Case Report

Mixed nutcracker syndrome with left renal vein duplication: A severe and exceptional presentation in an 18-year-old boy

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Abstract

The nutcracker syndrome (NCS) is rare and often misdiagnosed because it embraces an extended non-pathognomonic spectrum of symptoms that imply a difficult diagnosis. Ultimately it may be associated with substantial morbidity and even life-threatening events. Mixed NCS with renal vein duplication is an exceptional variety, have previously been reported to the best of our knowledge. We report a rare case of an 18-year-old boy who presented with a long history of abdominal, pelvic and left flank pain, fatigue and higher bilateral varicocele. Computed tomographic angiography, Doppler ultrasonography and venography were performed revealed left renal vein duplication with dilated retroaortic and preaortic branchs, entrapped respectively between the aorta and the vertebral column and in the aortico-mesenteric space, with extensive and complex varices of the deep pelvic venous plexus; promoting the mixed renal NCS. Auto transplantation of the left kidney was suggested, but refused by the patient; and only the varicocele was managed. The patient is still suffering from his severe initial symptoms. Diagnosis is difficult and should be considered in patients with inexplicable flank or abdominal pain. Our purpose is to raise clinician's awareness for this condition so that they will be more likely to diagnose it. This will facilitate prompt diagnosis and treatment.

Key Words: Abdominal and pelvic pain, duplication of the left renal vein, mixed nutcracker syndrome, varicocele

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INTRODUCTION

Nutcracker syndrome (NCS) is a rare entity caused by the left renal vein (LRV) entrapment,[1] most usually between the aorta and the superior mesenteric artery (SMA), known as anterior NCS. Sometimes a retroaortic position of the LRV also promotes an entrapment, this time between the aorta and the vertebral column, which is named posterior NCS.[2]

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This is a relatively common anatomical variance, in which the patient stays asymptomatic and it is often diagnosed in an occasional imaging exam. This syndrome can manifest by the left flank and abdominal pain, with or without macroscopic or microscopic haematuria. When the venous reflux caused by the LRV entrapment leads to the formation of collaterals this syndrome may be a cause of pelvic congestion syndrome characterized by an array of signs and symptoms such as lower abdominal pain and pelvic, perineal and lower limb varices.[3]

NCS with LRV Duplication is exceptional, only one case of LRV duplication with a dilated retroaortic branch was described as posterior NCS with renal vein duplication;[4] Here we present the first case of mixed NCS with renal vein duplication combining dilated retroaortic and preoaortic branchs, entrapped

respectively between the aorta and the vertebral column and in the AMS and the aorta (aortico-mesenteric space).

This illness can present quite differently evoking a heightened sense of alert in order to allow an accurate and in time diagnosis.

CASE REPORT

This was a case report of an 18-year-old boy presented to our institution with abdominal, pelvic and left flank pain and fatigue, which lasted for the 7 years prior to his first under hospital observation; with daily use of increasing doses of analgesics, including opioids, with no therapeutic success, despite a long investigational history combined with several imaging examinations and laboratory tests.

On physical examination, only a bilateral varicocele was detected visually through the scrotal skin. Initial laboratory tests revealed microhematuria and no other abnormalities.

The ultrasonography showed normal kidney measurements and echogenicity, excluding other anatomical defects.

The Doppler study revealed turbulent pattern of venous blood flow of the posterior and anterior LRV branchs behind the aorta.

Computed tomographic angiography (CTA) was performed revealed LRV duplication with dilated retroaortic and preoaortic branchs, entrapped respectively between the aorta and the vertebral column and in the aortico-mesenteric space. The post-anterior diameter of the hilar portion and that of the aortico-mesenteric and aortico-vertebral stenotic portions of the LRV were 9.8 mm and 1.7 mm and 11 mm and 8 mm, respectively. The promoting the anterior and posterior renal NCS [Figures 1-4].

