Nutcracker syndrome in a 20-year-old patient treated with intravascular stent placement: a case report

Louis-Olivier Gagnon, MD,[1] Yves Ponsot, MD,[1] Andrew Benko, MD,[2] Le Mai Tu, MD[1]

[1]Department of Urology, University of Sherbrooke, Sherbrooke, Quebec, Canada
[2]Department of Diagnostic Radiology, University of Sherbrooke, Sherbrooke, Quebec, Canada

GAGNON L-O, PONSOT Y, BENKO A, TU LM. Nutcracker syndrome in a 20-year-old patient treated with intravascular stent placement: a case report. The Canadian Journal of Urology. 2009;16(4):4765-4769.

Introduction: *Nutcracker syndrome (NCS) is a rare condition characterized by the entrapment of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the aorta. It was first described in 1950 and the term nutcracker was attributed by de Schepper in 1972.*
Clinical case: *A 20-year-old female patient was admitted to the urology department with a history of hematuria and left flank pain. Basic hematuria investigation was inconclusive. Further investigation revealed a nutcracker syndrome on retrograde phlebography. Surveillance was the option first chosen. Three years later and after several symptomatic episodes, an intravascular stenting procedure was performed.*
Discussion: *NCS is defined by the compression of the LRV between the SMA and the aorta. Clinical manifestations include left flank pain, hematuria and pelvic congestion syndrome (PCS). Diagnosis is based on history, physical examination, basic lab tests and imaging. Sequence of tests can be composed of Doppler ultrasound (DUS), computed tomography scan (CT scan) or magnetic resonance imaging study (MRI) and retrograde phlebography with pressure gradient to confirm the diagnosis. Management options include surveillance, intravascular and extravascular stenting procedures as well as open procedures.*
Conclusion: *NCS is a rare condition that represents a challenge for urologists in terms of accurate diagnosis and proper management.*

Key Words: interventional radiology, stent

Clinical case

A 20-year-old, otherwise healthy female patient was admitted to the urology department with a history of gross hematuria with clots present for 24 hours as well as mild left flank pain for 2 weeks. This was her third episode in 5 years. There were no associated symptoms; no lower urinary tract symptoms, no fever, no gastrointestinal symptoms.

The physical examination was noncontributory; all vital signs stable, no finding on abdominal exam, negative renal punch bilaterally.

Laboratories included a complete blood count (CBC) which showed a normal white blood cell count (WBC) and an hemoglobin of 123 g/l. The electrolytes and renal function tests were normal. The urinalysis demonstrated > 100 red blood cells (RBC). The urine culture was negative.

Accepted for publication June 2009

Address correspondence to Dr. Louis-Olivier Gagnon, Department of Urology, University of Sherbrooke, 3001, 12e Avenue Nord, Sherbrooke, Quebec J1H 5N4 Canada

A basic macroscopic hematuria work up was done including a renal ultrasound and a computed tomography scan (CT scan) which only showed a left 1.5 cm simple renal cyst. A cystoscopy was performed and came out normal.

Further investigation was done, in light of these normal results, in order to find the source of this recurrent bleeding. An arteriography demonstrated normal arterial findings, but interestingly showed a compression of the left renal vein (LRV) under the superior mesenteric artery (SMA) on the venous phase, compatible with a nutcracker syndrome (NCS). A retrograde phlebography was then performed showing a LRV stenosis with a pressure gradient of 4 mmHg, the pressure in the LRV proximal to the stenosis being 4 mmHg and the pressure in the inferior vena cava (IVC) being 0 mmHg, Figure 1. Revision of the CT scan, previously reported as noncontributory, revealed a narrowing of the LRV suggestive of a NCS, Figure 2.

With these results in hands, the diagnosis of NCS was made. The initial option chosen by the medical team and the patient was surveillance, mostly because the patient was not bothered enough by her symptoms to warrant an intervention.

From the time of diagnosis to 3 years after, the patient consulted several times. The severity and frequency of the episodes increased progressively.

