

Case Report

Endovascular stenting for nutcracker syndrome

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Abstract

Nutcracker syndrome (NCS) is a rare pathology manifested by pain or hematuria in males and females alike. It can be easily overlooked, and should be considered in young men or women with symptoms of extended duration. We present a case of a 54-year-old female with chronic lower abdominal pain radiating to the left thigh of 4 years in duration. Computed tomography (CT) eventually revealed engorged left renal, gonadal, and uterine veins due to compression between the superior mesenteric artery (SMA) and the abdominal aorta, consistent with NCS. After a successful endovascular stenting and a 6-month period of antiplatelet and anticoagulant therapy, the patient returned to stable health. NCS, while rare, should be suspected in patients of both sexes with persistent pain or hematuria. Copyright © 2013 Elsevier Taiwan LLC and the Chinese Medical Association. All rights reserved.

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1. Introduction

Nutcracker syndrome (NCS), or renal vein entrapment syndrome, is a rare and easily overlooked condition. It is characterized by external compression of the outflow from the left renal vein (LRV) into the inferior vena cava (IVC). Most often, it implies compression of the LRV between the aorta and the superior mesenteric artery (SMA), known as the anterior nutcracker. It may coincide with SMA syndrome—compression of the third portion of the duodenum by the abdominal aorta and the SMA.¹ The retro-aortic renal vein may be compressed between the aorta and the vertebral body, a condition known as posterior nutcracker.² The relationship between these structures is shown in Fig. 1. The LRV may also be compressed by nearby neoplasms, lymphadenopathy, or an enlarged abdominal aortic aneurysm. Endovascular technology may provide minimally invasive therapy to relieve the

symptoms of this compression syndrome. Herein, we report a case of successful stenting to treat NCS.

2. Case report

A 54-year-old female complained of frequent lower abdominal pain radiating to the left thigh for 4 years. Her gynecological record was pregnant 4 times, given birth 4 times (G4P4) with menopause 3 years previously. She denied any systemic disease, but had undergone a laparoscopic cholecystectomy procedure 7 years previously. During the past 4 years, the patient had several times visited local clinics and our hospital, including the emergency department, to address her abdominal pain. Physical examination showed lower abdominal tenderness, and laboratory examination disclosed mild anemia (hemoglobin ranging 10–13 g/dL) and several instances of microscopic hematuria.

The patient was referred to our colorectal and gastrointestinal clinic for further evaluation because of chronic symptoms. Colonoscopy revealed no abnormal findings; an upper gastrointestinal endoscopy showed only gastritis. Our neurological clinic was also consulted because abdominal pain

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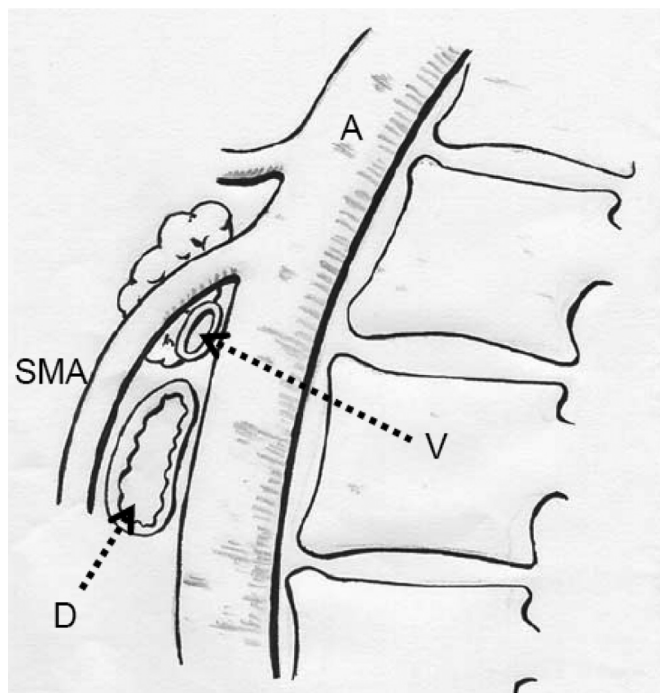


Fig. 1. The relationship between the abdominal aorta, SMA, LRV (V), and duodenum (D). LRV = left renal vein; SMA = superior mesenteric artery.

occasionally radiated to the left thigh, and magnetic resonance imaging (MRI) of the spine showed a bulging disc at the level of L5-S1, with mild dural sac compression. The patient was then transferred to the gynecologic clinic where transvaginal sonography showed suspected endometrial hyperplasia and fluid accumulation over the cul-de-sac. Pelvic congestion syndrome was diagnosed and conservative treatment was suggested. As symptoms failed to improve 2 months after medical treatment, the patient received multi-detector computed tomography (MDCT). It showed that the LRV was compressed between the superior mesenteric artery (SMA) and the abdominal aorta, with engorged left gonadal and uterine veins (Fig. 2A and B). The diameter of the LRV was 2.5×3.6 mm at its most compressed level, and 19.4×16.7 mm at its most dilated level; the diameter of the proximal gonadal vein was 7.6×7.4 mm. The patient was diagnosed with NCS and admitted for further intervention.