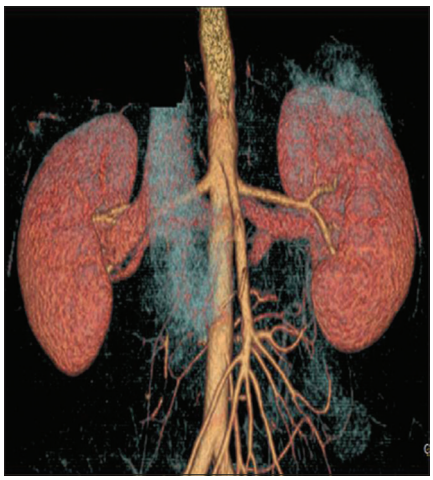

Figure 1: Computed tomographic angiography showed left renal vein duplication with dilatation of the hilar portion

Surgery for NCS by auto transplantation of the left kidney was suggested; however, the patient refused and was referred to the vascular and endovascular surgery service to evaluate the possibility of a minimally invasive treatment of varicocele such as a radiologic embolization. He was then submitted to venography, Through a puncture of the femoral vein, the LRV was catheterized showing clamping of the two LRV branchs, with a significant increase in its proximal diameter, besides extensive varices of the deep pelvic venous plexus, with inverted venous reflux in the left spermatic vein, which also had a diameter about 2-3 times larger than expected [Figures 5 and 6]. Endovascular therapeutic alternative was initially suggested by left anterograde embolization of varicocele and embolization of pelvic varices. However, this alternative has not been made because of the principal spermatic vein and the collateral veins were impossible to cannulate and because of the high occurrence of vital complications. And the patient was lost to follow up thereafter.

In another institution, surgical treatment of left varicocele was performed, through retroperitoneal approach with isolation and ligation of three large internal spermatic veins. The operator described several other small retroperitoneal collaterals, which were ligated and divided.

At 1 year later, he presented with the same initial symptoms with any improvement in both varicocele and spermogram. Bilateral varicocelectomy was performed by inguinal approach, the cords were dissected and several large spermatic veins were ligated.

At the follow-up, the varicocele improved significantly after the operation, but without improvement of the spermogram. The patient still refuses any surgical treatment of the NCS.

DISCUSSION

The most typical nutcracker morphological features imply compression of the LRV between the aorta and SMA, which

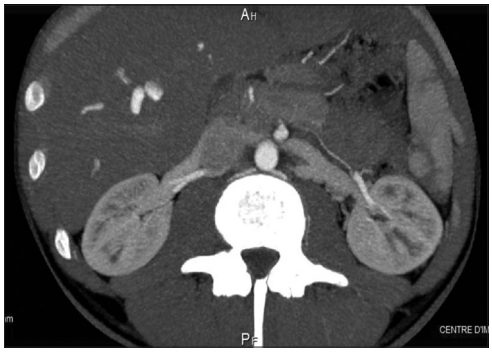

Figure 2: Computed tomographic angiography showed compression of the anterior branch of the left renal vein between the abdominal aorta and the superior mesenteric artery

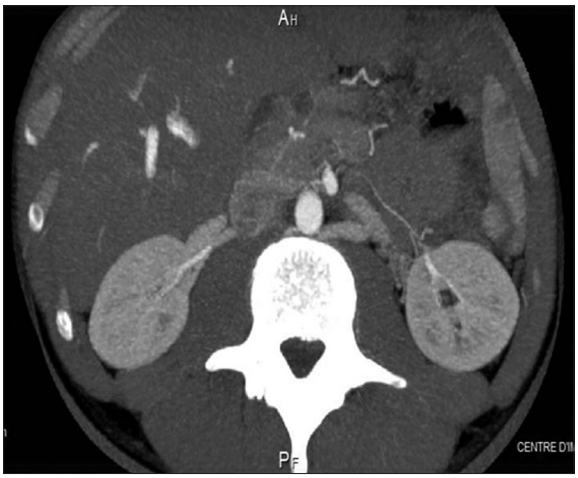

Figure 3: Computed tomographic angiography showed compression of the posterior branch of the left renal vein between the abdominal aorta and the vertebral column

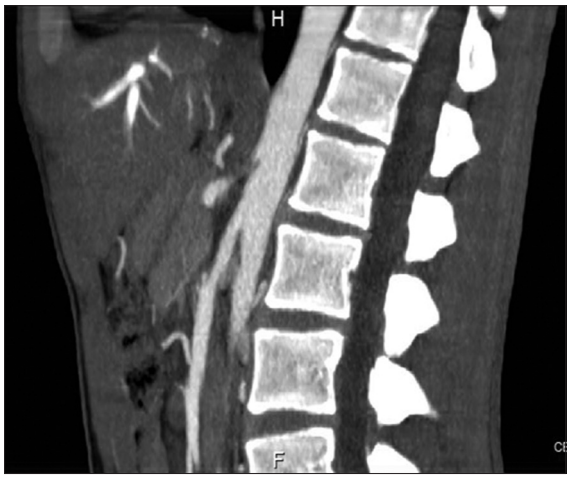

Figure 4: Computed tomographic angiography demonstrated the acute angle between the aorta and superior mesenteric artery at 12°; and showed the retroaortic and the preoaortic branchs, entrapped respectively between the aorta and the vertebral column, and between the abdominal aorta and the superior mesenteric artery

