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Left Renal Vein Compression Syndrome: Cracking the Nut of Clinical Dilemmas – Three Cases and Review of Literature

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Case series
Patient: Female, 36 • Female, 41 • Female, 36
Final Diagnosis: Left renal vein compression syndrome
Symptoms: Hematuria
Medication: —
Clinical Procedure: Percutaneous intervention • nephrectomy
Specialty: Nephrology

Objective: Unusual clinical course

Background: The term nutcracker phenomenon (NCP) elucidates anatomical structure and hemodynamics, whereas nutcracker syndrome (NCS) refers to clinical manifestations. We present three cases of similar clinical features of hematuria and flank pain with different clinical outcomes.

Care Report: Case 1: A 36-year-old Caucasian female with a past medical history (PMH) of HIV infection presented for evaluation of hematuria. Computed tomography (CT) without contrast showed pelvic venous congestion and narrowing of the extra-renal left renal vein (LRV). After the failure of conservative management, renal auto-transplantation was attempted but failed because of extensive venous collateral; the patient subsequently required a total hysterectomy due to recurrence of symptoms. Case 2: A 41-year-old Caucasian female with extensive PMH presented with chronic abdominal pain. A CT scan of the abdomen and pelvis showed pelvic venous congestion. The patient underwent angioplasty and stent placement of the LRV. Subsequently, a left ovarian vein embolization was performed. On follow-up visits, her symptoms improved. Case 3: A 36-year-old female with PMH of HIV infection, gastroesophageal reflux disease, and hypertension presented with hematuria and flank pain. Her venogram revealed 1 mm Hg pressure gradient across stenosis, suggestive of LRV hypertension. Over the months of her follow-up after discharge, her hematuria gradually decreased from daily to intermittent non-daily frequency, without any intervention.

Conclusions: The treatment of NCS includes observation, percutaneous angioplasty, open or endovascular surgery, or nephrectomy. In patients younger than 18 years of age, the best option is a conservative approach with observation for at least two years, as approximately 75% of patients have complete resolution of hematuria.

MeSH Keywords: Hematuria • Pelvic Pain • Renal Nutcracker Syndrome • Renal Veins

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/905324

Background

Nutcracker phenomenon (NCP) and nutcracker syndrome (NCS) are two terms that have been used interchangeably in the literature [1]. NCP is more commonly used to explain anatomical structure and hemodynamics, whereas NCS is used to refer to clinical manifestations [2]. Anatomically, the normal angle between the aorta and superior mesenteric artery (SMA) is close to 90° [3]. In NCS, there is compression of the left renal vein (LRV) in the angle formed by the aorta and the SMA, with the angle reduced to less than 35°. This phenomenon results in venous hypertension and congestion in the LRV and its tributaries and the left gonadal vein. Clinically, gross hematuria may be the only symptom; however, other common symptoms are left flank pain, proteinuria, and pelvic congestion in females and varicocele in males [4]. NCS can be diagnosed anytime from childhood to old age; and although the exact prevalence remains unknown, it is more frequently seen in young adults, and may be more prevalent in females [5]. Although, one study found no difference in prevalence between males and females [6]. Treatment of NCS mainly aims at decreasing LRV hypertension [3]. In this study, we present three distinct cases of similar clinical features of unexplained gross hematuria and flank pain with different clinical and management outcomes.

Case Report

Case 1

A 36-year-old Caucasian female with a past medical history (PMH) of human immunodeficiency virus (HIV) infection, who had been treated with anti-retroviral therapy (ARRT) for three years, presented to the outpatient office for the evaluation of blood in her urine for three months. She had a similar episode seven years earlier which resolved without any intervention. In addition to ARRT, she was also taking celecoxib for pain with previous use of ibuprofen as well. It was recommended that the patient stop celecoxib therapy. She had undergone cystoscopy and computed tomography (CT) urogram, which were both negative for renal calculi. Urinalysis revealed red blood cells (RBC) without any dysmorphic cells. Her infectious and rheumatologic serologic workup was negative for autoimmune diseases and hepatitis B and C viral infection. Her gynecological workup was also non-contributory. Her CT scan of the abdomen and pelvis was repeated along with a renal biopsy and LRV pressure measurement. The biopsy of the left kidney was unremarkable for any glomerular or interstitial pathology and negative for stains under microscopy, immunofluorescence, and electron microscopy. Her CT without contrast showed pelvic venous congestion syndrome, enlarged collateralized left ovarian

Figure 1. Enlarged and edematous left kidney (red arrows), dilated left renal vein (blue arrow), and dilated ovarian vein (yellow arrow).

