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Left Renal Vein Balloon Venoplasty without Stenting for Management of Symptomatic Nutcracker Syndrome

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Abstract

Nutcracker Syndrome (NCS) is a rare vascular anomaly involving left renal vein outflow entrapment most commonly secondary to compression between the aorta and the superior mesenteric artery. This can lead to chronic renal venous hypertension which can cause varicosity formation, gonadal vein reflux, hematuria, and if severe enough, renal failure. While open surgical reconstruction is heralded as the gold standard for treatment of this disorder, endovascular treatment is increasingly reported in recent literature. Herein, we present a unique approach to treatment of a severely symptomatic Nutcracker Syndrome via left renal vein outflow venoplasty without stenting performed in conjunction with embolization of refluxing varicosities.

Keywords: Left renal vein; Venoplasty; Stenting; Nutcracker syndrome

Introduction

Nutcracker Syndrome (NCS) or left renal vein entrapment syndrome is a rare congenital disorder involving compression of the left renal venous outflow. The most common scenario involves compression of the central-most segment of the left renal vein as it courses between the aorta and the Superior Mesenteric Artery (SMA). Compression can also take place with retroaortic (posterior nutcracker syndrome) and circumaortic left renal veins [1,2]. By far, the most commonly reported and the gold-standard treatment modality is open surgical reconstruction via left renal vein distal transposition, venolysis with patch venoplasty, or proximalization of the SMA [3]. While mid to long term outcomes using this approach remain satisfactory, the invasive nature of the operation and associated morbidity make it an undesirable first-line option. Recently, endovascular stenting of the central left renal vein has been reported more frequently with satisfactory short to mid-term results. However, there have been reports of stent fracture, migration, and thromboembolic complications that have questioned the durability and safety of stenting [4-7]. Here in, we present a case of a severely symptomatic Nutcracker Syndrome in a 23 year old female treated successfully via central left renal venoplasty with coil embolization of refluxing varicosities.

Background

A 23 year old female with a history of Irritable Bowel Syndrome (IBS) presented to our institution with a 3-dayhistoryof severe, throbbing, left flank pain. The pain was localized to the left flank and perinephric region, radiating to the left groin, and was associated with nausea. She reported similar episodes of severe flank and pelvic pain over the course of one year that was particularly worse during menstruation. She also had several episodes of menorrhagia. On presentation, she was a febrile, tachycardic (102 beats/min) with minimal costo-vertebral angle tenderness. The patient had evidence of microscopic hematuria with negative urine cultures and no evidence of pregnancy.

Computed Tomography Angiography (CTA) with delayed venography (CTV) revealed severe compression of the left renal vein outflow between the SMA and the aorta, a dilated left ovarian vein, and enlarged tortuous left-sided pelvic vessels. The angle between the SMA and the aorta on sagittal reconstructions measured approximately 20 degrees. A left renal vein duplex was obtained confirming significant central venous compression between the SMA and the aorta measuring approximately 4mm in diameter compared to a more peripheral maximum diameter of 19 mm (a ratio of approximately 1:5). Spectral wave form analysis at the point of compression revealed apulsatile venous signal with a velocity of 250 cm/second compared to caval velocities of approximately 60 cm/sec (a ratio of approximately 4:1). Given these findings, she was recommended to undergo

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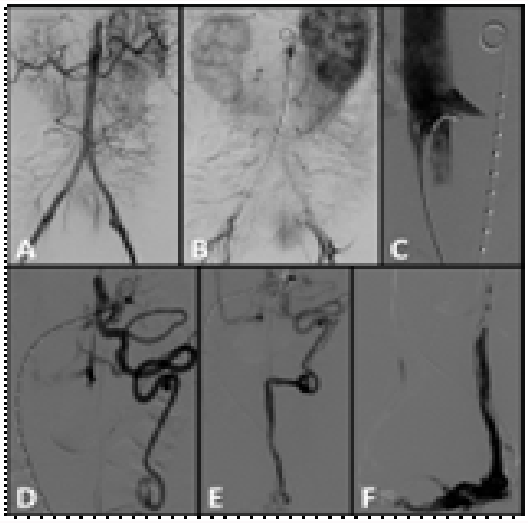

Figure 1A-B: Flush aortography demonstrates no evidence of arterial pathology but reveal evidence of left renal venous hypertension with sluggish washout of left nephrogram contrast. **C:** Selective left renal vein catheterization confirms central renal venous stenosis. **D-F:** The lesion is crossed and flush venography demonstrates significant renal vein varicosities with significant reflux into the left gonadal venous system.

diagnostic angiography as well as venography to better delineate the local angio architecture, rule out pelvic arteriovenous malformations, and assess extent of pelvic venous reflux. Endovascular therapy if deemed appropriate would be offered concomitantly.