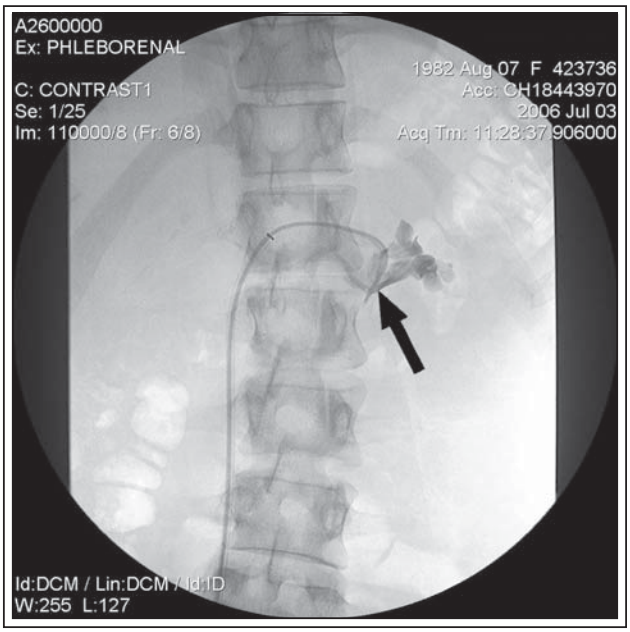

Figure 1. Phlebography demonstrated the compression on the LRV (arrow) causing a decreased flow, with a measured pressure gradient of 4 mmHg.

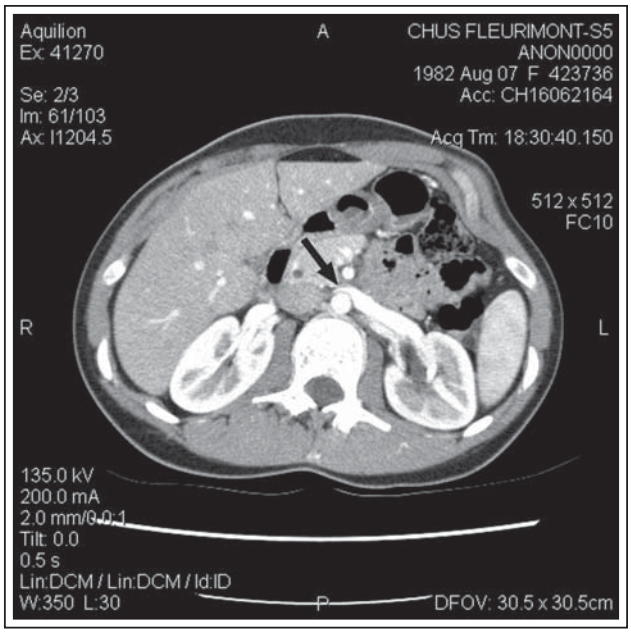

Figure 2. Revision of the CT scan, previously reported as noncontributory, revealed a narrowing of the LRV (arrow) suggestive of a NCS.

Three years after diagnosis, the patient decided to proceed with intravascular stent placement. Using a right femoral approach, a 14 mm x 40 mm nitinol stent was placed in the LRV and subsequently dilated with a 10 mm balloon. Phlebography done after stent placement demonstrated good stent position and a widely patent LRV, with a measured pressure gradient of 0 mmHg, the pressure in the LRV being 1 mmHg and the pressure in the IVC being 1 mmHg, Figure 3.

However, 2 months postop, the patient experienced a recurrence in symptoms. A phlebography done at that moment showed the stent displaced in the IVC, with impossibility of removing the stent through endovascular measures. A multidisciplinary discussion involving vascular surgery, interventional radiology and urology concluded that the stent should be left in place, the surgical risks outweighing the benefits, and a second stent should be installed in the LRV. The procedure was performed and well tolerated, Figure 4.

Now 2 years after this last procedure, the patient is doing well, without recurrence of pain or hematuria. Radiological monitoring done with an abdominal x-ray and a DUS shows a normal position of the stent in the LRV on the x-ray and a normal LRV flow on the ultrasound. The plan is to follow her clinically and radiologically on a yearly basis.

