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Case Report

Nutcracker Syndrome: About 05 Cases with Literature Review

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Abstract: The Nutcracker (NCP) phenomenon is the compression of the left renal vein (LRV) between the superior mesenteric artery (AMS) and the aorta, resulting in a decrease in blood flow from the LRV to the vena cava lower. The term Nutcracker Syndrome (NCS) is reserved for cases where this compression is associated to clinical manifestations. In this study, we report five cases of Nutcracker Syndrome admitted with abdomino-pelvic pain and discuss the clinical and radiological finding in the light of the litterature.

Keywords: Nutcracker syndrome - left renal vein - superior mesenteric artery - imaging.

INTRODUCTION

Nutcracker phenomenon (NCP) is a rare vascular disease entity, caused by contraction of the left renal vein (LRV), usually between the superior mesenteric artery (SMA) and the aorta. The nutcracker syndrome (NCS) results in some symptoms that range from mild proteinuria or asymptomatic microscopic hematuria to severe symptoms duo to pelvic congestion or nephrotic range proteinuria (Kurklinsky, A. K., & Rooke, T. W. (2010, June). Most patients become symptomatic in the second and third decades of their lives (Denham, S. L. W. *et al.*, 2013). Diagnostic algorithms include DUS, CT or magnetic resonance angiography.

OBSERVATION 1

A 38-year-old female patient with chronic abdominopelvic pain associated with, dysmenorrhea, exacerbated by standing and walking. ECBU revealed microscopic hematuria. The DUS showed pelvic varices. Abdominal CT scan revealed a compressed LRV between the aorta and the SMA (fig.-1).

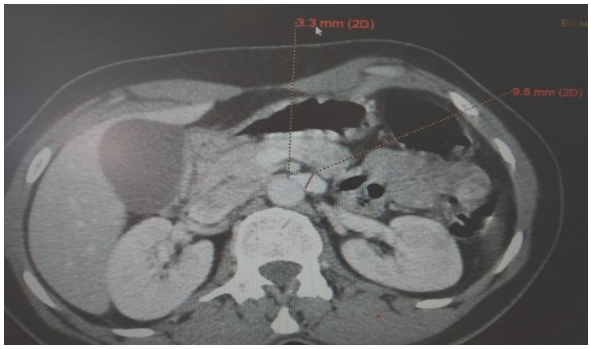

Fig.- 1. LRV is compressed between the aorta and the superior mesenteric artery.

OBSERVATION 2

A 46-year-old man, followed for hypogastric abdominal pain, especially left flank, for 02 years, moderately improved by symptomatic treatments, for which they become more and more resistant. An abdominal DUS showed no abnormalities that could explain these pains, whereas the testicular ultrasound showed a left varicocele The abdominal CT scan showed the LRV being pinched between the aorta and vertebral column (fig.- 2).

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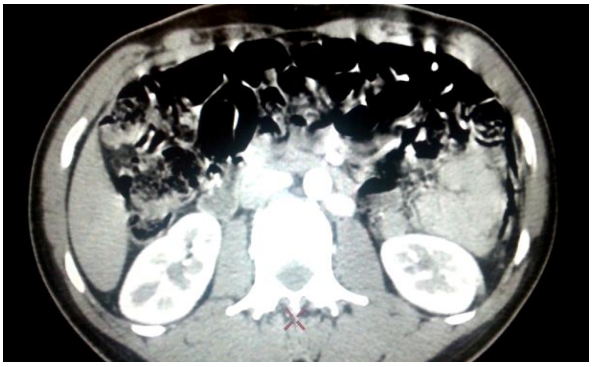

Fig.-2. LRV is compressed between the aorta and the vertebral column.

OBSERVATION 3

A 54-year-old female patient with chronic pelvic pain. The patient also reports notion of hematuria. The abdominopelvic US showed the presence of pelvic varices. Abdominal CT revealed distention of the hilar portion of the left renal vein and the ovarien vein (fig.-3).

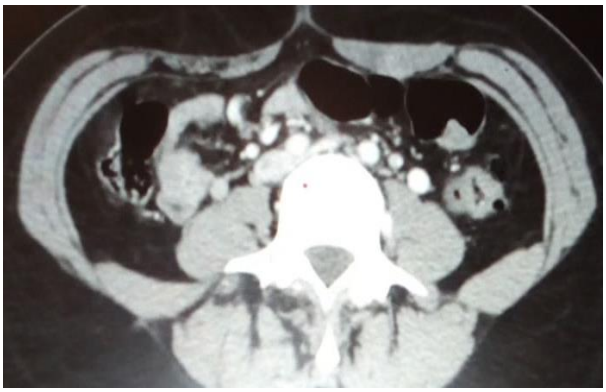

Fig.- 3.CT scan showing dlated left ovarien vein

OBSERVATION 4

A 48-year-old woman with no particular medical history who had chronic abdominopelvic pain associated with feeling of heaviness in lower abdomen, dysuria, dysmenorrhea. ECBU revealed microscopic hematuria. The abdominopelvic US showed pelvic varices. An abdominal CT a nutckcracker phenomen with pelvic congestion (fig.-4).