After admission, blood analysis showed normal white blood count, platelet count, and biochemistry, but mild anemia. Urine analysis was normal. A cardiovascular surgeon was consulted and surgical intervention was suggested. After being told the risks and benefits of both surgical and endovascular therapeutic alternatives, the patient agreed to undergo endovascular stenting. Under general anesthesia, the patient was placed supine on the sterilized and draped operative field. The right common femoral vein was gently punctured with an 18-gauge needle, a guidewire was inserted, followed by a 10-Fr/10-cm sheath. Heparin (3000 U) was administered intravenously, and activated clotting time checked hourly to ensure it remained above 200 seconds. A 0.035-inch/180-cm hydrophilic guidewire (Terumo Corporation, Tokyo, Japan) was

inserted into the IVC and the venography of the IVC was obtained via a pigtail catheter. The catheter and sheath were then exchanged for an 8-French (Fr)/55-cm Mach1 peripheral catheter (Boston Scientific Inc., Natick, MA, USA). The catheter was advanced to the junction of the IVC and LRV, then the LRV was cannulated with a Terumo 0.035-inch guidewire, followed by a CHG 2.5 and Vanschie 2 catheter (Cook Medical Inc., Brisbane, Australia).

A selective left renal venogram was performed, which identified abundant large collaterals of the left adrenal and renolumbar veins. A 0.035-inch Amplatz super stiff wire (Boston Scientific Inc., Natick, MA, USA) was advanced into the LRV after the catheter and the long sheath was advanced, followed by the dilator. The stenosis lesion was pre-dilated with an 8.0 mm \times 80 mm balloon (Wanda standard; Boston Scientific Inc., Natick, MA, USA). The lesion length was measured with the balloon and a 14 mm \times 4 cm stent (Zilver 635 self-expanding stent; Cook Medical Inc., Brisbane, Australia) was advanced and deployed. The venography was checked again and revealed a widened LRV with decreased collateral filling. The whole procedure was tolerated well by the patient. The duration of fluorography was 33 minutes, the dosage was 232 mGy, and 50 mL of contrast volume was used.

After the procedure, we arbitrarily chose to treat this patient with aspirin (100 mg/day), clopidogrel (Plavix, 75 mg/day) and 1-week low-molecular-weight heparin (Clexane, 0.2 cm³ subcutaneously twice/day). No postoperative complications occurred, and computed tomography (CT) on the 3rd day postoperatively showed the patient's stent had remained immobile, without migration (Fig. 2C and D). With stable vital signs, the patient was discharged on a 6-month regimen of aspirin (100 mg/day), clopidogrel (Plavix, 75 mg/day), and coumadin (Warfarin, 2.5 mg/day), and thereafter monthly followed up at the outpatient department.

3. Discussion

NCS is an infrequently seen and easily overlooked condition. The severity of this syndrome varies from the virtually asymptomatic cases, sometimes involving microscopic hematuria, to gross hematuria and severe pelvic congestion.^{3,4} NCS should be considered in young men or women with symptoms of extended duration during our practice.⁵ In this case, microscopic hematuria was found several times, but this symptom could not be linked to the patient's abdominal pain, the two most common symptoms of NCS. Consequently, we had not yet formally diagnosed NCS, and the syndrome remained. Varicocele is another common symptom, and the LRV is compressed in more than half of those patients with varicocele.^{6,7} Hence, NCS should be routinely excluded as a possible cause of varicocele.

Diagnosis of NCS, both challenging and commonly delayed, requires a high index of suspicion and can be accomplished with Doppler sonography, CT, MRI, or retrograde phlebography. It is confirmed by a pressure gradient of >3 mmHg across the lesion.^{5,8,9} Doppler ultrasonography or MDCT may be used as the initial diagnostic test in patients with suspected NCS. Doppler ultrasonography can assess the