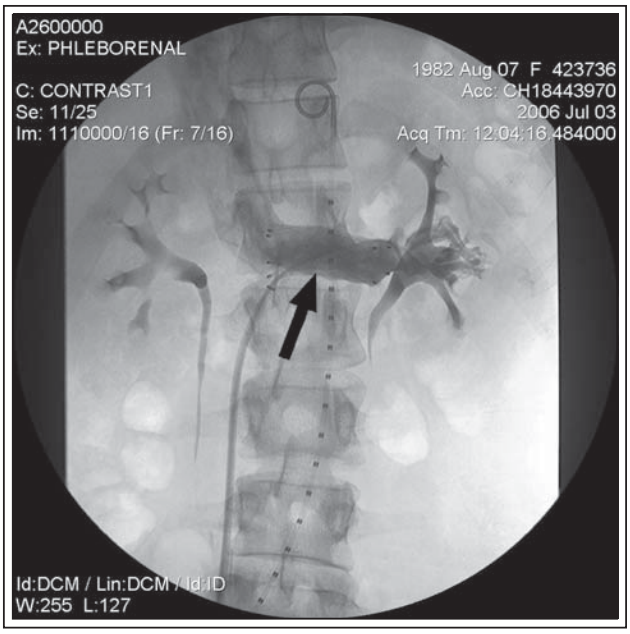

Figure 3. Phlebography done after stent placement (arrow) demonstrated good stent position and a widely patent LRV, with a measured pressure gradient of 0 mmHg.

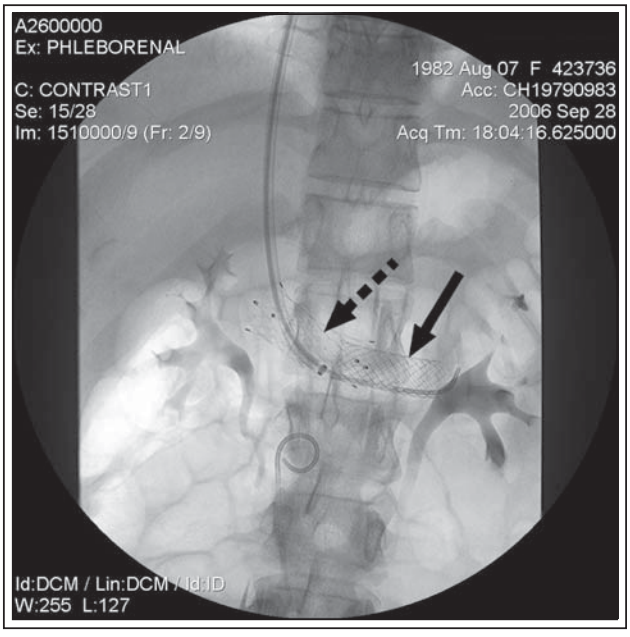

Figure 4. Phlebography done after the installation of a second stent in the LRV showed the normal position of the second stent in the LRV (arrow) and the abnormal position of the first stent displaced in the IVC (dotted arrow).

Discussion

NCS is a rare condition characterized by the entrapment of the LRV between the SMA and the aorta. It was first described in 1950 by El Sadr.[1] In 1972, de Schepper published a report on a 16-year-old boy with macroscopic hematuria in which he gave the name 'nutcracker syndrome' to the disease. [2] De Schepper believed the LRV between the aorta and the SMA resembled a nut between the jaws of a nutcracker.

NCS is a rare condition with only a few reported cases. It is more prevalent in the female community and affects more commonly thin and tall patients aged between 20 to 40 years old.[3]

NCS is classified as either anterior or posterior. Anterior NCS is the most common and is defined by compression of the LRV between the SMA and the aorta. The normal angle between the SMA and the aorta is approximately 90 degrees and the SMA usually courses 4-5 mm ventrally, Figure 5.[4] In anterior NCS, a configuration abnormality with the origin of the SMA causes the SMA to have an initial steep caudal descent as well as an acute angle with the aorta. As for posterior NCS, it is defined by the retro-aortic course of the LRV causing a compression of the LRV between the aorta and the vertebral column.