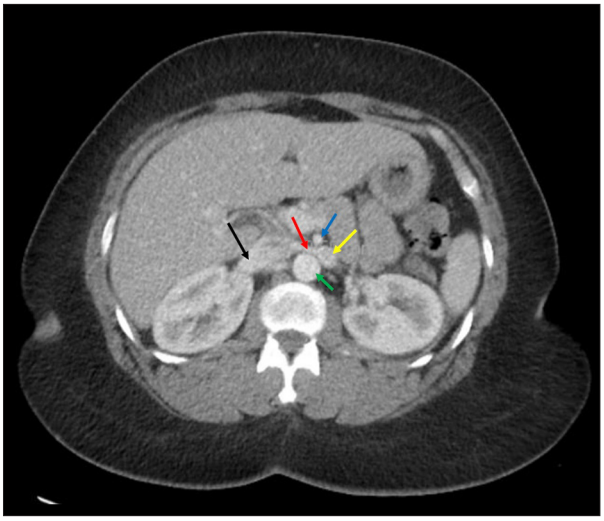

Figure 2. Normal right renal vein (black arrow) compared to compressed left renal vein (red arrow) between the aorta (green arrow) and the superior mesenteric artery (blue arrow). Yellow arrow shows pre compression dilated left renal vein.

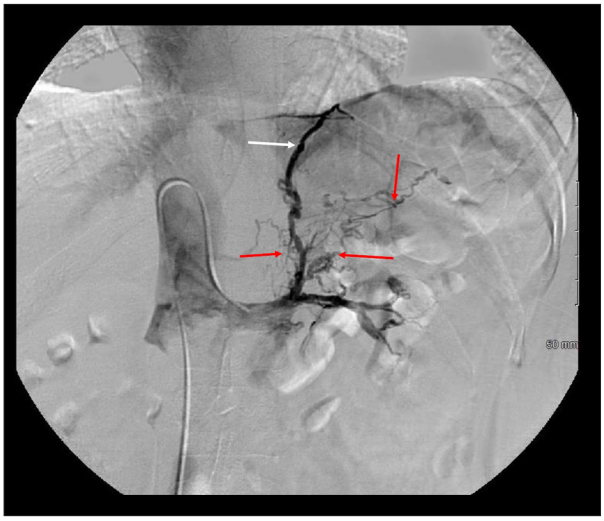

Figure 3. Arrows showing formation of multiple collaterals (red arrows) and the dilated left adrenal vein (white arrow).

