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Case report

Nutcracker syndrome: A rare cause of chronic pelvic pain and left back pain ☆,☆☆

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ABSTRACT

The Nutcracker Syndrome is a rare and often unrecognized cause of chronic pelvic pain and left back pain. These symptoms are due to the left renal vein compression between the aorta and the superior mesenteric artery (anterior nutcracker) or between the aorta and the spine (posterior nutcracker). The variety of clinical manifestations make the diagnosis difficult and commonly delayed. Therefore, imaging plays a key role in correcting the diagnosis by confirming the left renal vein stenosis and ruling out any differential diagnosis. Treatment options are discussed by a multidisciplinary team involving urologists, nephrologists and vascular surgeons for each patient. We report the case of 2 patients presenting chronic pelvic and lower back pain in whom clinical investigation and CT imaging findings were consistent with a nutcracker syndrome.

Introduction

Anterior nutcracker syndrome consists of all manifestations caused by the compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA) [1]. First described in 1937 by pathologist Grant, this syndrome remains unfortunately unknown to many physicians and often unrecognized. The nutcracker syndrome is associated with a symptom complex that can be misleading, it is dominated by hematuria, which may or may not be associated with left back pain, pelvic congestion syndrome for women and varicocele

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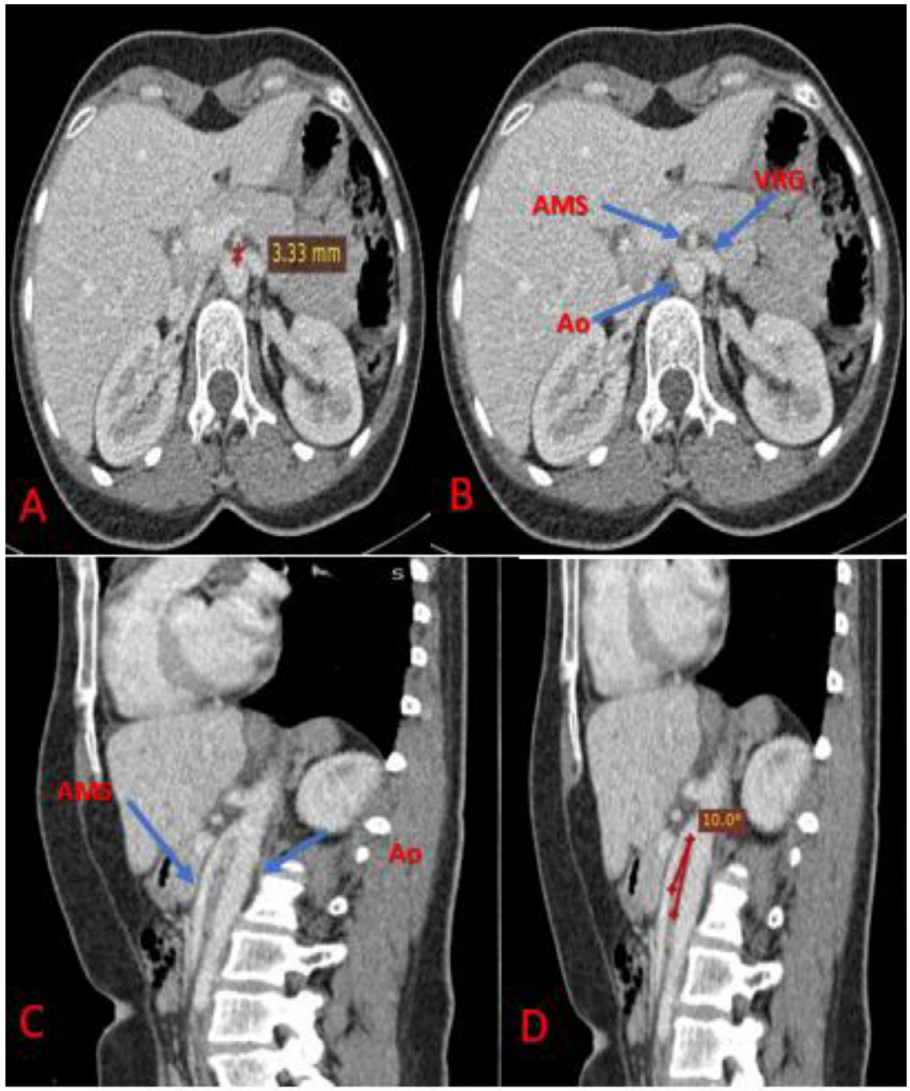

