Management of Nutcracker Syndrome: Left Renal Vein Transposition

Shihan Feng, OMS-4[a], Robert Murrell, OMS-4[a], Michael Deiparine, MD,[b]
Kansas City University of Medicine and Biosciences[a], Midwest Aortic and Vascular Institute[b]
Kansas City, Missouri

Introduction

Left renal vein compression or entrapment with compromised outflow from the left renal vein (LRV) into the inferior vena cava (IVC) from extrinsic compression is often referred to as nutcracker syndrome. Anterior nutcracker syndrome with the entrapment of the left renal vein between the aorta and superior mesenteric artery (SMA) is the most common presentation. Less commonly a posterior nutcracker syndrome can be seen with a retro-aortic left renal vein being compressed between the aorta and vertebral body.[2]

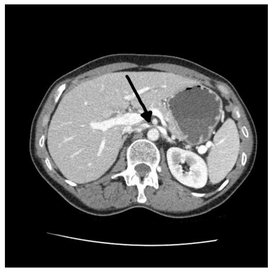
Data from published case reports show a wide patient age range with the majority of patients being middle aged women.[1][3] The exact cause of nutcracker syndrome is unknown. Different theories have suggested possible causes including an acute angle between the aorta and SMA, an abnormal or variant branching of the SMA or LRV, and entrapment by fibrolymphatic tissue.[1][3]

The obstruction of blood flow in the LRV can be asymptomatic and self-limiting, especially in the pediatric population. However if it becomes symptomatic, it commonly presents as hematuria and abdominal or left flank pain that radiates to the thigh and buttock.[1]

Treatment options range from surveillance and conservative management in less severe cases to various types of open or endovascular surgical procedures. The surgical technique chosen depends upon each individual's anatomy and how likely that procedure is to relieve the patient's symptoms.[2]

Case Report

A 53-year-old female initially presented to her primary care physician (PCP) with a one-month history of moderate to severe sharp left lower quadrant abdominal pain with radiation to the left flank and associated hematuria. There was no aggravating or alleviating factors. The patient also reported a 2-3-month history of pelvic pain and pressure. There were no other associated symptoms. The patient has a past medical history of hyperlipidemia, endometrial polyp, menorrhagia, lumbar radiculopathy affecting left lower extremity, and psoriasis. A CT scan of the abdomen and pelvis was ordered by her PCP, which showed LRV compression between the SMA and aorta with an adjacent dilated left lumbar collateral, left ovarian vein, and left pelvic vein (**Figure 1 &2**).



Figures 1 (left) and 2 (right)- Axial and sagittal CT sections showing compression of the LRV between the SMA and aorta.

The patient was then referred to urology for workup of her hematuria. A cystoscopy was performed and the workup for microscopic hematuria was negative. The patient was eventually referred to vascular surgery for further investigation and intervention. LRV entrapment syndrome was ultimately diagnosed. Treatment options were discussed with the patient, which included endovascular stenting versus transposition of the LRV. It was determined that the left renal vein was so tightly compressed that a transposition would be the better option for the patient. Stenting was considered, but there was a concern that the stent would not adequately open up the flow channel and would thrombose because of the severe extrinsic compression by the SMA. A mutual agreement was reached with the patient, and she was scheduled for an elective LRV transposition to adequately open up the left renal vein and alleviate the compression.

Case Report

During the surgery, a midline incision was made to enter the abdomen. Upon entering the abdomen, the ligament of Treitz was mobilized and the duodenum was retracted. The inferior vena cava (IVC) and aorta were identified. The LRV was subsequently exposed to where the large gonadal and lumbar veins were. The stump was oversewn on the left gonadal vein (LGV) and the left adrenal vein was ligated (**Figure 3**). Four-thousand units of Heparin were administered. The IVC was clamped with a side-biting clamp and division of the LRV off of the IVC was achieved. After oversewing where the LRV was removed from the IVC, the left renal vein was then moved distally along the IVC to a location where the renal vein could reach easily. Lateral venography on the IVC was done and the end-to-side anastomosis was flushed out prior completing the suture line between the LRV and IVC (**Figure 4**). Prior to closing the abdominal incision, good flow was visualized through the vein and into the IVC, and the vein sat within the abdominal cavity without undue tension. Retroperitoneum, abdominal fascia, and skin were closed with appropriate sutures. Standard steri-strips and dry sterile dressing were applied to the incision site.

The patient tolerated the procedure well. She experienced some intermittent nausea and vomiting postoperatively which resolved before discharge on postoperative day 3. At her two-week and four-week follow up appointments, she did not report any signs of hematuria, sharp left lower quadrant pain, or pelvic pain or pressure. The incision healed well without sign of infection, hematoma, or hernia. Follow-up renal arterial and venous ultrasound completed at six-months postop showed the LRV widely patent (**Figure 5**).

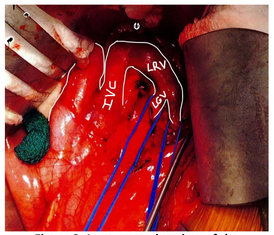


Figure 3- Intra-operative view of the oversewn left gonadal vein stump

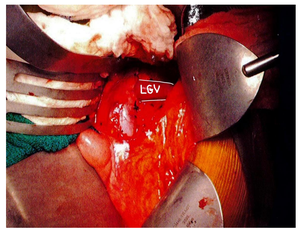


Figure 4- Intra-operative view of the end-to-side anastomosis

Figure 5- Six-month post-operative renal arterial and venous ultrasound

Discussion

The left renal vein can receive up to 12-15% of cardiac output. If outflow obstruction occurs, this can lead to distension and swelling of small collateral vessels. The renal venous hypertension can result in swelling of the left kidney, which can cause left renal colic, flank and abdominal pain, nausea, and vomiting. Other symptoms may include gross or microscopic hematuria, varicoceles in men due to an incompetent gonadal vein, and pelvic varices with pelvic congestion syndrome in women. The diagnosis is often missed or delayed due to the lack of standard diagnostic criteria and variable presentation of symptoms, leading to possible underestimation of the true prevalence of the condition.

