Left renal vein transposition for posterior Nutcracker syndrome

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ABSTRACT

Nutcracker syndrome (NCS) is a rare cause of pelvic venous congestion syndrome and is secondary to either compression of the left renal vein in its normal anatomic position by the superior mesenteric artery and aorta or less commonly when the left renal vein is in a retroaortic position, compressed between the aorta and the spine. We herein present a unique case of NCS in a female patient with a history of chronic pelvic pain and venous congestion. We also review the literature and discuss the diagnostic modalities, differential diagnosis, and various open surgical and endovascular options for NCS. (J Vasc Surg Cases and Innovative Techniques 2021;7:243-6.)

Keywords: Posterior Nutcracker syndrome; Pelvic congestion; Left renal vein transposition

Nutcracker syndrome (NCS) can be divided into two types: anterior and posterior. The presentation can range from asymptomatic hematuria to symptomatic flank pain, pelvic congestion (females), varicocele enlargement (males), and renal vein thrombosis. Anterior NCS (aNCS) accounts for the majority of cases and is characterized by the left renal vein (LRV) in its normal anatomic position compressed between the superior mesenteric artery and the aorta. Posterior NCS (pNCS) is an uncommon anatomic variant in which the LRV courses posterior to the aorta and is compressed between the aorta and the lumbar vertebra. The incidence of retroaortic LRV is rare (<2%), and the subsequent development of NCS is exceedingly rare with fewer than 30 documented cases in the literature.[1,2]

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CASE REPORT

A 30-year-old woman with a history of ulcerative colitis presented with chronic and worsening pelvic and left flank pain. Physical examination revealed mild tenderness to palpation in the left lower quadrant without lower extremity edema or varices. She denied gross hematuria, but reported a recent episode of tea-colored urine right before her admission. Laboratory values included a normal creatinine (0.72 mg/dL) and normocytic anemia (hemoglobin of 10.7 g/dL; mean corpuscular volume of 89.5 fL). Urinalysis was negative for red blood cells. A computed tomography (CT) scan demonstrated a peripherally dilated retroaortic LRV (Fig 1). A pelvic venogram with isolated LRV and ovarian vein cannulation demonstrated reflux into the dilated left ovarian vein. With and without Valsalva, contrast refluxed into the left ovarian vein with opacification across pelvic collaterals and uterine plexus with subsequent filling of the right ovarian vein and the inferior vena cava (IVC; Fig 2). Pullback pressure gradient from the peripheral LRV to the IVC measured greater than 10 mm Hg.

A midline transperitoneal approach was used to expose the infrarenal aorta. Mobilization of the aorta to allow full exposure of the LRV required ligation of multiple lumbar arteries. The LRV was identified in a significantly caudal trajectory. The exposure and control of the IVC, LRV, and aorta before transposition is shown in Fig 3. The LRV was transected at its confluence with the IVC and the venotomy was closed with two-layer running 4-0 monofilament suture. The LRV was then transposed to a more cranial position and an end-to-side anastomosis was performed with 6-0 monofilament (Fig 4). The left gonadal vein was subsequently ligated.

The patient recovered without complication and was discharged home on postoperative day 5 with 81 mg acetylsalicylic acid. At the 2-month follow-up, the patient had returned to work and full activity. She was pain-free and tolerating a normal diet.

DISCUSSION

NCS is an uncommon cause of abdominal and pelvic pain. This is partly due to the vague compilation of symptoms (pelvic and/or flank pain, hematuria, gonadal varices, pelvic congestion). Much of the literature describing NCS is limited to case reports and small series. Because of its infrequent description in the literature, the optimal treatment of pNCS is uncertain.

Definitive diagnostic methods consist of doppler ultrasound examination, CT scan, magnetic resonance

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Fig 1. Computed tomography scan demonstrating retroaortic left renal vein (LRV) position.