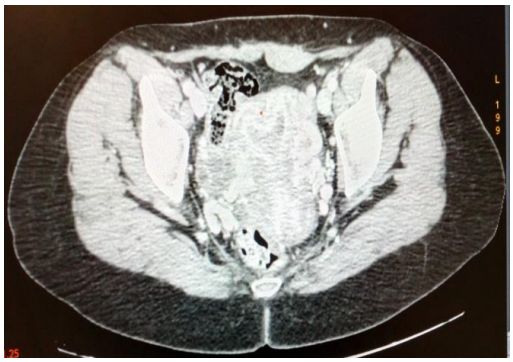

Fig.- 4. CT scan showing pelvic congestion.

OBSERVATION 5

A 38-year-old woman presented with pains of the iliac fossa and left flank, which had been evolving for six years, associated with a feeling of pelvic heaviness. DUS revealed a dilated VRG with pelvic varices. These findings were confirmed by an abdominal CT scan (fig- 5).

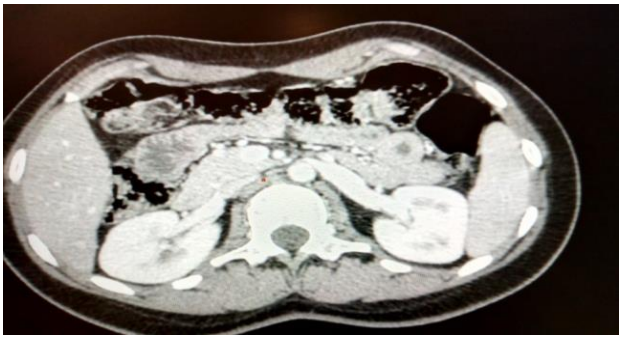

Fig.- 5. CT scan showing nutcracker phenomenon. LRV is compressed between the aorta and the SMA . LRV with distended hilar portion

DISCUSSION

Nutcracker syndrome describes the compression of VRG between the SMA and the aorta, or more rarely, between the aorta and the vertebral column (Kurklinsky, A. K., & Rooke, T. W. 2010 ; Wang, Y., Zhou, Y., & Liu, C. Y. 2016). Both types can be exceptionally associated in case of VRG duplication. SCN is characterized by a decrease in blood flow from VRG to inferior vena cava due to extrinsic compression of VRG. The term SCN should be reserved for patients with clinical symptoms associated with these anatomical features because there are similar anatomical variants that have no clinical implications and should be these cases rather speak of nutcracker phenomenon (Pournasiri, Z. 2016).

The first clinical report of this phenomenon was by El-Sadr and Mina in 1950. The term nutcracker is usually credited to de Schepper (1972), although it was first used by Chait et al (1971); the earliest pathologic description belongs to the anatomist Grant (1937) (Sador, E. L. 1950). The etiological assumptions of the SCN include (1. Kurklinsky, A. K., & Rooke, T. W. 2010):

- A posterior renal ptosis.
- An abnormally high path of VRG.
- An abnormality of insertion of the superior mesenteric artery on the aorta.
- Various processes at the origin of VRG compression.

Because of the variability of symptoms and absence of consensus on diagnostic criteria, the exact prevalence of NCP is unknown but may be slightly higher in females, as shown in our series: 4 women and one man. Patients' age can range from childhood to the seventh decade of life, but most symptomatic patients are in their IIaire or third decade of life (Shin, J. I., &

Lee, J. S. 2005). The average age of our patients was 44 years old.

The frequency and severity of the syndrome vary from asymptomatic microhematuria to severe pelvic congestion. Although some patients have severe and persistent symptoms, many, especially children, are asymptomatic. Symptoms are often aggravated by physical activity and commonly include hematuria, pain or gonadal vein syndrome, varicocele (case of the 4th observation), orthostatic proteinuria, and orthostatic intolerance. 02 women in our series had microscopic hematuria and one woman had macroscopic hematuria.

However, hematuria is not necessary for the diagnosis of CNS. Patients with NCS often present with abdominal pain and left flank pain, and this is the case in all our patients.

BMI correlates positively with NCP and NCP may manifest after weight loss, although many individuals with relatively low BMI and aortomesenteric angles have no signs of NCP (Buschi, A. J.*et al.*, 1980). On the basis of analogy with the SMA syndrome, a decrease in retroperitoneal fat is believed to reduce the AMA and cause NCP. Increased BMI is associated with decreased prevalence of varicoceles. Mild to moderate proteinuria has also been observed in approximately half of patients with NCS (Sawant, D. A., & Moore, T. F. 2015).