Fig. 1 – Case 1 Abdomino-pelvic CT scan with injection of PDC in sagittal and axial section showing compression of the left renal vein in the aorto-mesenteric space reduced to 3 mm with VRG stenosis of 73% (Figs. 1A and B). The sagittal reconstruction shows an aorto-mesenteric angle of 10° (Figs. 1C and D) AMS, Superior mesenteric artery; Ao, Aorta; VRG, Left renal vein; VOG, Left ovarian vein.

for men. The diagnosis is suggested on clinical and laboratory findings then confirmed on imaging. Management is multifactorial and multidisciplinary. Through these 2 cases, we show the key role of imaging to better recognize the nutcracker syndrome.

Observation

Observation 1

A 36-year-old women, mother of 2 children with history of nasopharyngeal cancer treated with radiation therapy, presented with chronic left lower back pain treated with analgesics without any improvement. Lumbar spine plain films and iterative abdominal pelvic ultrasound showed no abnormalities. All laboratory tests performed (complete blood count, metabolic panel, urinalysis, vaginal swab, C reactive protein test) came back normal.

The patient was then referred to our hospital to investigate for any distant disease recurrence. The cervical thoracic abdominal pelvic contrast enhanced CT showed no signs of distant metastasis. However, it revealed compression of the LRV within an aorto-mesenteric angle of 10° and reduced aorto-mesenteric space to 3 mm with loss of the surrounding fat. This resulted in a 73% stenosis of the LRV compared to the hilar segment (Fig. 1).

It was associated with a dilation of the left ovarian vein measuring up to 9 mm in caliber and ipsilateral utero-ovarian varices (Fig. 2). After CT examination, we then re-interviewed the patient who reported having chronic pelvic pain along with left back pain, dyspareunia but no notion of hematuria.