Materials and Methods

The procedure was performed in the hybrid operating suite under general endotracheal anesthesia. Flush aortography via right common femoral artery puncture revealed no evidence of high-flow arteriovenous malformations, uterine leiomyomas, or congenital renal arterial anomalies. However, delayed phase venographic runs revealed delayed emptying of the left renal nephrogram relative to the normal right side suggesting a venous outflow compromise (Figure 1A and B).

At this point, the left common femoral vein was accessed and diagnostic iliocavogram revealed no abnormalities. Selective catheterization of the Left Renal Vein (LRV) revealed a high-grade stenosis at the LRV outflow close to its insertion into the Inferior Vena Cava (IVC) (Figure 1C). The area of stenosis was crossed using an angled glidewire and a 5-Fr Cobra catheter (Terumo, Tokyo, Japan) exchanged for a 5-Fr marked pigtail catheter positioned as far peripherally as possible into the LRV. Diagnostic venogram in multiple projections including anteroposterior, oblique, and sagittal windows revealed significant stagnation of LRV outflow due to central LRV stenosis with subsequent formation of large LRV refluxing varicosities extending into the left pelvic circulation with subsequent drainage into the accessory hemiazygous system. In addition, a large, refluxing left gonadal vein was also noted with extensive pooling of venous blood into the left para-uterine region collateralizing in this area with the right side (Figure 1D-F).A pressure gradient of 10mmHg was noted across the area of LRV stenosis.

We initially proceeded with balloon venoplasty of the LRV stenosis using a 10 x 40 mm Mustang balloon (Boston Scientific, Marlborough, MA) (Figure 2). Post venoplasty showed brisk, unobstructed flow through the previously stenotic area with significantly decreased filling of the LRV varicosities (Figure 3) and no evidence of recoil or

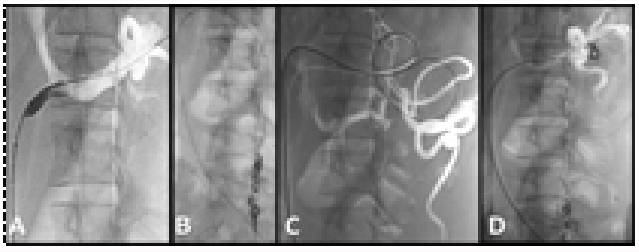

Figure 2A: Significant wasting noted during balloon venoplasty of the central left renal vein. **B:** Selective catheterization and sheath insertion with deposition of pushable coils into the left gonadal vein. **C-D:** Selective catheterization of the left renal vein varicosities with coil embolization.

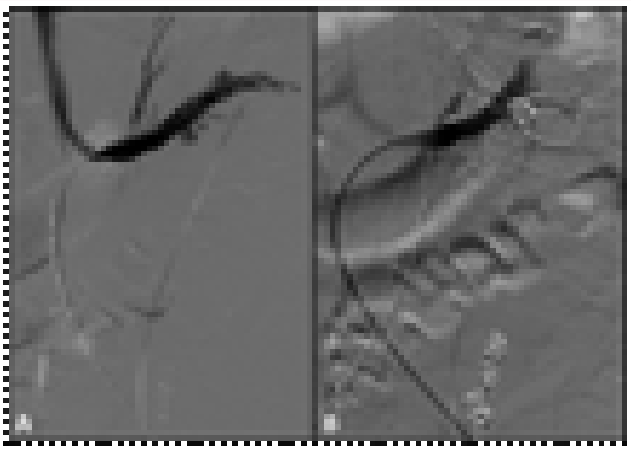

Figure 3A-B: Completion venography shows brisk washout of contrast through left renal view with no visualization of the varicosities and gonadal vein reflux even on delayed venous runs.

restenosis across the treated area. There was also noted to be improved flow through the adrenal and lumbar tributaries. No further pressure differential was noted across the venoplastied area.

Given the presence of incompetent left gonadal vein and LRV varicosities causing significant pelvic congestion syndrome, decision was made to proceed with coil embolization of these veins. These veins were selectively catheterized using a 5-French Berenstein catheter (Angio-dynamic Co, Queensboro, NY) with subsequent insertion of a6-French 55 cm Raabe Sheath (Cook Medical, Bloomington, IN). Left gonadal vein embolization was performed via3 densely packed pushable 0.035 platform coils (Interlock, Boston Scientific, Marlborough, MA) (Figure 2B). Similarly, the large dominant LRV varicosity was accessed and coil embolized with a single pushable coil (Figure 2C and D). Completion venogram showed elimination of pelvic reflux with maintained brisk, unobstructed flow through the LRV into the IVC (Figure 3). There were no perioperative complications. Patient was admitted for overnight observation. She reported elimination of left flank pain at the time of discharge. She was seen in follow-up at 2 weeks, 3 months, 6 months, with no recurrence of symptoms. Follow-up LRV duplex revealed a velocity gradient of less than 3 with a LRV outflow diameter ranging from 6-15 mm. In absence of recurrence of symptoms, she will be followed annually with LRV duplex surveillance. Consent to publish this manuscript was formally obtained from the patient.

