Case Report

Nutcracker Syndrome- How It Was Presented and Managed? - A Rare Case Report

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Abstract
Anterior Nutcracker syndrome (NCS), also referred to as left renal vein entrapment syndrome, occurs due to extrinsic compression of the left renal vein between the abdominal aorta and superior mesenteric artery. NCS is a rare condition, and prevalence is equal among both genders. Clinically it may be asymptomatic, but the typical manifestations of this syndrome are hematuria, back pain, pelvic or flank pain on the left side, orthostatic proteinuria, and left- sided varicocele in male patients. A variety of imaging techniques like renal ultrasound, computed tomography renal angiography, magnetic resonance imagings (MRI) help to diagnose NCS. The treatment options, including conservative and surgical approaches, depend on the severity of symptoms.
Keywords: anterior Nutcracker syndrome, clinical features, diagnosis, treatment.

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Introduction

The name derives from the fact that, in the sagittal plane and/or transverse plane, the superior mesenteric artery (SMA), abdominal aorta (AA) and also spinal column appear to be a nutcracker crushing a nut (the renal vein). There are two forms of nutcracker syndrome (NCS) based on how the left renal vein (LRV) travels, anterior NCS being entrapment or compression of the LRV by the SMA and AA and posterior NCS being compression of the LRV by AA and spinal Column [1]. Anterior NCS is also known as LRV entrapment syndrome was first defined anatomically by Grant in 1937 as follows: " the left renal vein, as it lies between the aorta and superior mesenteric artery, resembles a nut between the jaws of a nutcracker'' [2]. The first clinical report of the nutcracker phenomenon was described by EI Sadr and Mina in 1950 [3].The precise prevalence of NCS globally is not known due to a lack of definitive diagnostic criteria [1]. Although NCS can occur at any age, the prevalence of NCS peaks in the second to fourth decades of life. NCS has been found to be higher in women; however, recent data suggest that it affects both sexes equally [4,5]. Anterior NCS is more common than posterior NCS. Anterior NCS is a classical variant that refers to extrinsic compression of the LRV between the AA and SMA, which impedes outflow of blood from the LRV into the inferior vena cava (IVC) [6], often accompanied by medial aorto-mesenteric narrowing and lateral hilar dilatation.

Other less common causes of anterior NCS include lordosis, reduced retroperitoneal and mesenteric fat tissue, pancreatic neoplasm and compression by adjacent lymphadenopathy [1,7].

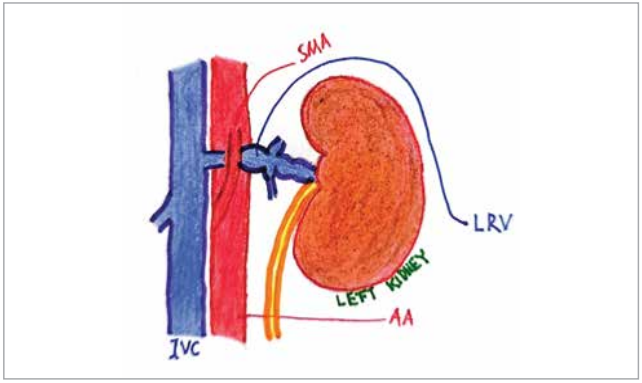


Figure 1: Anterior Nutcracker Syndrome

IVC: inferior vena cava; SMA: superior mesenteric artery; AA: abdominal aorta; LRV: left renal vein.

The syndrome cannot be regarded as a hereditary disease, despite it occurs in siblings [3]. Clinically it may be asymptomatic, but the typical manifestations of this syndrome are hematuria, back pain, pelvic pain, flank pain

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on the left side in women, orthostatic proteinuria, and left-sided varicocele in male patients.There is no diagnostic criteria for anterior NCS. The diagnosis isbased on(a) symptoms,primarily a history of hematuria and flank pain; (b) urine studies like urine analysis and urine phase contrast microscopy; and (c) imaging results such as Doppler ultrasound, Computed tomography (CT) renal angiography, and magnetic resonance imaging (MRI) [1,3,4,7]. The management of the NCS is dependent on the severity of symptoms and age of the patient. The management options include conservative and surgical approaches. Herein we present a case of anterior NCS, managed surgically.