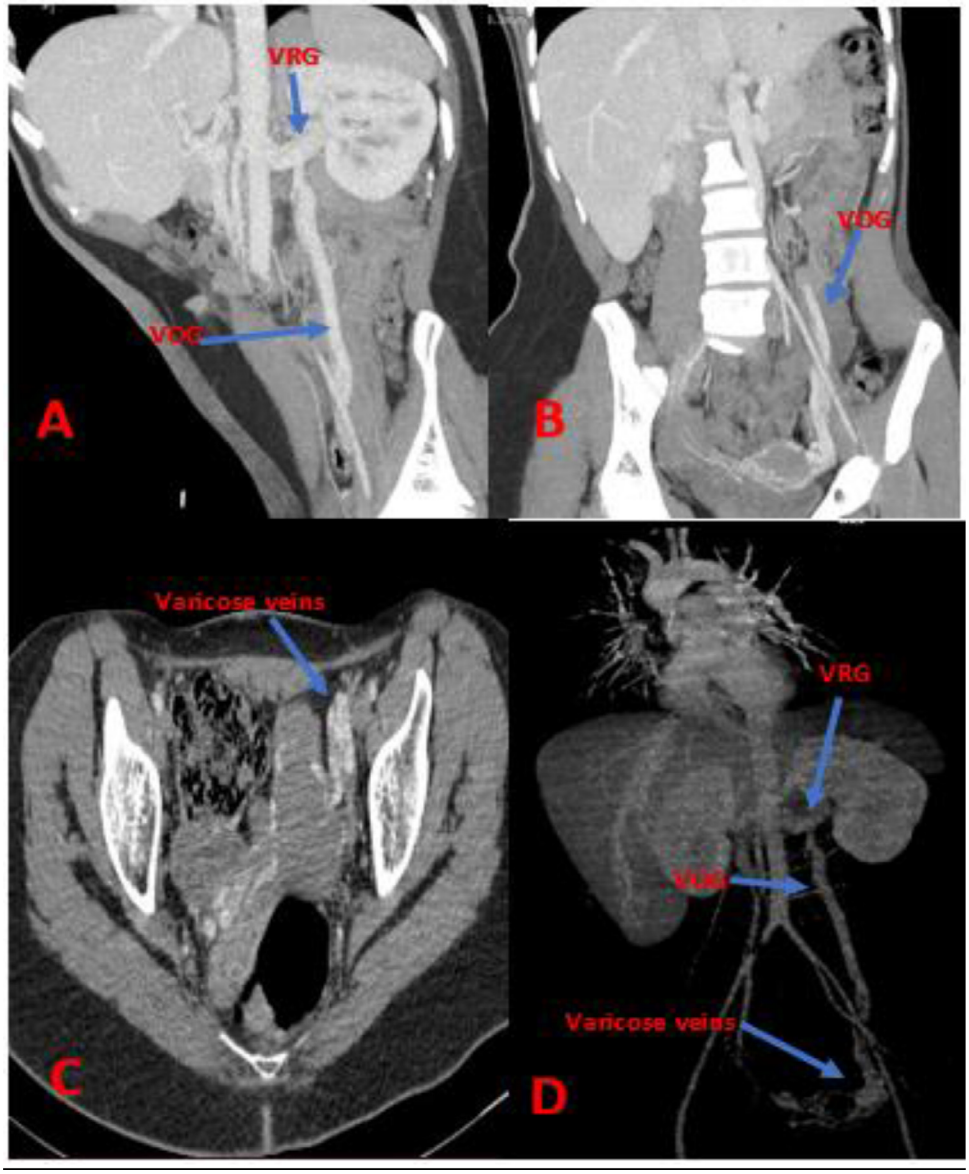

Fig. 2 – – Case 1 Abdomino-pelvic CT scan with axial PDC slice injection (Fig. 2C), in sagittal and oblique coronal reconstruction in MIP (Figs. 2A and B) and VR (Fig. 2D) showing dilation of the left renal and ovarian veins and utero-ovarian varices ipsilateral.

Regarding these clinico-biological and radiological findings, the final diagnosis of the nutcracker syndrome was retained and the patient referred to a urological and vascular consult for further evaluation and treatment.

Observation 2

A 54-year-old multiparous women with history of breast cancer with liver metastasis under palliative chemotherapy, presented with chronic pelvic pain, a feeling of heaviness in the pelvis and atypical left lower back pain, with no other associated symptoms. She consulted several times and performed multiple tests: inflammatory assessment (CRP, complete blood count), infectious assessment (urinalysis, vaginal swab), ionogram, renal assessment (urea, creatinine), and radiological assessment (abdominal ultrasound, plain abdominal film, lumbar spine x-ray, thoracic abdominal pelvic CT scan), all came back normal except for a liver riddled with metastatic nodules.

The symptomatology was thought to be linked either to the neoplastic, psychological context or to nephro lithiasis eliminated during miction.

She was then referred to our institution for a thoracic abdominal pelvic contrast enhanced CT for her known cancer follow up. CT showed a stable disease with no appearance of new suspicious lesions.

Further analysis showed a compression of the LRV in the aorto-mesenteric space reduced to 4 mm with an aorto-mesenteric angle of 12° and 75% stenosis in the aorto-mesenteric segment compared to the hilar segment (Fig. 3). It was associated with a dilation of the left ovarian vein up to 10