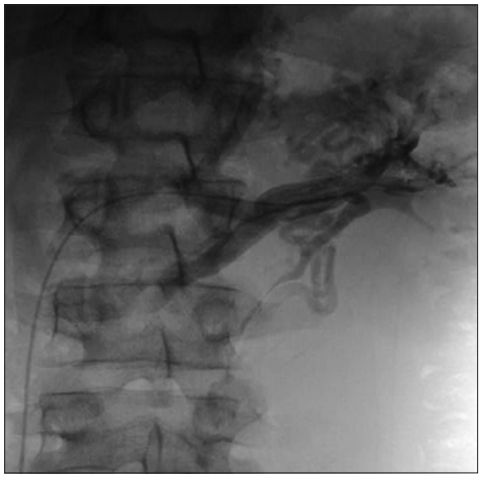

Figure 5: Diagnostic phlebography (left) with a 4F cobra catheter. Higher grade pelvic varicosities with contralateral filling of the left spermatic vein and confirmed the extrinsic compression of the two branchs of the LRV with spermatic and collaterals venous reflux

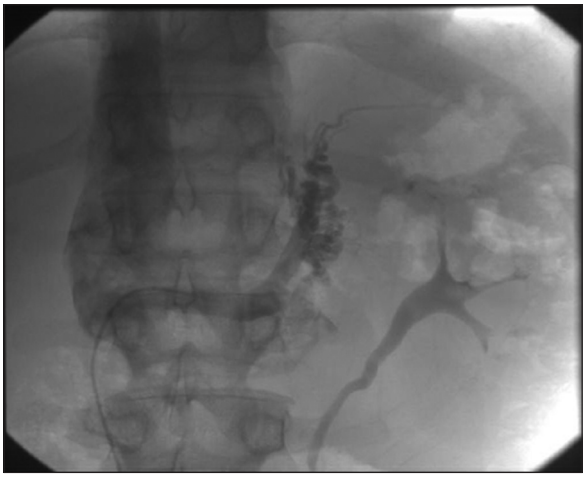

Figure 6: Diagnostic phlebography (left) showed dilation of the left spermatic vein

is known as anterior nutcracker. The retroaortic renal vein may be compressed between the aorta and the vertebral body, which is called posterior nutcracker. NCS with LRV Duplication is exceptional, only one case of LRV duplication with a dilated retroaortic branch, entrapped between the aorta and the vertebral column was described as posterior NCS with renal vein duplication;[4] Here we present the first case of mixed NCS with renal vein duplication combining dilated retroaortic and preoaortic branchs, entrapped respectively between the aorta and the vertebral column and in the AMS and the aorta (aortico-mesenteric space).

Epidemiologically, the NCS occurs more frequently in women and is diagnosed within the 3rd and 4th decade of life; our case however was in an 18-year-old male. Its exact incidence and prevalence is difficult to define since the majority of patients are asymptomatic and are diagnosed incidentally on imaging ordered for other reasons.

When the diagnosis is suspected, the detection of the NCS can be made by several newer imaging modalities; such as CTA and magnetic resonance angiography that have powerful abilities in providing accurate, three-dimensional reconstructive images in any desired plane and allowing better identification of these anomalies. Spectral Doppler ultrasound can give more details of the hemodynamics of the LRV at the site of compression.[5]

In the face of a strong suspicion, performing a cystoscopy in the acute phase with visualization of unilateral left hematuria.

With regard to more invasive investigations, retrograde venography and video angiography to determine the

renocaval pressure gradient will give a precise diagnosis of the compression. Venography will demonstrate the area where the compression is, the existence of collateral circulation in periureteral vessels, reflux into the renal vein branches and the stagnation of contrast in the renal vein.[6]

After confirming the diagnosis of NCS different treatment alternatives can be chosen taking into account the symptoms of each individual patient and the morphological features of the NCS.