Case presentation

A 25-year-old male presented to the nephrology outpatient department with ahistory of passage of painless red-colored urine associated with persistent bilateral flank pain for about 2 months. The flank pain was severe in nature for the last seven days, prior to the visit to the nephrology clinic. He had also complained of back pain for the same duration. He had no history of fever, weight loss, or bleeding tendency. The patient was normotensive, non-diabetic and had no history of burning micturition, urgency & frequency of micturition or recurrent urinary tract infections, surgery or history of trauma, no family history of renal disease, allergic disease, or drug abuse. With this history, some laboratory and radiological tests were requested by the nephrologist and diagnosed as NCS. Laboratory investigations showed: serum hemoglobin of11.7 g/dl, INR of 1.98, and serum creatinine of 1.1 mg/dl. Urine analysis revealedtrace albumin with 60-70 red blood cells per high power field (RBC/HPF), urine for dysmorphic cells of 2-3%, and a urine culture result of no growth. Other biochemical reports were shown within the normal range.

Ultrasonography of the kidney, ureter and bladder (KUB) region with post-void residual (PVR) showed the LRV appears significantly compressed between the AA and SMA, with the diameter being about 8.0 mm in the pre-compressed LRV and about 2.7 mm in the aorto-mesenteric segment of the vein, with the ratio of diameter being about 2.96. The SMA branching angle with the AA appears reduced to about 21.3 degrees, likely anterior NCS. Then he was referred to the cardiovascular surgery outpatient department for surgical correction. Thereafter, he got admitted to our cardiovascular surgery unit through the outpatient department as a diagnosed case of anterior NCS to undergo surgery.

After admission, on physical examination, he was well-looking. The patient had undergone cardiovascularand respiratory system examinations: pulse rate was 88 beats/min, blood pressure was 120/80 mmHg, respiratory rate was 22/min, and SpO2 was 97% on room air. Chest X-rays and echocardiography showed normal.The electrocardiogram showed sinus rhythm.

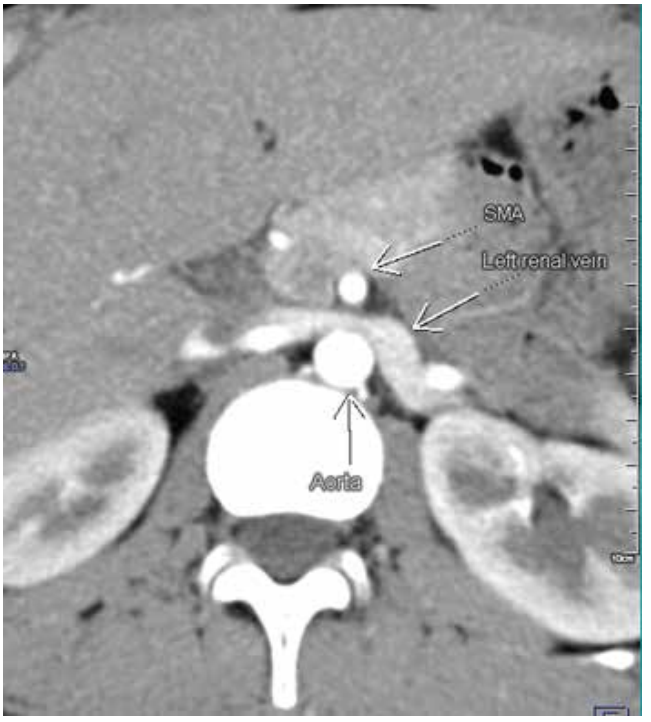


Figure 2A: ANS (Compression of left renal vein between AO and SMA).

ANS: anterior nutcracker syndrome; AO: aorta; SMA: superior mesenteric artery.

For further evaluation of the patient, a CT scan of KUB with intravenous contrast was done, which revealed compression of the LRV between the AA and SMA. Therefore, the diagnosis of anterior NCS was confirmed.