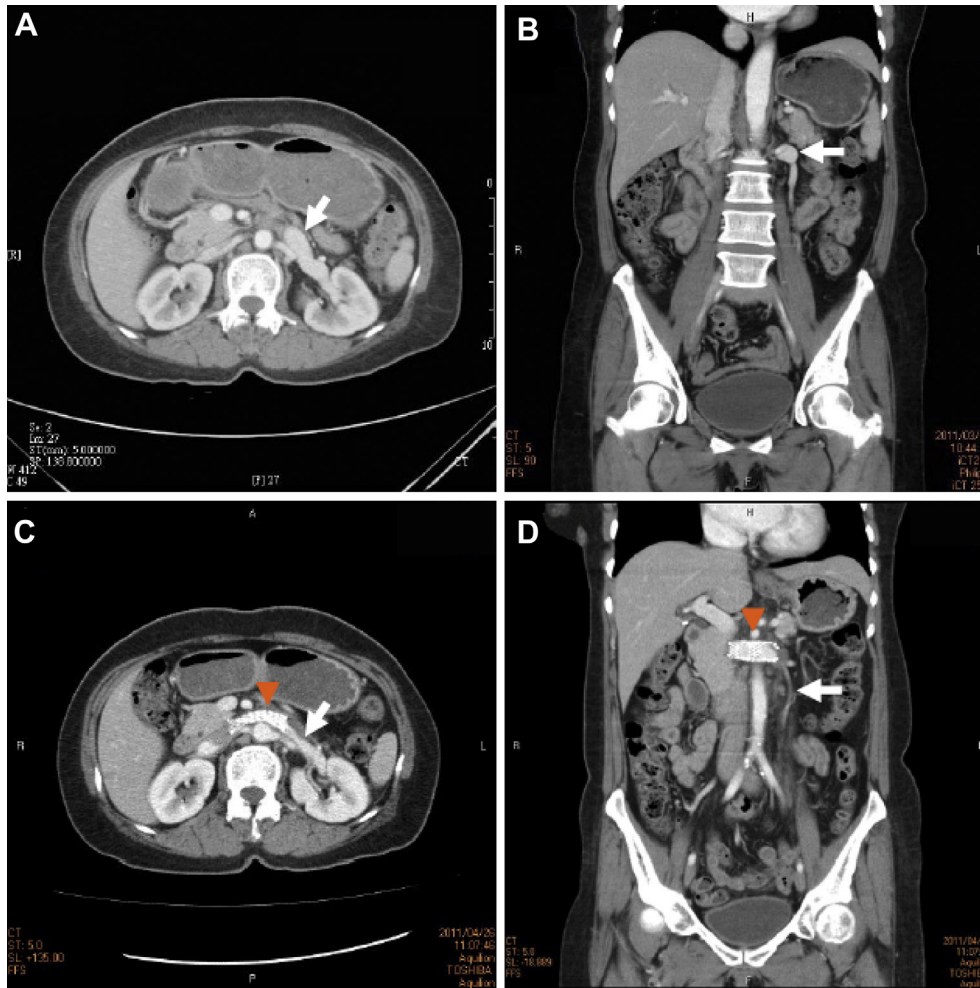


Fig. 2. CT images of the LRV before and after endovascular stenting. Engorged left renal and gonadal veins (white arrow) in (A) and (B) before stenting. LRV and gonadal veins with decreased diameter (white arrow) in (C) and (D). The orange arrow head shows the stent. CT = computed tomography; LRV = left renal vein.

antero-posterior diameter and peak velocity of the LRV in two places: (1) at the level of the renal hilum; and (2) at the point where the LRV crosses between the aorta and the SMA. Diagnosis of nutcracker phenomenon should be considered in cases in which the antero-posterior diameter and peak velocity at these two points exceeds the norm by a factor of four.^{10,11} In this case, we did not conduct pre-operative Doppler sonography or an intra-operative hemodynamic study because MDCT provided obvious anatomical confirmation (compression of LRV and varicose veins) of NCS.

Traditionally, persistent or anemizing hematuria, lumbar or incapacitating pelvic pain, or the presence of severe congestive pelvic symptoms are treatment indications for NCS, with the exclusion of other differential diagnoses.^{12,13} A variety of surgical approaches have been developed to decrease LRV hypertension or pelvic venous reflux, and these have been well reviewed by Menard et al.¹³ A report of 23 cases by Wang et al shows that LRV transposition is an efficient surgical approach with an acceptable risk of complications. However, in some cases, pelvic pain may persist despite removal of the obstruction to the renal venous backflow.¹⁴ Intravascular stenting to treat NCS using expandable metallic stents was first reported by

Neste et al.¹⁵ Sporadic cases of intravascular stenting have since been reported using a variety of stents, but with relatively short follow-up. This patient may be the first reported case of NCS treated with endovascular stenting in Taiwan. Currently, percutaneous angioplasty and stenting for symptomatic LRV compression for NCS is an excellent, effective, and safe alternative to surgery.¹⁶ As yet, no consensus has been reached concerning the optimal size and type of stent (balloon-expandable or self-expanding). Chen et al have shown, in a longer follow-up of 61 patients, that endovascular stenting is a primary option for NCS.¹⁷ However, further follow-up and investigation will be required to determine if the durability of this approach compares favorably to current open options, because of concerns including stent migration,^{12,18} thrombosis,¹⁹ deformity, and erosions. Another issue is the danger associated with the post-stenting use of anticoagulants and antiplatelets.

In conclusion, NCS is a rare pathology manifested by pain with or without microscopic or gross hematuria. Ultimately, NCS should be suspected in young men or women with symptoms of lengthy duration, and endovascular stenting for symptomatic LRV compression may be an effective and safe treatment.

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