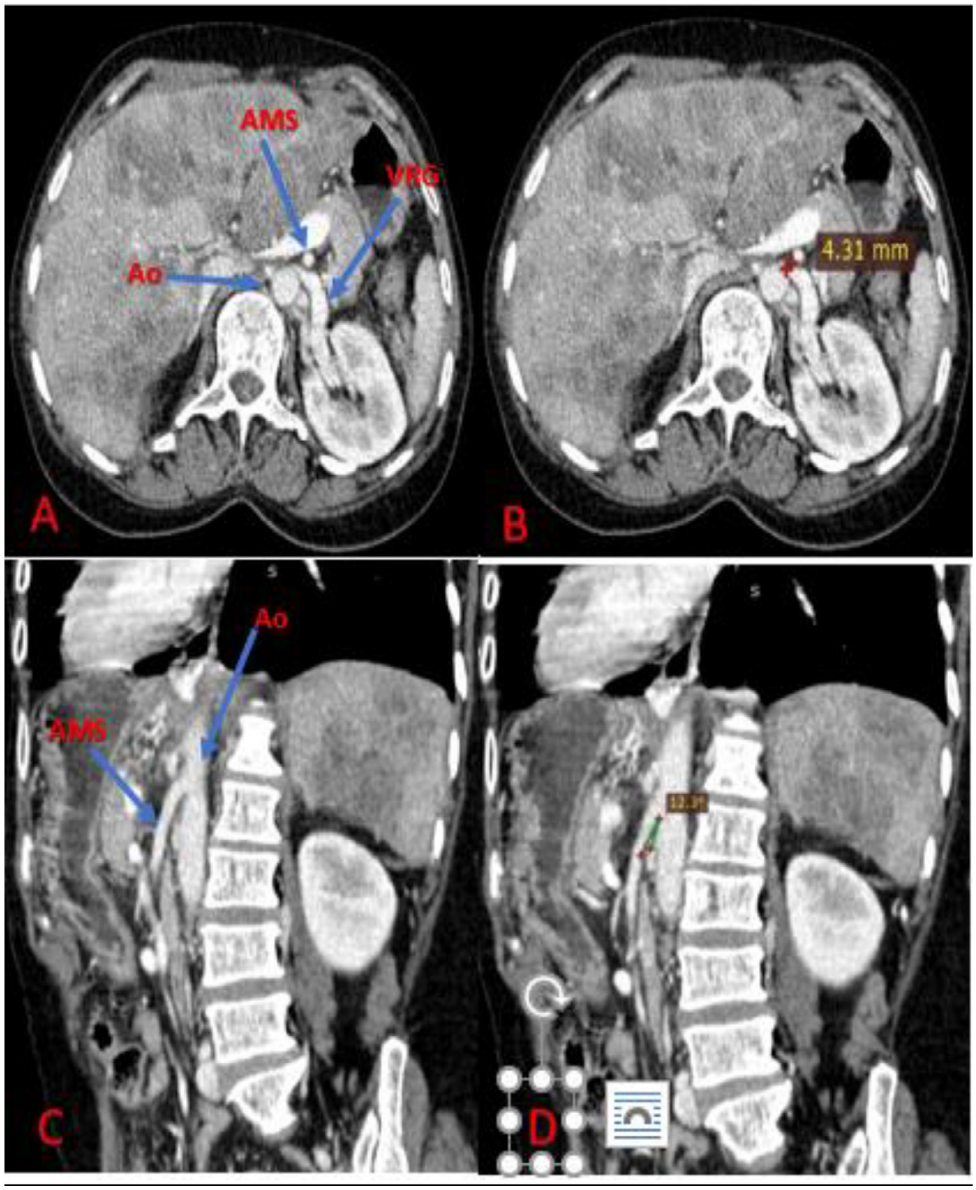

Fig. 3 – Case 2 Abdomino-pelvic CT scan with PDC injection in axial section (Figs. 3A and B) with oblique sagittal reconstruction (Figs. 3C and D) showing compression of the left renal vein in the aorto-mesenteric space reduced to 4 mm with an aorto-mesenteric angle of 10° and a stenosis of the VRG of 75%. Note: liver damage.

mm in caliber and ipsilateral utero-ovarian varices (Fig. 4). The diagnosis of the previous nutcracker syndrome was retained and the patient was referred to the urological and vascular surgeon team for further management.

Discussion

Nutcracker syndrome was first described in 1937 by pathologist Grant [2]. It was not until 1950 that El Sadr and Mina reported the first case. In 1972 De Schepper gave it the name Nutcracker Syndrome [3]. It has an anatomical definition and a precise pathogenesis: compression of the LRV between the SMA and the aorta: Anterior nutcracker or between the aorta and the spine: Posterior nutcracker [4]. This results in dilation of the left renal and gonadal veins with development of collateral venous circulation (Pyelocaliceal, peri-ureteral and gonadal) due to congestion and increased venous pressure. It is a rare and underestimated syndrome, which mainly affects young adults with a vast majority of women [5, 6]. Its prevalence remains unknown, probably because it is often misdiagnosed and commonly asymptomatic. Several etiopathogenic hypotheses have been reported in the literature: dorsolateral ptosis of the left kidney with stretching of the LRV, too low or lateral origin of SMA implanted at a steep angulation while the normal angle of insertion is approximately 90°. The risk factors described are: anatomical variants (left renal duplicity, horseshoe kidneys, ectopic origin of gonadal veins), important weight loss, hyper lordosis, absence of peritoneal fat,