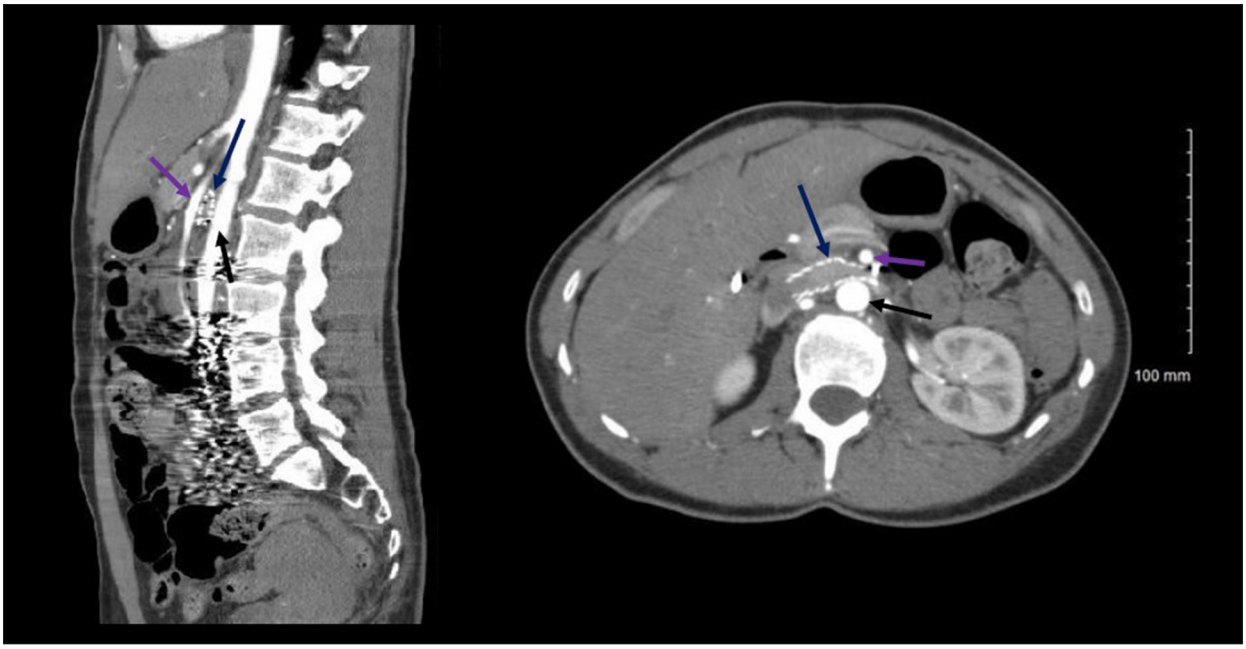

Figure 4. The stent within the compressed left renal vein (blue arrow), between the superior mesenteric artery (purple arrow) and the aorta (black arrow).

vein, and lower pole LRV (Figure 1). The extra-renal LRV was markedly narrowed, measuring 1.6 mm compared to 13 mm on the right (Figure 2). It was compressed between the SMA and the aorta and the take-off of the left renal artery. A renal auto-transplantation was attempted but failed because of extensive venous collateral (Figure 3); the patient later required a total hysterectomy due to the recurrence of symptoms. Her renal function remained stable after four years with a solitary kidney.

Case 2

A 41-year-old Caucasian female with PMH of cholecystectomy, tubal ligation, interstitial cystitis, median arcuate ligament syndrome status post dissection, and ligation with celiac artery compression presented with chronic abdominal pain. A CT scan of the abdomen and pelvis showed pelvic venous congestion. An angiography was performed which revealed

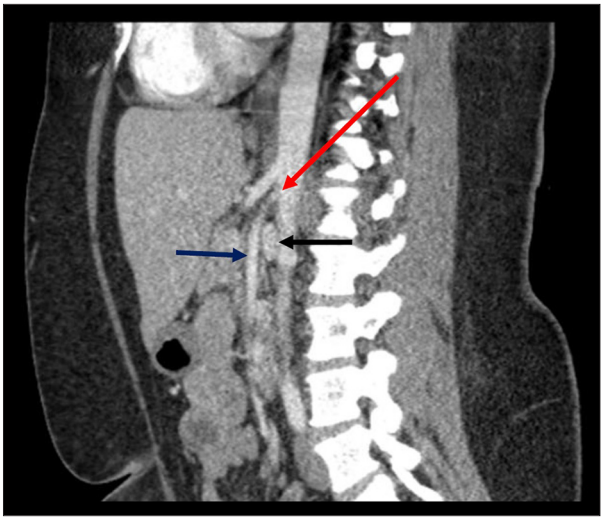

Figure 5. The acute angle of superior mesenteric artery with aorta (red arrow) compressed left renal vein (black arrow) and superior mesenteric artery (blue arrow).

stenosis of the LRV. Pre-stenosis pressure in the LRV was 18 mm Hg and post-stenosis pressure was 9 mm Hg with a gradient of 9 mm Hg (normal <1 mm Hg) suggesting LRV hypertension. The patient was managed with angioplasty of the left renal vein with the deployment of a 14 mm×4 cm nitinol stent (Figure 4). Subsequently, a left ovarian vein embolization was performed. On subsequent visits, the patient reported improvement in her symptoms.

Case 3

A 36-year-old female with PMH of HIV infection, gastroesophageal reflux disease, and hypertension presented with hematuria and persistent non-radiating flank pain of five months. Her outpatient serological workup for potential infectious and rheumatologic causes for autoimmune etiologies and conditions were negative. She was admitted to the hospital to undergo CT-guided biopsy of her left kidney, which was also unremarkable. Her CT of the abdomen with contrast disclosed a narrowed angle between the SMA and the aorta of 32° (normal=90°) (Figure 5) with distance measuring 6 mm (normal >8 mm) and the IR venogram revealed a pressure gradient across the area of compression of 1 mm Hg (normal <1 mm Hg) suggestive of LRV hypertension. Symptoms of hematuria gradually decreased over the months following her discharge, from daily to intermittent non-daily frequency, without any intervention.