Several imaging methods such as Doppler ultrasonography, computed tomography angiography, magnetic resonance angiography and retrograde venography with measurement of pressure gradient between LRV and inferior vena cava and hilar-aortomesenteric LRV diameter ratio > 4 are used to diagnose NCS. Although usually performed only in severe cases, the criterion standard for diagnosing NCS is venography with renocaval pressure gradient determination (Gulleroglu. K. *et al.*, 2014).

Doppler ultrasound, therefore, should be used as the first-line imaging modality for the diagnosis of NCS in both children and adults. The sensitivity and specificity of DUS ranges from 69% to 90% and 89% to 100%, respectively (Kurklinsky, A. K., & Rooke, T. W. 2010). Nutcracker syndrome DUS criteria are based on both the ratio of the peak velocity in the narrowed and dilated portions of the LRV and the ratio of the AP diameter of the LRV in the dilated and narrowed portions. The peak velocity ratio cutoff for NCS ranges from 4.1 to 5.0 (narrowed/dilated), and the AP diameter ratio cutoff ranges from 4.0 to 5.0 (dilated/narrowed) (Denham, S. L. W. *et al.*, 2013).

NCS is a rare condition, which if left untreated may cause thrombosis of the LRV and damage to the left kidney. Although underdiagnosed, there are several treatment options. The patients diagnosed with NCS should be managed according to their age and clinical symptoms. Usually, patients younger than 18 years of age are more frequently treated conservatively, since a majority of such patients will show complete remission in their clinical symptoms including hematuria because of the development of collateral veins and increase in BMI, which is an important factor in the changing renal hemodynamics (Sharp, G., & Glenn, D. A. 2015 ; Nickavar, A. 2016). NCS patients with mild hematuria or with spontaneous resolution of hematuria cancer be managed with conservative therapy. However, both stenting and open surgical intervention for correcting anatomical anomaly, including procedures such as transposition of the LRV or SMA, nephropexy, intravascular and extravascular stent implantation, gonadocaval bypass, renal autotransplantation, and nephrectomy, are indicated for patients with significant pain, renal insufficiency, and severe, persistent life-threatening hematuria. However, selection criteria are not well defined (Pournasiri, Z. 2016).

CONCLUSION

The Nutcracker syndrome is a rare anatomo-clinical entity, to reconise before symptoms such as hematuria and unexplained chronic abdominal pain, in children as in adults. CT angiography and magnetic resonance angiography remain the main stay of diagnosis of this syndrome, but Doppler retains the advantage of being an easy and non-irradiating method, with precise diagnostic criteria.

DECLARATIONS D'INTERETS

Les auteurs déclarent ne pas avoir aucun conflit d'intérêts avec ce manuscrit.

REFERENCES

1. Kurklinsky, A. K., & Rooke, T. W. (2010, June). Nutcracker phenomenon and nutcracker syndrome. In *Mayo Clinic Proceedings* (Vol. 85, No. 6, pp. 552-559). Elsevier.
2. Denham, S. L. W., Hester, F. A., & Weber, T. M. (2013). Abdominal pain of vascular origin: nutcracker syndrome. *Ultrasound quarterly*, *29*(3), 263-265.
3. Wang, Y., Zhou, Y., & Liu, C. Y. (2016). A rare case of nutcracker phenomenon with nephrotic syndrome. *International urology and nephrology*, *48*(4), 631.
4. Pournasiri, Z. (2016). The nutcracker syndrome as a rare cause of chronic abdominal pain: a case report. J Compr Ped,7(3),39741.
5. Sador, E. L. (1950). Anatomical and surgical aspects in the operative management of varicoceles. *Urol. Cutan. Rev.*, *54*, 257-262.
6. Shin, J. I., & Lee, J. S. (2005). Nutcracker phenomenon or nutcracker syndrome?. *Nephrology Dialysis Transplantation*, *20*(9), 2015-2015.
7. Buschi, A. J., Harrison, R., Norman, A., Brenbridge, A. G., Williamson, B. R., Gentry, R. R., & Cole, R. (1980). Distended left renal vein: CT/sonographic normal variant. *American Journal of Roentgenology*, *135*(2), 339-342.

8. Sawant, D. A., & Moore, T. F. (2015). An unusual course of segmental renal artery displays a rare case of hilar nutcracker phenomenon. *Case reports in medicine*, *2015*.
9. Gulleroglu. K., Gulleroglu, B., Baskin, E.,& syndrome, N. (2014). World J Nephrol,63(4),277-81.
10. Sharp, G., & Glenn, D. A. (2015). Differential diagnosis of haematuria following a motor vehicle collision: nutcracker syndrome. Case Rep Surg,7(4),91-82.
11. Nickavar, A. (2016).Nutcracker syndrome; a rare cause of hematuria. J Nephropathol,5(4),144–145.