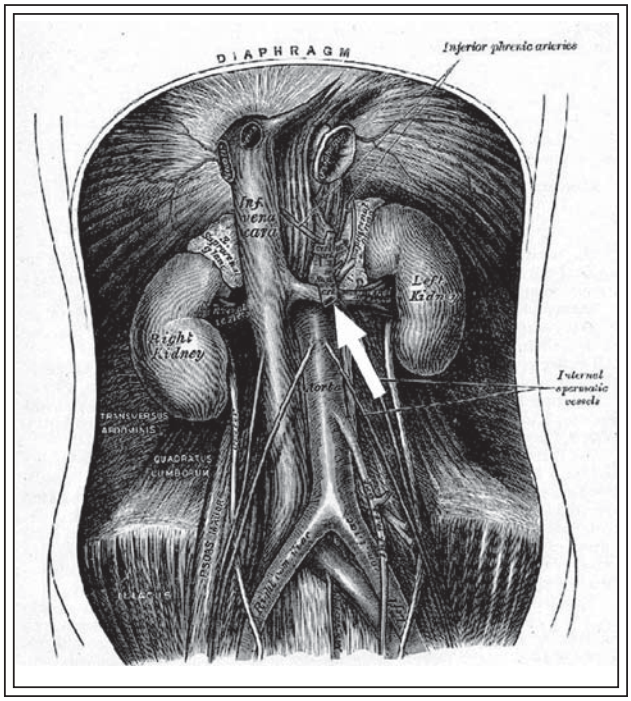

Figure 5. Anatomy of the vasculature of the retroperitoneum showing the normal course of the LRV between the SMA and the aorta. (arrow).

Common manifestations for NCS include left flank pain, usually associated with the passage of clots, and hematuria, either microscopic or macroscopic, caused by the rupture of renal veins in the collecting system. Alternate manifestation is the 'pelvic congestion syndrome' (PCS). It consists of pelvic pain, dyspareunia, dysmenorrhea, vulvar varices, varicoceles, due to left gonadal vein reflux.

Another diagnosis to consider in the case of unexplained recurrent flank pain and hematuria is the loin pain hematuria syndrome (LPHS). It is characterized by recurrent flank pain, either unilateral or bilateral, hematuria and occasional low-grade fever. LPHS is a rare condition that has no known etiology and therefore remains a diagnosis of exclusion. Renal autotransplantation with renal denervation has been reported as an effective procedure for the most severe cases.[5]

Work up for NCS includes a series of tests such as Doppler ultra-sound (DUS), CT scan or magnetic resonance imaging (MRI) angiography and retrograde phlebography.[6] DUS is the initial diagnostic test. Sensitivity is 78% and specificity 100%. Measurements include anteroposterior diameter and peak velocity at two points. CT and MRI angiography are of interest for the anatomic delineation of the NCS. As for retrograde phlebography, it is the gold standard and is used to confirm the diagnosis of NCS. It allows visualization of the point of compression on the LRV. Diagnosis is made with a pressure gradient measurement of > 3 mmHg between the LRV proximal to the stenosis and the IVC.

Management options include surveillance, intravascular and extravascular stenting procedures as well as open procedures. Surveillance constitutes a valuable choice in selected cases with mild symptoms, such as microscopic hematuria and short periods of painless gross hematuria. A close follow up is required in this situation.

Stent placement, either intravascular or extravascular, represents an alternative to surveillance in more severe cases. The intravascular stenting procedure corresponds to placement of an expandable metallic stent under angiography. Neste et al first reported the intervention in 1996. [7] It is simple, minimally invasive and efficient, according to Zhang et al.[8] In 2001, they compared three patients with SMA transposition and three patients with endovascular stent placements, on 4 to 54 months follow up. All the patients became asymptomatic. In regards to complications, they include fibromuscular hyperplasia as well as proximal migration. As for the extravascular stenting procedure, it corresponds to ring forced polytetrafluoroethylene (PTFE) graft placement around LRV by either open or laparoscopic technique. Barnes et al. first reported the intervention in 1988.[9] Scultetus et al reported the lap-assisted variant in 2001.[10]

The last alternative in refractory cases is open surgery. In 1974, Pastershank described the first open surgery for the treatment of NCS as he practiced a fibrous tunnel division.[11] Also, LRV transposition has been practiced numerous times and has a good outcome according to a study done by Hohenfellner et al in 2002.[12] It consists of the re-anastomosis of the LRV with the IVC at a lower level. Other options for open surgery include renal autotranplantation, SMA transposition and anterior nephropexy.