Discussion

Venous entrapment syndromes such as May-Thurner Syndrome,

Thoracic Outlet Syndrome, and NCS are increasingly recognized and reported in the recent literature in large part due to enhanced endovascular diagnostic capabilities and growing awareness amongst treating specialists [8-10]. The Hamburg Classification Scheme categorizes these venous entrapment syndromes as congenital, obstructive venous anomalies and labels them as truncal vascular malformations given their central location and embryological patterns of development [11]. NCS involves hemodynamically significant compression of the LRV outflow near its insertion point into the IVC most commonly secondary o a hyperacute angle between the SMA and the aorta. NCS can also occur with retroaortic and circumaortic LRV configurations [1,2]. Symptoms, when present include most commonly left flank and lower abdominal pain, hematuria (both microscopic and gross), and pelvic congestion syndrome [10,12,13]. The latter develops secondary to the formation of incompetent varicosities and development of reflux within the left gonadal vein.

Diagnosis is mainly one of exclusion after the onset of symptoms which can be generalized and non-specific or potentially ignored and brushed off as psychogenic in origin as was the case in our patient who was given a diagnosis of IBS prior to vascular surgery evaluation and subsequent workup. Once more common diagnoses such as primary pelvic congestion syndrome, neoplasm, iatrogenesis, and medical renal pathology (especially autoimmune and post-infectious etiologies) have been ruled out, clinical suspicion for NCS rises most often due to the incidental finding of LRV pathology on imaging performed for work up of other etiologies. Once the diagnosis of NCS is entertained, LRV duplex in combination with Computed Tomography Venography (CTV) or Magnetic Resonance Venography (MRV) is strongly recommended for confirmation of the diagnosis and assessment of the hemodynamic significance of the lesion. What will determine the need for diagnostic venography is presence of several key distinctive features on dedicated LRV duplex ultrasound. These include LRV compression ratio of greater than 5, compared to the dilated peripheral portion, and compressed velocity ratio of greater than 5 compared to adjacent IVC velocities [3,10]. The third definitive criteria for diagnosis of NCS requires selective LRV catheterization with venography and measurement of pressure gradients across the area of compression. A pressure gradient of greater the3 is considered hemodynamically significant [3,14]. Venography can also facilitate use of intravascular ultrasound (IVUS) and multiplanar imaging for more accurate assessment of degree and hemodynamic significance of the stenosis, venous drainage patterns, and extent of pelvic reflux if present [15].

Open surgical reconstruction is the current gold-standard modality for intervention, with favorable mid to long term outcomes noted in several reports [14,16]. The most common open approach is an inferior transposition of the LRV [3]. Distal transposition, with end to side reanastomosis, has shown to provide good patency out comes in pediatric patients [17]. However, the morbidity associated with the invasive nature of these open procedures continues to make them a less desirable first-line option. Indeed, when presented with potential options for treatment, our patient refused all open interventions given associated morbidity. Endovascular intervention, on the other hand, is a newer, less invasive, fairly effective approach for treatment of symptomatic NCS. Historically and by and large, endovascular therapy has comprised of stent deployment across the area of compression often requiring extension into the IVC [2,4]. Stent use has seen favorable outcomes in several case reports and series, but significant concerns remain with their use in the young population owing mostly to reports of in-stent thrombosis and stent migration [4-7]. Furthermore, long-term outcomes remain unknown. The dynamic configuration of LRV wall circumference makes it suboptimal for stent placement. Proper apposition is often difficult, and therefore formation of gutters as well as stent migration with fracture and thromboembolic phenomenon are not uncommon [4-7,18]. In a large series by Chen et al. [7] perioperative stent complications requiring operative intervention and stent migration into the right atrium have occurred.

Venoplasty alone without stent placement has not been reported commonly in the literature. Indeed we encountered only a single case in the English language literature reporting the use of venoplasty for treatment of symptomatic NCS. Similar to the current case, Bekou and colleagues reported a case of NCS with severe pelvic congestion treated with LRV venoplasty and ovarian vein embolization [19]. Their patient was asymptomatic at 18-month follow-up with widely patent LRV, as confirmed by LRV duplex.

Not unlike symptomatic varicose veins of the lower extremities, pelvic varicosities associated with NCS develop as a result of venous hypertension caused by obstruction and subsequent reflux. Alleviation of both obstruction and reflux is necessary to achieve maximum therapeutic benefit. In this case, we have demonstrated that LRV venoplasty alone without routine stenting in conjunction with embolization of the refluxing gonadal vein and LRV varicosities can be highly effective in alleviating LRV hypertension and associated NCS symptoms. It forgoes the need for stent surveillance and maintenance and obviates the need for a major morbid operation. While larger experience and longer follow ups are needed before any stern recommendations can be made, it is safe to propose that this approach may serve as an acceptable first-line treatment modality for symptomatic NCS before stenting or open surgical revisions are entertained.

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