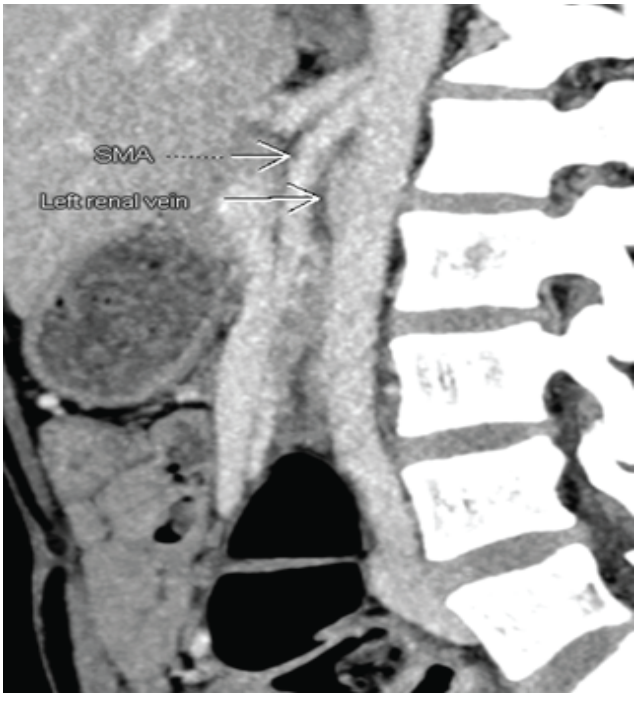


Figure 2B: ANS (Compression of left renal vein between AO and SMA).

ANS: anterior nutcracker syndrome; AO: aorta; SMA: superior mesenteric artery.

Then the patient was scheduled for surgical correction. Written informed consent was obtained from the patient and his parents. Surgery was performed with endotracheal intubation under general anesthesia (G/A). With all aseptic precautions and proper positioning, the abdomen was opened through a midline incision extending from the xiphisternum to the pubic symphysis. Upon retraction of intestinal loops, the aortic bifurcation, SMA, and inferior mesenteric artery were identified. The IVC, left renal artery, and LRV along with the right renal artery, were identified.

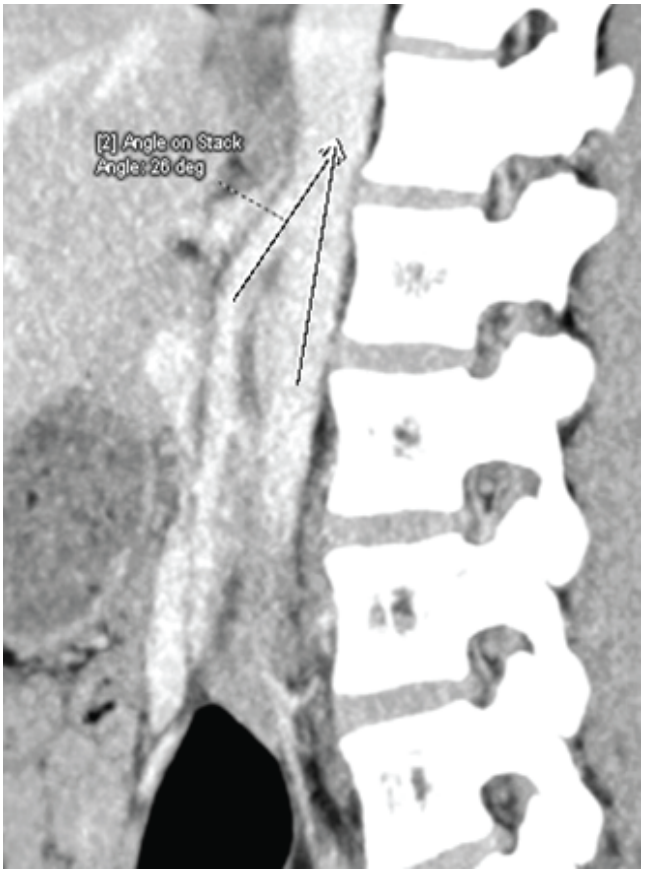


Figure 3: Decreased Aortomesenteric angle (AMA).