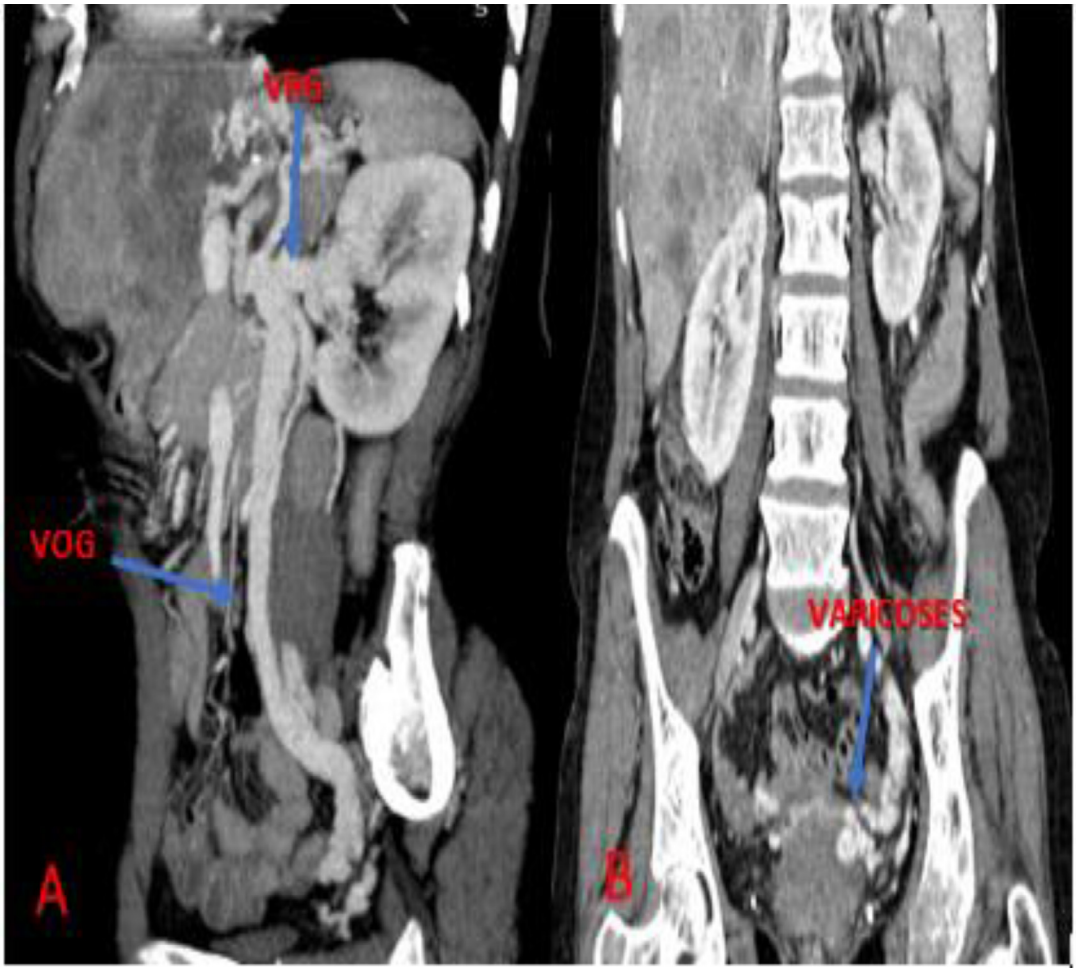

Fig. 4 – Case 2 Abdomino-pelvic CT scan with injection of PDC coronal section (Fig. 4A) and sagittal reconstruction in PIM (Fig. 4B) showing dilation of the left renal and ovarian veins associated with ipsilateral utero-ovarian varices.

retro peritoneal fibrosis and all situations with increased venous pressure [7].