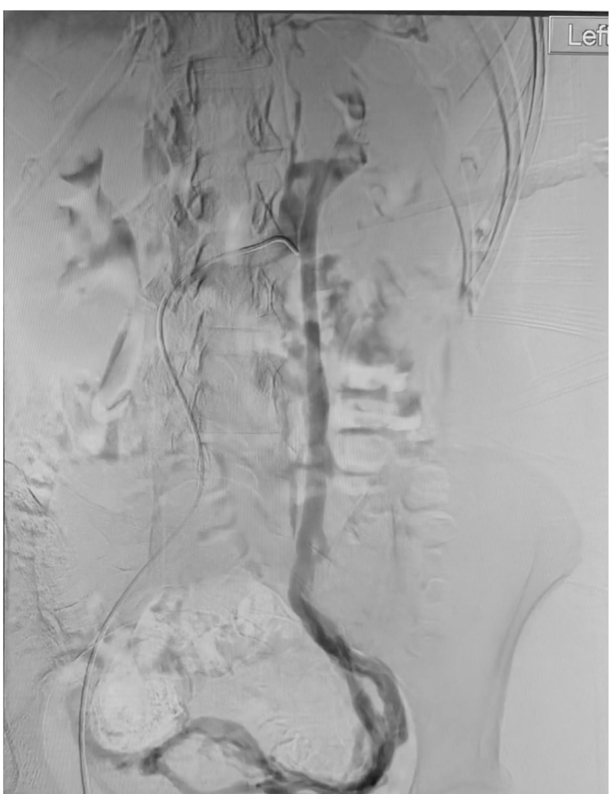

Fig 2. Left renal vein (LRV) venography with reflux into left gonadal vein and across uterine venous plexus.

imaging, venography, and intravascular ultrasound (IVUS).[3] Results from doppler ultrasound examination can have high sensitivity and specificity (69%-90% and 89%-100%, respectively); however, results are limited by patient body habitus and experience of the ultrasonographer. Both CT scan and magnetic resonance imaging

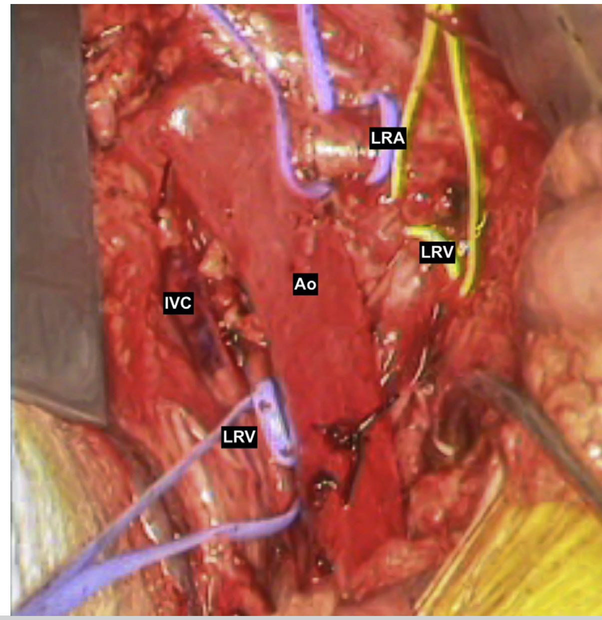

Fig 3. Predivision anatomy. *Ao*, Aorta; *IVC*, inferior vena cava; *LRA*, left renal artery; *LRV*, left renal vein.

assess the LRV for degree of compression and surrounding anatomy. CT criteria include compression ratio of more than 2.25 (diameter compressed LRV/diameter non-compressed LRV), a beak angle of less than 32°, a LRV diameter ratio (hilar to aortomesenteric) of more than 4.9°, and the angle between SMA and aorta of less than $<$ 41°.[4,5] Venography with IVUS is considered the gold standard for diagnosis. Renocaval pullback pressure gradient is frequently used during venography. A normal pressure gradient (LRV minus IVC) is less than 1 mm Hg, whereas a difference of greater than 3 mm Hg is diagnostic for NCS. When comparing these two methods, IVUS demonstrates superior specificity over venography alone (90% vs 62%) as a result of the wide variation of observed renocaval pullback pressure gradients seen in both the healthy and NCS populations.[4,6]