The diagnostic work-up for NCS focuses on a step-wise approach. It is important initially to exclude other possible more common diagnoses. The first step begins with a history of presenting symptoms and thorough physical examination. Hematuria and abdominal/flank pain are the most common presentations. Therefore, full workup will usually initially include urinalysis, urine cultures, possibly cystoscopy and CT angiogram or magnetic resonance imaging (MRI) to evaluate for common renal causes of hematuria. Non-invasive doppler ultrasonography should be used as the first diagnostic assessment tool if NCS is suspected clinically.[1] Several studies have shown that anteroposterior diameter (AP) ratio between the distended and narrowed portion of the LRV and peak velocity (PV) ratio between the distended and narrowed portion of the LRV are good evaluating criteria for diagnosis of NCS. Although different studies used different cutoff ratios, the majority of the studies have a cutoff above 4 for both AP and PV ratios for indication of NCS .[1][3][5] CT angiography/MR angiography in the sagittal plane can be used to measure the aortomesenteric angle (AMA). The typical range for a normal AMA should be between 38° to 56°, and studies have shown it to be less than 16° in NCS patients. LRV compression and dilation can also be visualized through these imaging modalities.[1][5] The presence of an elevated renocaval pullback pressure gradient is diagnostic of NCS. Normally, there is minimal to no pressure gradient between the IVC and LRV. Presentation of increased LRV pressure, with a pressure gradient greater than 3 mmHg via venacavogram in situations where NCS is suspected, can confirm the diagnosis.[3][5][6]

Once NCS is diagnosed in a patient, the next step is to determine the method of treatment that is most suitable for the patient. The initial goal of intervention is to reduce hypertension in the LRV. Factors that are taken into consideration when determining the treatment methods include the severity and stage of the symptoms, as well as the age of the patient.[7] Conservative management is typically recommended for patients who are asymptomatic, have mild symptoms, or are younger than the age of 18. In the pediatric population, symptoms are often self-limited and spontaneously resolve due to normal physical development. Studies have shown that up to 75% of patients younger than 18 will have complete resolution of hematuria.[1] Many open surgical techniques have been used to treat Anterior NCS, including but not limited to LRV transposition, patch venoplasty without LRV transposition, gonadal vein transposition, saphenous vein bypass, and extravascular stenting. Open surgical technique for Posterior NCS includes anterior transposition of the left renal vein. Endovascular stenting is also another treatment option for NCS.[8]

Discussion

LRV transposition is the most commonly used approach currently and remains a safe and effective treatment of patients with NCS. It was first performed in the early 1980's. Studies have shown that LRV transposition has led to improved long-term outcomes and greater resolution of flank pain, hematuria, varicocele, and pelvic varicosities with low rates of morbidity as compared to conservative and other surgical managements. Above 80% of the patients with LRV transposition showed resolution of hematuria and flank pain.[5] However, studies have shown that one in three patients after open repair required reintervention, most frequently LRV stenting. Open reconstruction should be tailored to the patient's anatomy, and placement of vein cuff or patch may help reduce restenosis after open repair.[9][10][11] It is our practice to use ultrasound postoperatively to follow patients after left renal vein transposition to evaluate for possible development of LRV postoperative stenosis. Extravascular stenting with ringed polytetrafluoroethylene (PTFE) graft wrapped around the left renal vein has also been described. Patients will need to be on anticoagulation therapy like any other stenting procedures.[12]

Endovascular stenting is a less invasive procedural approach to treat NCS. The first endovascular stent placement for the treatment of NCS was described in 1996. Recent studies have shown that endovascular treatment is a safe, effective, and minimally invasive technique that provides long-term patency rates for patients with NCS. Stenting helps provide relief for the symptoms associated with NCS. Studies have shown that the renocaval pullback pressure gradient normalizes post-procedure for many patients. However, it has been reported that persistence of hematuria in patients despite the reduction of compression or entrapment and normalization of the pressure gradient does occur.[5] Further experience and data need to be collected and analyzed before accepting this procedure as the superior treatment for NCS over LRV transposition. Complications of stenting like any endovascular surgery include stent migration, restenosis, and formation of a clot.[13][14][15] Due to lack of standard protocol and guideline for stent type and sizing, specifically for NCS, it can further increase the chance of migration and the potential of re-stenting. To reduce the possibility of thrombosis and maintain stent patency, patients with endovascular approach will need to be placed on anticoagulation therapy.[5] Our own experience with endovascular treatment of nutcracker syndrome has had mixed results.

Nutcracker Syndrome is a rare condition that remains difficult to diagnose. Diagnostic and management options will be constantly changing and improving as more data becomes available. Treatment decisions should not solely made on radiographic evidence alone: The patients' age, anatomy, severity of symptoms, and the patient's wishes all need to be taken into consideration to determine the proper course of action . If surgical intervention is ultimately necessary, LRV transposition remains a relatively safe and effective method for long term symptom resolution. Further investigation is needed on comparison of long-term outcomes regarding different common methods of treatment, like endovascular stenting approach versus transposition.

Conclusion

Nutcracker syndrome is a rare and underdiagnosed condition due to lack of standard diagnostic protocols. Once diagnosed, patients experiencing severe symptoms can be successfully treated with left renal vein transposition with complete resolution of symptoms.