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CASE REPORT OPEN ACCESS

NUTCRACKER SYNDROME AND STENOSIS OF THE URETEROPELVIC JUNCTION: CASE REPORT

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ABSTRACT

Nutcracker syndrome (CNS) is characterized by compression of the left renal vein (LRV) between the abdominal aorta and the superior mesenteric artery (SMA) due to a reduction in the aortic-mesenteric angle. Stenosis of the ureteropelvic junction (UPJ) occurs due to obstruction of urinary flow at the anatomical confluence of the proximal ureter with the renal pelvis, due to intrinsic or extrinsic forces, and may be congenital or acquired. We will describe the case of a 52-year-old woman, without previous comorbidities, complaining of pain in the left abdominal flank, nausea and sudden onset vomiting. During the investigation, after carrying out several tests, including tomography and scintigraphy, it was observed that the patient presents a rare association of these two pathologies, with few cases reported in the literature.

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INTRODUCTION

Nutcracker syndrome (NCS) is characterized by compression of the left renal vein (LRV) between the abdominal aorta and the superior mesenteric artery (SMA) due to a reduction in the aortic-mesenteric angle. Ureteropelvic junction (UPJ) stenosis, which can be congenital or acquired, occurs due to obstruction of urinary flow at the anatomical confluence of the proximal ureter with the renal pelvis [1]. To date, we have observed few cases reported in the NCS literature in association with ureteropelvic junction stenosis.

OBJECTIVE AND METHODS

To describe the imaging findings of a patient with a rare association of Nutcracker syndrome and ureteropelvic junction stenosis.

DISCUSSION

A 52-year-old woman, with no previous comorbidities, arrives at our service complaining of intense pain in the left flank of sudden onset, associated with chills, increased urinary frequency, nausea and vomiting for approximately 3 to 4 months. She previously sought hospital service in her neighborhood, where symptomatic medication and antibiotics were prescribed, and her condition improved. She is currently asymptomatic. At our service, the patient underwent urinary tract ultrasound, revealing an enlarged left kidney and changes suggestive of hydronephrosis and/or polycystic disease with significant cortical thinning. Stenosis of the UPJ and changes suggestive of associated pyelonephritis were also noted. Based on the findings of the tests mentioned above, renal scintigraphy with DSMA (dimercaptosuccinic acid) was requested to evaluate the renal function of the affected kidney, in addition to a new tomography of the abdomen and pelvis with contrast. The scintigraphy revealed normal glomerular and cortical tubular function on the right and moderate/severe depression on the left, in addition to dilation of the left pyelocaliceal system, with a pattern suggestive of obstruction on this side. Computed tomography of the abdomen and pelvis with contrast showed a reduction in the aorto-mesenteric space (angle of 26º) with thinning/stenosis of the left renal vein and thinning/stenosis of the third/fourth duodenal portion. Consequently, a moderate ectasia of the left gonadal vein was characterized, with pelvic varicose veins also on the left. It was demonstrated that the dilated left gonadal vein promoted compression over the upper third of the left ureter, at the level of the left ureteropelvic junction, with marked

ureterohydronephrosis upstream and diffuse renal cortical thinning on the left. It was highlighted that the left kidney showed a delay in the elimination of the contrast medium, showing signs of nephropathy. In NCS, the most common clinical manifestations are left flank pain, pelvic pain, hematuria and gonadal varices.

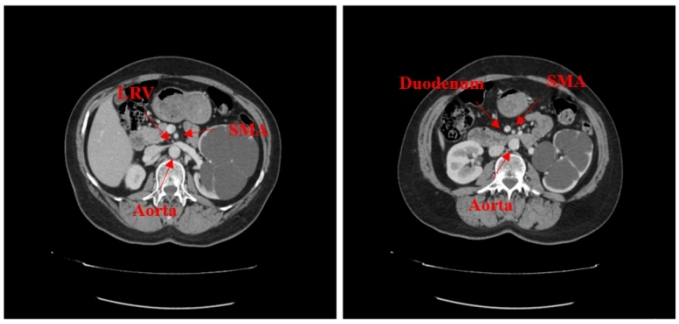

Figure 1. Computed tomography of the upper abdomen, in the arterial phase of the contrast medium, in axial section: In the left-sided image, a notable stenosis of the left renal vein (LRV) is evident between the aorta and the superior mesenteric artery (SMA). Simultaneously, the left kidney appears dilated with hydronephrosis and significant parenchymal thinning. The right-sided image reveals compression of the third portion of the duodenum between the aorta and the superior mesenteric artery (SMA), along with associated changes in the left kidney.

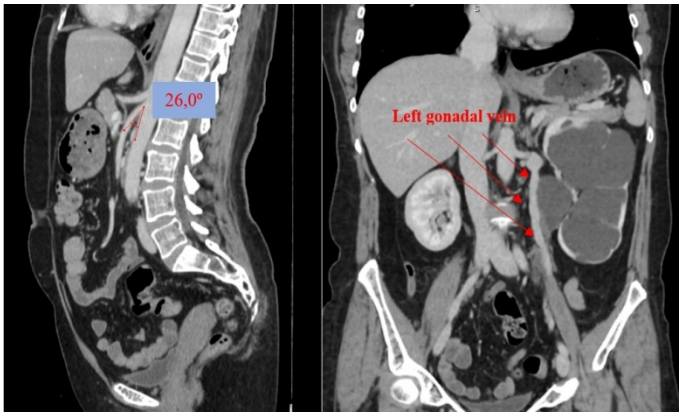

Figure 2. Computed tomography of the abdomen and pelvis: On the left we have a sagittal section in the arterial phase of the contrast medium showing the reduced aorto-mesenteric angle (26 degrees). On the right we have a coronal section in the portal phase of the contrast medium that clearly characterizes the ectasia and early enhancement of the left gonadal vein, this left gonadal vein promoting compression on the left proximal ureter, at the level of the ureteropelvic junction, with consequent hydronephrosis upstream and thinning of the left renal parenchyma.