The LRV was found grossly dilated, entrapped within the aorta and SMA. Careful dissection was done to mobilize the LRV from surrounding structures. Heparin was given. A side biting clamp was applied at the IVC, and the LRV was transected along with the portion of the IVC. IVC repaired on side biting clamp with6-0 prolene in double layers. The LRV was then reimplanted with the distal portion of IVC, just distal to the SMA. The left kidney was then mobilized medially. Hemostasis was achieved. The wound was closed in layers after keeping one drain tube in situ. The patient was shifted to ICU with stable hemodynamic condition. He was extubated on the 'o' post-operative day, and the subsequent post-operative period was uneventful.

The patient was discharged from the hospital on the 8thday after surgery in good condition.

Follow-up

At the 3 and half -year follow-up, the patient was doing well and the CT Urography/ CT intravenous pyelogram revealed increased angle between aorta and SMA.



Figure 4A: Compression of LRV released at first follow up period. LRV: left renal vein



Figure 4B: Compression of LRV released (One month after operation)

LRV: left renal vein



Figure 5: Increased Aortomesenteric angle: Postoperative computed tomography scan

Discussion

NCS is a form of venous compression syndrome that affects the LRV, referred to as left renal vein entrapment syndrome. This type of syndrome is a rare condition.The terms nutcracker phenomenon and syndrome are used interchangeably.Aorto-mesenteric constriction of the LRV that is asymptomatic is referred to as "Nutcracker phenomenon," while specific symptoms associated with Nutcracker anatomical morphology are identified as "Nutcracker syndrome"[1]. The first clinical report of the nutcracker phenomenon was described by EI Sadr and Mina in 1950 [3], and later, in 1972, a Belgian radiologist, De Schepper, named the condition NCS [8]. NCS can occur at any age, ranging from childhood to eighthdecade of life but the prevalence peaks in the second to fourth decades of life [5]. It is assumed that the elongation of the renal vein and the reduction of the angle between the SMA and AA may be associated with nephroptosis above or decreased retroperitoneal adipose tissue.

It has also been hypothesized that the rapid growth of the vertebral bodies and height that happens during adolescence causes the aorto-mesenteric angle (AMA) to decrease [9].
NCS has two variants: anterior nutcracker syndrome, which is a classical variant, and posterior NCS, which is retroaortic or circumaortic, a rare variant. Other variants include right-sided nutcracker syndrome, LRV duplication, LRV compression by a dilated left inferior vena cava, and LRV compression between the SMA and the right renal artery [10]. Hematuria is the main symptom of this syndrome, occurring in about 80% of the patients due to rupture of the thin-walled septum separating smaller veins fromthe renal collecting system, which is disrupted by high blood pressure in collaterals and subsequent venous sinuses close to the renal calyces [1,10]. Other symptoms include back pain, pelvic pain, flank pain on the left side in women, orthostatic proteinuria, and left sided varicocele (9.5%) in male patients.

NCS is confused with other diseases like nephrolithiasis and bladder cancer because of common symptoms. So, after careful history taking, physical examination and imaging results, include doppler ultrasonography, CT scan, MRI can help to diagnose NCS.

Diagnostic criteria are based on CT imaging results and Doppler ultrasoundstudy. CT imaging includes (1) AMA (aortomesenteric angle) of <35-39˚is significant for definitive diagnosis of NCS; thenormal value is 45-95˚. The abrupt narrowing of the LRV between the AA and SMA, the characteristic “beak sign” in the axial plane, is reported to have an 88.9% specificity and 91.7% sensitivity [1], (2) An LRV diameter ratio (Hilar-Aortomesenteric ratio) of ≥ 4.9 (66.7% sensitivity and 100% specificity) is recommended for the highest diagnostic accuracy [11]. In Doppler ultrasonography, reno-caval pressure gradients ≥3 mmHg indicates venous hypertension, consideredfor diagnosis of NCS [12]. MRI has the benefit of no radiation and multiplanar imaging, which gives better delineation of soft tissue anatomy in the region of the compression and other abnormalities, such as dorsolateral torsion of the left kidney, abnormally high course and compression or pre-stenotic dilatation of the LRV, abnormal configuration of SMA, and peri-renal or gonadal vein varices. T2-TRUFI (True Fast Imaging with Steady-State Free Precession) provided the best imaging quality [13].

