow).

The final diagnosis was Nutcracker Syndrome. Then, the patient was referred for surgery. A median supra and infraumbilical incision were made, diaeresis by planes and access to the abdominal cavity. A terminolateral anastomosis of the LRV was performed in the inferior vena cava (IVC), 2 cm above the commom iliac vein, and another anastomosis between the left and right gonadal veins (Figure 4). Thereby, a good decompressive effect was achieved.

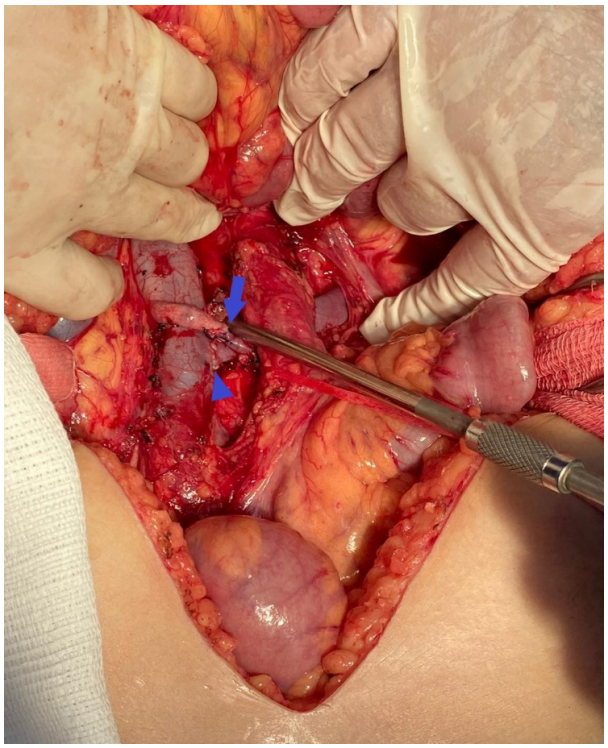

Figure 4. A double shunt was performed. One between the LRV and the distal IVC (arrowhead) and another between the left and right gonadal vein (arrow).

DISCUSSION

In 1950, *El-Sadr and Mina* named the term 'nutcracker phenomenon' (NCP) as the anatomic compression of the LRV that can occur between SMA and the aorta[1,2]. This phenomenon is characterized by impeded outflow from the LRV into the inferior vena cava due to extrinsic compression. A rarer condition is posterior nutcracker, when the LRV is compressed between the aorta and the vertebral body. The term 'nutcracker syndrome', first described by *De Schepper* in 1972, is preferred over phenomenon if the vascular compression becomes symptomatic[2,3].

The exact prevalence of NCS is unknown, partly because of an absence of definitive diagnostic criteria and partly because of the variability in symptomatic presentation[4]. Most symptomatic patients are in their second and third decades of life and a second peak of NCS occurs in middle-aged women[5].

Most common etiology is the angle between the SMA and abdominal aorta less than 45°, causing LRV compression. Less common pathologies and conditions leading to NCS as a result of compression of the LRV are: pancreatic neoplasms, para-aortic lymphadenopathy, retroperitoneal tumor, abdominal aortic aneurysm, lordosis, left renal ptosis, reduced retroperitoneal and mesenteric fat and pregnancy with a gravid uterus compressing the renal vasculature[6,7]. Lower body mass index (BMI) also correlates positively with NCP, more likely after weight loss. Based on analogy with the SMA syndrome, a decrease in retroperitoneal fat is believed to reduce the aortomesenteric angle and cause NCP[8].

The clinical presentation ranges from asymptomatic, in most cases, to episodes of macroscopic hematuria, proteinuria, renovascular hypertension, flank pain, dyspareunia, dysmenorrhea, and pelvic varicose veins[7]. This symptomatic presentation reflects the degree of LRV compression, renocaval hypertension, and development of collateral circulation. Symptoms are often aggravated by physical activity[8]. In this context, the most common symptom is hematuria, and it is due to elevated LRV pressure, resulting in the rupture of thin-walled septum between the varices and the collecting system in the renal fornix. Pain is a result of the inflammatory cascade triggered by venous hypertension. Flank and abdominal pain are the consequences of that inflammatory process[5].

Diagnosis is challenging and generally is made after exclusion of other more common causes. Considering the rarity of this disease, it becomes necessary a high index of suspicion. Contrast-enhanced Computed Tomography (CECT) is generally used to evaluate patients with hematuria of unknown causes and, in this case, an angle less than 45º between aorta and SMA[7] is a common finding and accurate criteria. Magnetic Resonance Imaging (MRI) angiography can also be performed. Confirmation of the syndrome is by US Doppler, the most widely used method, in which a venous pressure gradient between the LRV and the inferior vena cava ≥ 3 mmHg and five times increase in maximum flow velocity in the LRV as it passes the SMA, in relation to the renal hilum[7], can be seen. Venography with measurement of the renal vein pressure gradient is the gold-standard exam, but its invasive nature makes it a later resource that is very often unnecessary for diagnosis (Figure 5). To rule out more common renal conditions, diagnostic methods include blood examinations, urinalysis, urine culture, cytology, urethrocystoscopy, CT urography, and renal biopsy[8]. NCS can be suspected from the clinical history and a urine erythrocyte morphology of predominantly isomorphic erythrocytes, also, a cystoscopic finding of left-sided hematuria is a supporting evidence for this disease[9].