There are also reports of orthostatic proteinuria and renovascular hypertension. In asymptomatic patients, the term used is nutcracker phenomenon[2]. Compression of the VRE occurs mainly between the VMS branch and the aorta, however cases may also occur in which the VRE has a retro-aortic location and is compressed between the aorta and the vertebrae. This compression determines a significant reduction in the caliber of the VRE, causing obstruction of the flow to the inferior vena cava (IVC) and consequently secondary left renal venous hypertension. In some situations in the NCS, the third duodenal portion follows anteriorly the VRE and is also compressed between the aorta and SMA, in this case known as superior mesenteric artery syndrome or Wilkie syndrome[3]. This syndrome is more common in women and involves patients with ages ranging from childhood to the elderly, with a higher prevalence in patients between 20 - 30 years old and middle-aged adults[4]. The diagnosis of NCS is initially carried out after excluding other causes that could trigger the patient's complaints. A careful analysis, a complete clinical history, physical and laboratory examination are necessary[5]. Confirmation is done through imaging tests, including: renal ultrasound with Doppler, where a five-fold increase in the maximum flow velocity in the VRE as it passes through the SMA in relation to its hilar portion infers the disease; in contrast-enhanced computed tomography it is possible to measure the compression rate of the VRE, measure the aorto-mesenteric angle, with values lower than 45º being abnormal (range between 35 – 56º) and evaluate the presence of collateral pathways, the main one being the left gonadal vein, which will present early enhancement during the portal venous phase; magnetic resonance imaging presents similar findings to tomography; and renal angiography, which is currently more used for treatment as it is an invasive method, shows a venous gradient between the VRE and the inferior vena cava (IVC) $\geq$ 3 mmHg[2,4]. Treatment in asymptomatic patients under 18 years of age can initially be carried out conservatively for 24 months. In adults, initial follow-up for 6 months is recommended before the surgical approach. There are several techniques, including: VRE transposition, left kidney autotransplantation, SMA transposition, nephropexy, renocaval shunt and, in refractory cases, nephrectomy[4].

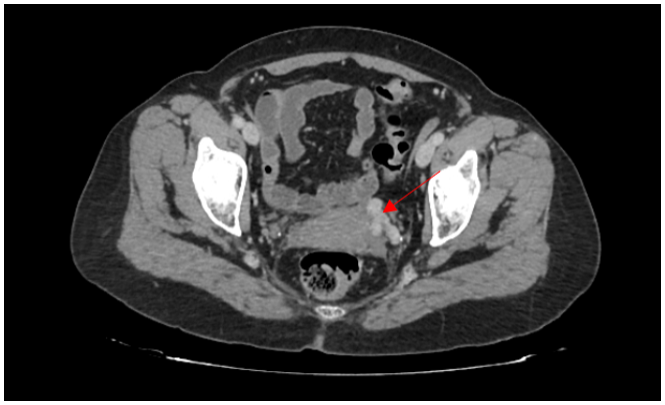

Figure 3. Computed tomography of the pelvis, in the contrast medium portal phase, in axial section: We characterize the parauterine pelvic varices on the left (red arrow), drawing attention to this focal and unilateral location of the pelvic varices, denoting signs of possible changes in the upper structures

Stenosis of the ureteropelvic junction can be present in both the pediatric population and adults, but with different causes. Obstruction occurs due to intrinsic or extrinsic forces and may be congenital or acquired[5]. The congenital cause, common in the neonatal period, is generally idiopathic/unknown or due to extrinsic compression of the ureter. In adults, as in the case of the patient under study, the most common causes are renal trauma, obstructive stones, sequelae of pyelitis, intrinsic neoplasms (such as urothelial carcinoma of the upper tract) and extrinsic compression. Symptoms of UPJ obstruction in adults vary, ranging from asymptomatic to vague and mild or sudden and severe complaints. Furthermore, in many patients the diagnosis is identified incidentally when the renal tract is examined for other reasons[5]. When present, signs and symptoms include flank pain, nausea and vomiting, hematuria, infection and palpable mass[1]. Diagnosis is made by magnetic resonance imaging, computed tomography, ultrasound, retrograde urography and scintigraphy, the latter being the gold standard for evaluating obstruction and renal function[6]. Treatment aims to provide adequate drainage of the affected kidney and relieve obstruction. Indications for surgical intervention are: infection, severe and recurrent flank pain or nausea, routine stone formation and loss of kidney function. It includes three groups of procedures: open surgical procedures, endoscopic procedures (anterograde or retrograde procedures) and laparoscopic[1,7].

CONCLUSION

Nutcracker syndrome that evolves with ureteropelvic junction stenosis is a rare and infrequent finding, with both presenting similar

symptoms. An adequate investigation with an accurate diagnosis favors early intervention, significantly improving the patient's prognosis in an attempt to keep the kidney intact and with good function. Imaging exams play a fundamental role in the exact diagnosis, with computed tomography being one of the methods of choice for elucidating the case. It is crucial for radiologists to be well-prepared to promptly make this type of diagnosis, facilitating more effective patient management.

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