Table 1 showing the baseline characteristics of patients, clinical presentation, imaging results and the intervention performed.

Discussion

NCS is more often considered an important differential diagnosis of gross hematuria now than it was in earlier years, yet most patients remain undiagnosed. The precise prevalence of NCS remains unknown, but it is believed to be higher in females [7]. It can be seen from the first decade of life until the seventh decade of life, but it is usually most symptomatic during the second and third decade of life [8]. The first case of LRV compression was reported in 1950 by El-Sadr and Mina [9]. De Schepper was the first physician to use the term "nutcracker syndrome" in 1972 [10], based on the observation that the LRV gets compressed between the aorta and the SMA in a "nutcracker" like fashion, where one can consider the aorta and the SMA as the two levers of the nutcracker. Extrinsic compression of LRV can also occur from pancreatic neoplasm, para-aortic lymphadenopathy, retroperitoneal tumor, and abdominal aortic aneurysm [11,12]. Less often, LRV takes a retro-aortic route and is compressed between the aorta and vertebral body [13,14], which also known as posterior NCS, in comparison to conventional NCS which is also known as anterior NCS. The anatomy of LRV and the adjacent structures and their variations, along with the hemodynamic study of LRV, are critical in diagnosing NCS. In addition, sometimes the right renal vein can be compressed during pregnancy causing right side NCS [15].

The SMA-aorta angle can vary even within the same individual with a change in different body parameters, such as body mass index (BMI), due to mesenteric and retroperitoneal fat content surrounding the angle, the curvature of vertebral column as lordosis pushes the aorta making the angle narrow, or ptosis of the left kidney. The average normal diameter of a LRV is 4 to 5 mm [16]. The presentation of NCS can vary from asymptomatic hematuria to severe pelvic congestion [17]. Hematuria usually occurs in the unilateral kidney due to the rupture of varices secondary to venous congestion and hypertension in the collecting system [18]; and during cystoscopy, the left ureteric orifice is usually seen to contain blood [19].

The treatment options for NCS vary and may range from close observation to percutaneous angioplasty, or open or endovascular surgery to nephrectomy. When deciding on the treatment plan, and before offering treatment, careful patient selection is important. It is imperative to differentiate between NCP and NCS to avoid overly aggressive treatment. It is also extremely vital to assess the extent of the condition and the development of venous collaterals; the development of collaterals is associated with high risk of treatment failure, especially when angioplasty is desired. In patients with advanced NCS, renal auto-transplantation should be the last resort and be performed in only highly specialized centers because venous collaterals make venous anastomosis difficult to perform.

Table 1. Baseline characteristics of patients, clinical presentation, imaging results and the intervention performed.

	Patient 1	Patient 2	Patient 3
Demographics			
Age (yrs)	36	41	36
Gender	Female	Female	Female
Ethnicity	Caucasian	Caucasian	African American
Clinical features			
Hematuria	Present	Unknown	Present
Pelvic pain and left renal colic	Present	Present	Present
Diagnostic tests			
CT evidence of LRV compression	Present	Present	Present
Doppler criteria	Present	Present	Absent
Management			
Initial recommendation	Conservative	Angioplasty with stent	Conservative
Final treatment	Failed renal auto transplantation Eventually TAH for symptom resolution	Angioplasty with stent	Conservative
Outcome	Pelvic congestion symptoms persist	Improvement of symptoms	Complete resolution

CT – computed tomography; LRV – left renal vein; TAH – total abdominal hysterectomy.

In young patients (younger than 18 years of age), the best option is a conservative approach [4] with observation for at least two years, as approximately 75% of patients will have complete resolution of their hematuria [4,20–23]. As seen in our case studies, one patient underwent drastic measures for her symptoms without resolution and eventually underwent hysterectomy, another patient had improvement in symptoms after the deployment of a stent, while the third patient did not require any intervention and was managed conservatively with complete resolution of symptoms within a few months.

Conclusions

Our case series can remind clinicians to consider NCS in patients who have unexplained hematuria and pelvic congestion syndrome. The treatment varies from conservative management, to minimally invasive procedure, to nephrectomy. These patients should be managed conservatively for at least 24 months because of the high rate of spontaneous resolution.

Conflict of interests

None.

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