Management of Nutcracker Syndrome: Left Renal Vein Transposition

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Case Report

A 53-year-old female initially presented to her primary care physician (PCP) with a one-month history of moderate to severe left lower quadrant abdominal pain with radiation to the left flank and associated hematuria. There was no aggravating or alleviating factors. The patient reported no history of polycystic or hypertension. There were no other associated symptoms. The patient has a past medical history of hypertension, endometrial polyp, menorrhagia, lumbar radiculopathy affecting left lower extremity, and left gonadal vein stump associated with her left spermatic vein being compressed between the aorta and vertebral body.\(^1\)

Data from published case reports show a wide patient age range with the majority of patients being middle-aged women.\(^2\) The exact cause of nutcracker syndrome is unknown. Different theories have suggested possible causes including an acute angle between the aorta and the LRV, an abnormally branched or varying branch of the SMA, and entrapment by fibrolipomatosic tissue.\(^3\)

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.\(^3\)

Treatment options range from surveillance and conservative management in less severe cases to various types of open or endovascular surgical procedures. The medical treatment choice depends on each individual’s anatomy and how likely that procedure is to relieve the patient’s symptoms.\(^7\)

**Case Report**

The patient, which included endovascular stenting versus transposition of the LRV. It was determined that the left renal vein was too 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 thrombosis because of the severe extrinsic compression by the SMA. A mutual agreement was reached with the patient, and she was scheduled for an elective UV transposition to adequately open up the left renal vein and alleviate the compression.

The procedure was performed via a midline incision to enter the abdomen. Upon entering the peritoneal cavity, the left gonadal vein stump was noted (Figure 1). Thereafter, the extrinsic pressure was released, and the LRV was subsequently exposed. The LRV stump was oversewn on the left side and the LRV was transposed anterior to the SMA. The procedure is to relieve the patient’s symptoms.\(^1\)

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 venacaval junction and IVC were identified. The LRV was subsequently exposed to where the large gonadal vein (LGV) and the left adrenal vein was ligated (Figure 2). Four thousand units of heparin were administered. The incision was closed with a suture around the vein stump. Extraluminal pressure was measured in the IVC prior to completing the suture line between the LRV and IVC (Figure 4). Prior to closing the abdominal incision, good vasculature was visualized through the incision and into the IVC, and the vein sat within the abdominal cavity without undue entrapment or compression. The patient was discharged the next day and returned for six-week follow-up (Figure 5).

Further experience and data need to be collected and analyzed before guideline for stent type and sizing, specifically for NCS, it can further increase the chance of entrapment or thrombosis.\(^10\)

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 be 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.\(^13\)

If surgical intervention is ultimately necessary, LV 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.

**Discussion**

LV 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 LV transposition has led to improved symptom resolution and greater resolution of flank pain, hematuria, varicocele, and pelvic varicosities with low rates of complications.\(^2\) It has been shown to offer more long-term benefit over other surgical management techniques. Above 80% of the patients with LV transposition showed resolution of hematuria and flank pain.\(^1\) However, studies have shown that patients after open repair required reinvention, most frequently LV stenting. Open resection should be tailored to the patient’s anatomy, and placement of vein cuff or patch help may reduce hemostasis after open repair.\(^14\) It is our practice to use ultrasound postoperatively to follow patients after left renal transposition to evaluate for possible development of LV postoperative stenosis. Extravascular stenting with tapered polytetrafluoroethylene (PTFE) graft wrapped around the left renal vein has also been described. Patients will need to be on anticoagulopathy therapy like any other stenting procedures.\(^14\)

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 mean fenestration, a less invasive approach to open renal causes of hematuria, and needle aspiration with pelvic congestion syndrome in women. The diagnosis is often missed or delayed due to the lack of standard diagnostic criteria and varied 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 history of presenting symptoms and thorough physical examination. Hematuria and abdominal/pelvic pain are the most common presentations. Therefore, full workup will usually include imaging (renal ultrasound, intravenous pyelography, radionuclide renal scans, and CT angiography), cystoscopy or ureteropyeloscopy (UPJ), and contrast-enhanced CT/angiogram of the lumbar arteries. Non-invasive Doppler ultrasonography should be used as the first diagnostic assessment tool of NCS is suspected clinically.\(^2\) Several studies have shown that anteroposterior diameter (AP) ratio is the most important parameter for common renal cause of hematuria. Normal AP ratio is less than 4.2. Several studies have shown criteria like a normal AP should be less than 3.5, and studies have shown it to be less than 3 in NCS patients. LV transposition and dilation can also be visualized through these imaging modalities.\(^1\) The presence of an elevated renovascular pulse pressure gradient is diagnostic of NCS. Normally, there is minimal to no pressure gradient between the IVC and LRV. Measurement of increased LV pressure, with a pressure gradient greater than 5 mmHg on venousogram in situations where diagnosis can be confirmed.\(^2\)

Once NCS is diagnosed, the next step is to determine the method of treatment that is most suitable for the patient. The initial step 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.\(^3\) Conservative management is typically recommended for patients who are asymptomatic, have mild symptoms, or are younger than the age of 30 years. In the pediatric population, patients are often self-limited and spontaneously resolve in situations where blood pressure is controlled.\(^2\)

The left renal vein can receive up to 12% of cardiac output. If outflow obstruction occurs, this can lead to extension and swelling of small-caliber vessels. The renal venous hypertension can result in swelling of the left kidney, which can cause left renal vein clots, 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 varicosities with pelvic congestion syndrome in women. The diagnosis is often missed or delayed due to the lack of standard diagnostic criteria and varied presentation of symptoms, leading to possible underestimation of the true prevalence of the condition.\(^1\)

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**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.