Treatment of the NCS is controversial and there are several available options, including follow-up, conservative treatment and surgical therapy. Endoscopic proceedings include external or internal stenting.[7] Open surgical procedures include the transposition of LRV[8] transposition of the SMA[9] renal auto transplantation[10] gonadocaval bypass[11] or nephrectomy when there is severe organ damage.[12] Recently, NCS was treated with a retroperitoneal laparoscopic nephrectomy with *ex vitro* autograft repair and auto transplantation[13] and abdominal aortic transposition.[14]

In our case, the suitable option is transperitoneal nephrectomy with *ex vitro* autograft repair and auto transplantation because of the complex morphological features of this mixed NCS. This option was refused by the patient and only treatment of varicocele by antegrade radiologic embolization was accepted but was technically impossible because of the difficulty to cannulate the branchs of the LRV and the collateral veins. The treatment of varicocele by the retroperitoneal approach (palomo technic) was failed with early recurrence that we explain by the existence of several collateral veins of the spermatic vein no ligated. Inguinal varicocelectomy was successful without recurrence of varicocele at 4 months of follow-up. Our patient still suffer of the initial presentation with abdominal and pelvic pain and fatigue, with daily use of analgesics.

CONCLUSION

We present here a case of an 18-year-old boy with severe abdominal pain promoted by an antero-posterior NCS. Our case report shows an exceptional cause and presentation of a NCS. This is a rare entity, with diverse presentations that provides a variety of signs and symptoms. This evokes the necessity of a heightened sense of alert when investigating abdominal pain in children and adolescents in order to achieve a correct and attempted diagnosis.

REFERENCES

1. El-Sadr AR, Mina E. Anatomical and surgical aspects in the operative management of varicocele. Urol Cutaneous Rev 1950;54:257-62.
2. Shin JI, Park JM, Lee SM, Shin YH, Kim JH, Lee JS, *et al*. Factors affecting spontaneous resolution of hematuria in childhood nutcracker syndrome. Pediatr Nephrol 2005;20:609-13.
3. Rudloff U, Holmes RJ, Prem JT, Faust GR, Moldwin R, Siegel D. Mesoaortic compression of the left renal vein (nutcracker syndrome): Case reports and review of the literature. Ann Vasc Surg 2006;20:120-9.
4. Preza Fernandes J, Amorim R, Gomes MJ, Oliveira V, Reis A, Ribeiro-Castro J. Posterior nutcracker syndrome with left renal vein duplication: A rare cause of haematuria in a 12-year-old boy. Case Rep Urol 2012;2012:849681.
5. Luo XL, Qian GN, Xiao H, Zhao CL, Zhou XD. Posterior nutcracker syndrome associated with interrupted left inferior vena cava with azygos continuation and retroaortic right renal vein. Korean J Radiol 2012;13:345-9.
6. Ferreira M, Lanziotti L, Abuhadba G, Monteiro M, Capotorto L, Spicacci JL. Chronic pelvic pain: The role of the nutcracker syndrome. J Vasc Bras 2008;7:76-9.
7. Barnes RW, Fleisher HL 3rd, Redman JF, Smith JW, Harshfield DL, Ferris EJ. Mesoaortic compression of the left renal vein (the so-called nutcracker syndrome): Repair by a new stenting procedure. J Vasc Surg 1988;8:415-21.
8. Stewart BH, Reiman G. Left renal venous hypertension "nutcracker" syndrome. Managed by direct renocaval reimplantation. Urology 1982;20:365-9.
9. Thompson PN, Darling RC 3rd, Chang BB, Shah DM, Leather RP. A case of nutcracker syndrome: Treatment by mesoaortic transposition. J Vasc Surg 1992;16:663-5.
10. Chuang CK, Chu SH, Lai PC. The nutcracker syndrome managed by autotransplantation. J Urol 1997;157:1833-4.
11. d'Archambeau O, Maes M, De Schepper AM. The pelvic congestion syndrome: Role of the "nutcracker phenomenon" and results of endovascular treatment. JBR-BTR 2004;87:1-8.
12. Russo D, Minutotto R, Iacavino V. Gross hematuria of un-unknown origin: The nutcracker syndrome. Am J Kidney Dis 1998;32:23-9.
13. Xu D, Liu Y, Gao Y, Zhang L, Wang J, Che J, *et al*. Management of renal nutcracker syndrome by retroperitoneal laparoscopic nephrectomy with *ex vivo* autograft repair and autotransplantation: A case report and review of the literature. J Med Case Rep 2009;3:82.
14. Li X, Ji C, Guo H. Abdominal aortic transposition as a treatment alternative for posterior nutcracker syndrome. Int J Urol 2012;19:1043-4.

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