The management of this syndrome is still debatable. There are currently no clear treatment guidelines due to limited data. Thus, it should be contingent upon the individual case, age of the patient, status of each patient, and severity of symptom [1,7].

The treatment options include conservative and surgical approaches. Conservative treatment is considered with mild hematuria or with mild and tolerable symptoms and in patients aged <18years. In growing individuals, 30% of patients with NCS exhibit a resolution of their symptoms in 24 months due to an increase in mesenteric fat stores and fibrous tissue near the SMA origin, which releases the entrapped LRV.ACE inhibitors, specifically “Alacepril” and low dose aspirin are used to improve orthostatic proteinuria and renal perfusion [5,7]. Surgical approaches may be considered for patients with recurrent gross

hematuria; anemia, severe flank or abdominal pain,impaired renal function, including persistent orthostatic proteinuria, and varicocele formation, and also considered for patients with persistent symptoms where a conservative approach is ineffective after 24 months in patients < 18 years of age and after 6 months in adults [14,15].

The open surgical methods for managing NCS that have been published include LRV transposition, patch venoplasty without LRV transposition, nephropexy, gonadal vein transposition, and renal auto transplantation [1], but the most common intervention is LRV transposition.The first documented case of surgical treatment for NCS was performed by Pastershank in 1974 on a 34-year-old man who had hematuria and flank pain. He did this by releasing the fibrous entrapment that had compressed the LRV between the aorta and SMA [16]. LRV transposition andrenal auto transplantation are two recent open vascular techniques. LRV transposition is carried out through a midline transperitoneal approach, consisting of excision of the veins at the IVC junction with reimplantation distal to the SMA,but it has some risks of postoperative complications like bleeding, small bowel adhesions, paralytic ileus, and vessel thrombosis [17]. Renal auto transplantation required much more dissection and is probably associated with a higher chance of complications. Endovascular, a minimally invasive approach is also another option for the treatment of NCS but have risk of stent migration into the right atrium, IVC and hilar LRV [18].

Endovascular treatment, which is a minimally invasive approach, is another option for treating nutcracker syndrome in recent years; however, it carries the risk of stent migration into the right atrium, IVC, and hilar LRV, stent thrombosis, and has limited data on its efficacy. So, LRV transposition is currently the most often used surgical procedure for NCS and is considered as the gold standard in management.

Our patient was 25 years old and had complaints of painless hematuria with bilateral flank pain. Ultrasonography of the KUB region with PVR showed the left renal vein appears significantly compressed between the abdominal aorta and superior mesenteric artery (SMA) and an AMA (aortomesenteric angle) of 21.3˚,was diagnosed as anterior NCS treated surgically.

Anterior NCS represents a critical yet frequently underdiagnosed entity. Its variable clinical presentation mandates a high index of suspicion, often requiring a multi-modality diagnostic approach that moves beyond simple imaging. The integration of Doppler ultrasound with cross sectional studies of CT or MR angiography provides the essential anatomical context, while invasive venography remains the definitive standard for assessing hemodynamic significance via renocaval pressure gradients. Treatment mut be tailored to the individual patient, balancing conservative observation for pediatric or asymptomatic cases. While surgical transposition of the LRV offers the most durable, gold-standard solution. Less invasive endovascular stenting provides a valuable alternative, although long-term patency data is still maturing.
Ultimately, advancing awareness and standardizing diagnostic thresholds are paramount to improving the identification of NCS, allowing for timely, personalized intervention and better functional outcomes for patient struggling with this challenging vascular compression disorder.

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Conflict of interest
The authors declare that they have no conflict of interest for publication.

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