Females with NCS are occasionally diagnosed with pelvic congestion syndrome (PCS). Both syndromes cause increased venous pressure in the pelvic vessels resulting in overlapping symptoms and often display pelvic varices on three-dimensional imaging.[7] PCS is described in the obstetric and gynecologic literature in the setting of chronic pelvic pain and is accompanied by symptoms of dysmenorrhea, dyspareunia, and urinary urgency. The etiologies set these two apart as PCS is due to pelvic venous insufficiency with one of many causes (dysfunctional valves, venous kinking from uterine malposition, structural and hormonal changes of parity, etc), whereas NCS is specific for congestion resulting from renal vein compression and retrograde flow through the uterine plexus. This distinction is important because the mainstay of treatment for PCS includes the embolization of

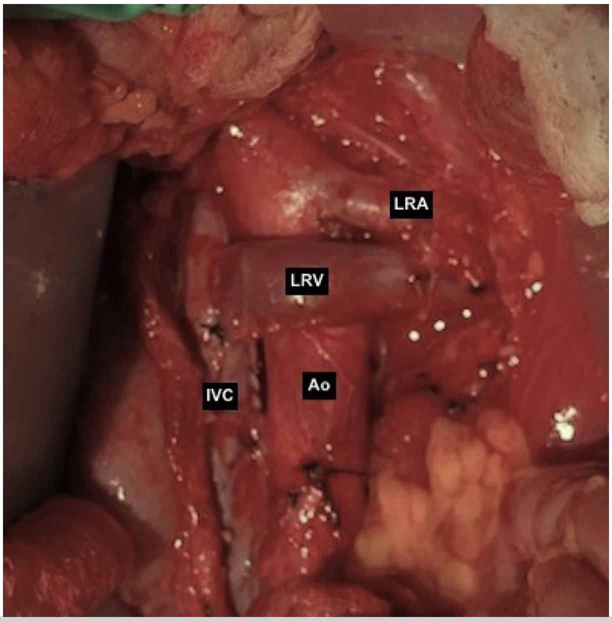

Fig 4. Anatomy after left renal vein (*LRV*) anterior transposition. *Ao,* Aorta; *LRA,* left renal artery; *IVC,* inferior vena cava.

incompetent veins, whereas the optimal treatment of NCS focuses on improving outflow of the LRV.

Multiple techniques have been described to treat pNCS, including conservative management,[8-10] LRV transposition,[11-14] left gonadal vein transposition,[15] nephrectomy with autotransplantation,[16] LRV venoplasty and stenting,[17,18] hybrid LRV transposition with stenting,[19] and extravascular stenting.[20]

Given our patient's severe symptoms and impacted quality of life, the decision to intervene was made. The first and most widely described technique is anterior LRV transposition. This procedure provides favorable patency rates with minimal need for reintervention. A large case series describing outcomes from LRV transposition from a heterogenous group of pNCS and aNCS had an 87% resolution of symptoms and 20% reintervention rate at 30 days for thrombosis and stenosis.[21] A true analysis of patients with pNCS is limited given they represented only 5% of the patients in the cohort. A second series described 30 patients with aNCS who underwent endovascular stenting. They reported symptomatic relief and no additional interventions at the 3-month follow-up.[22] Other series with longer follow-up document risk of stent migration into the IVC or heart in up to 6.7% of patients.[23,24] It has been suggested that a normal renal vein can dilate to more than 50% in size with the Valsalva maneuver.[25] Although the restricted LRV in NCS may not expand to this degree, this may be a helpful intraoperative study to evaluate for the potential for migration. Data specifically representing stenting for pNCS are limited, however, larger studies examining venous stenting in the iliac veins due to external compression does suggest that successful long-term patency rates can be achieved.[26]

Although stenting of larger veins is done with successful long-term patency rates, we were concerned about the compression between the aorta and vertebra and its impacts on a stent for the long-term future. For this reason, we chose an open repair. We have treated several patients with aNCS using an endovascular approach with excellent results at 5 years. However, we argue that this rare condition and unusual course of the vein lend themselves ideally to an open surgical repair.

CONCLUSIONS

After reviewing various open and endovascular options for intervention, we proceeded with an open approach for the most durable approach to pNCS. Although there are multiple described endovascular alternatives, long-term patency data for LRV compression between the lumbar spine and the aorta is limited. Our young, active patient has returned to her presurgical quality of life and activity level.

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