Conclusion

NCS is a rare condition that represents a challenge for urologists in terms of accurate diagnosis and proper management. Diagnosis is based on history, physical examination, basic lab tests and imaging. Sequence of tests can be composed of Doppler ultrasound, CT or MRI angiography and retrograde phlebography with pressure gradient to confirm the diagnosis. Management options include surveillance, intravascular and extravascular stenting procedures as well as open procedures. Stenting procedures represent promising treatment options for NCS. □

References

1. El Sadr AR, Mina A. Anatomical and surgical aspects in the operative management of varicoceles. *Urol Cut Rev* 1950;54(5): 257-262.
2. de Schepper A. Nutcracker fenomeen van da vena renalis en veneuz pathologie van de linker nier. *J Belge Radiol* 1972;55: 507-511.
3. Gorospe EC, Aigbe MO. Nutcracker syndrome: a rare cause of hematuria. *Scientific World Journal* 2006; 30;6:745-746.
4. Gray H. Anatomy of the human body. Bartleby.com. 2000.
5. Dube GK, Hamilton SE, Ratner LE, Nasr SH, Radhakrishnan J. Loin pain hematuria syndrome. *Kidney Int* 2006;70(12): 2152-2155.
6. Ahmed DK, Sampath R, Khan MS et al. Current trends in the diagnosis and management of renal nutcracker syndrome: a review. *Eur J Vasc Endovasc Surg* 2006;31(4):410-416.
7. Neste MG, Narasimham DL, Belcher KK. Endovascular stent placement as a treatment for renal venous hypertension. *J Vasc Interv Radiol* 1996;7(6):859-861.
8. Zhang HK, Shen LG, Li M et al. Diagnosis and treatment of left renal venal entrapment syndrome. *Chin J Gener Surg (Chin)* 2001;16:511.
9. Barnes RW, Fleisher HL III, 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(4):415-421.

10. Scultetus AH, Villavicencio JL, Gillespie DL. The nutcracker syndrome: its role in the pelvic venous disorders. *J Vasc Surg* 2001;34(5):812-819
11. Pastershank SP. Left renal vein obstruction by a superior mesenteric artery. *J Can Assoc Radiol* 1974;25(1):52-54.
12. Hohenfellner M, D'Elia G, Hampel C et al. Transposition of the left renal vein for treatment of the nutcracker phenomenon: long-term follow-up. *Urology* 2002;59(3):354-357.

EDITORIAL COMMENT

The authors present an interesting case of a 20-year-old female patient who was diagnosed with Nutcracker Syndrome. This condition, characterized by left flank pain, hematuria and pelvic congestion syndrome, is often difficult to diagnose. The possibility that this condition has an origin in the bladder is certainly possible as a rare presentation of Interstitial Cystitis/Painful Bladder Syndrome. Symptoms can wax and wane for many years prior to diagnosis. Additionally, patients may present with signs of narcotic addiction and psychiatric disturbances such as anxiety and depression. A complete evaluation of the upper and lower urinary tract is mandatory to rule out other causes of hematuria, particularly small renal masses and obstructing stones in caliceal diverticuli.

Treatment for Nutcracker Syndrome can be challenging. The authors describe their success with intravascular stent placement. However, open surgery may be required for refractory cases. Our experience with similar patients presenting with Loin Pain Hematuria Syndrome indicates that additional benefit can be obtained with renal autotransplantation. This procedure was performed in patients who failed conservative management with surveillance and oral analgesics. In those patients, long term relief was achieved with renal auto-transplantation.[1]

References

1. Pukenas BA, Zaslau S. Loin Pain Hematuria syndrome: case series. *WV Med J* 2003;99(5):192-1993.

Stanley Zaslau, MD, MBA, FACS
Associate Professor
Program Director
Division of Urology
West Virginia University
Morgantown, West Virginia