Management options range from observation to nephrectomy, depending on the severity of symptoms. In case presenting with mild hematuria or with mild and tolerable symptoms, conservative management is recommended. However, surgery may be considered for gross hematuria (especially if recurrent); for severe symptoms including flank pain or abdominal pain, anemia, autonomic dysfunction, impairment of renal function including persistent orthostatic proteinuria, varicocele formation; and for ineffective conservative measures after 24 months in patient aged less than 18 years and after 6 months in adults[6,8]. Conservative management is preferred in patient aged 18 years or younger, as growing individuals may experience symptom resolution because of an increase in intra-abdominal fat and fibrous tissue at the SMA origin, releasing the entrapped LRV[6] (Figure 5). Medical therapy has also been employed including angiotensin converting enzyme inhibitors, specifically Alacepril, to improve orthostatic proteinuria and aspirin to improve renal perfusion[6].

Open surgery intervention for anterior NCS includes: transposition of the LRV, which is the most common and effective surgery for treatment, left kidney autotransplantion, transposition of the SMA, nephropexy, nephrectomy, renocaval bypass and transposition of the left gonadal vein[7]. Laparoscopic extravascular stent placement is an alternative to open surgical operation because it allows a direct visualization of the surgical field with no LRV clamping[9]. The endovascular approach can also be used, by the embolization of the left gonadal vein and endovascular stent placement; these techniques are gaining strength recently and ever more being used, as more data is published. Furthermore, because of its unusual condition, the paucity of evidence, lack of data and long-term follow up makes it difficult to establish specific diagnostic criteria and, also, standard treatment options[9]. However, many experts consider the open surgical approach the standard of care for the treatment of NCS[10].

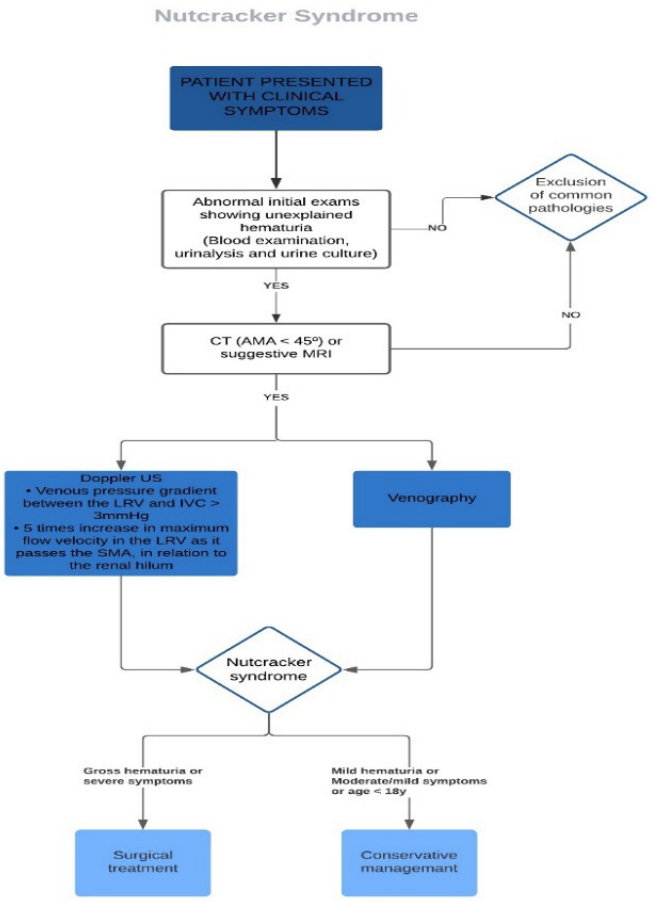

Figure 5. NCS algorithm of diagnosis and management.

CONCLUSION

NCS should be considered as a diagnosis in patients with unexplained hematuria, either gross or microscopic. Although uncommon, it's necessary a high degree of suspicion to make this important exclusion diagnosis and reduce the morbidity associated with it, including the risk of chronic kidney disease from long-term renal vein hypertension and the risk of LRV thrombosis.

Authors participation: We inform for due purposes that the article was prepared jointly by the group of authors with the following degree of participation: Data collection – *Khouri TEF; Faria IR.* Organizational orientation and on the essence, argumentation and relevance of the work – *Khouri TEF.* Analysis, research of articles, reading and exclusion of research not relevant to the involvement of the chosen theme – *Khouri TEF.* Reading and writing the content – *Khouri TEF; Faria IR.* Revision of the text regarding integrity and veracity as the sources used - *Khouri TEF; Faria IR; Miguel GPS.* Thus, the group of authors certifies joint participation in the preparation of the article, hoping to contribute to the theme in question.

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