The term nutcracker syndrome is reserved for patients with clinical symptoms associated with these anatomical features because similar anatomical variants without clinical repercussions are not uncommon and do not require any management. In these cases we speak of the Nutcracker phenomenon [8]. The main symptom is micro or macroscopic hematuria, often intermittent. Other manifestations have been described: pelvic pain, left lower back pain, orthostatic proteinuria, pelvic or atypical varices of the lower extremities, and pelvic congestion syndrome in women and varicocele in men. Systemic manifestations have been reported in adolescents such as headache, abdominal pain, and tachycardia [9]. This polymorphism also contributes to the delay in diagnosis, bringing up first the most frequent causes of chronic pelvic pain (endometriosis, adenomyosis, fibromas, infectious, tumoral disease) and lumbar pain (renal colic, pyelonephritis, herniated disc, osteoarthritis, tumors, infections)

The diagnosis cannot be conceived without imaging. Indeed, imaging eliminates the differential diagnoses, which are multiple, and confirms the diagnosis. Renal Doppler ultrasound is a non-invasive diagnostic tool with good sensitivity and specificity of 89-100% but requires an experienced operator. In a 2007 study by de Shin et al on 149 children with hematuria of unknown etiology, Doppler ultrasound was considered normal. The re-examination by an experienced radiologist revealed a nutcracker syndrome in 60 children [10, 11]. Renal Doppler ultrasound shows a decrease in the diameter of the LRV at the level of the aortomenteric clamp with increased circulatory velocities between the renal hilum and the level of compression. However, abdominal computed tomography (CT) with injection of contrast media is the modality of choice to visualize the compression of the LRV and its hemodynamic consequences. The characteristic sign is the visualization of a beak corresponding to compression of the LRV by the aorto-mesenteric steep angle. On sagittal reconstructions, the normal angle formed by the SMA and the aorta should be between 35°-56°. A narrowing is confirmed when this angle is between 7°-22°. The SMA-aorta distance is reduced, measuring between 2-8 mm with disappearance of the normal surrounding retroperitoneal fat, while the normal distance is 10-28 mm [12].

Also, computed tomography makes it possible to quantify the degree of stenosis of the LRV, which is greater than 70% in the aorto-mesenteric segment compared to the hilar segment; to measure the ratio between the diameter of the hilar and aorto-mesenteric LRV's segments, which is greater than 4 compared to a normal ratio of 2.

Furthermore, it shows a dilation of the renal and gonadal veins and visualizes pelvic varices.

Magnetic resonance imaging shows similar findings with excellent anatomical analysis thanks to high soft tissue contrast and multiplanar capability using sagittal and coronal slices [9].

Lumbar venography used to be the gold standard to confirm the diagnosis, but it remains an invasive examination supplanted by cross-sectional imaging. In fact, it is now performed for treatment purposes rather than diagnostic ones. It measures the pressure gradient between the IVC and the LRV, which is greater than 3 mm Hg in case of a nutcracker, for a normal value of 0-1 mm Hg [11].

Currently there is no standard regarding treatment, it is discussed depending on each patient's background and on the severity of symptoms. There are many therapeutic modalities, their primary goal is to alleviate LRV outflow obstruction and hypertension. Surgery is only reserved for disabling cases or failure of other modalities. Some teams opt for endovascular treatment by embolization or placement of a stent at the level of the LRV [13]. In moderate cases in young patients or with mild symptoms, a wait-and-see strategy is recommended.

Conclusion

Nutcracker syndrome is a rare entity. Lack of awareness of this syndrome leads to diagnostic delays. It should be considered in front of a chronic pelvic pain syndrome, an atypical lower back pain associated or not with hematuria. It is one of the causes of pelvic congestion syndrome in women and varicocele in men.

Imaging plays a major role to make the correct diagnosis, computed tomography being the modality of choice.

Endovascular treatment by placing a stent in the LRV has become the first therapeutic option performed in highly specialized centers.

Patient consent

Written informed consent has been obtained from the patients for the publication of this case report and any accompanying photographs.

This case report is an incidental finding in the course of clinical work and has no ethical implications.

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