

Case Report; Concomitant Left Nutcracker Syndrome and Right Ureteropelvic Junction Obstruction

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Abstract: We report rare case of concomitant left nutcracker syndrome and right ureteropelvic junction obstruction (UPJO) on adult female patient diagnosed by CT urogram after she presented with intermittent bilateral flank pain. For this we did Anderson-Hynes pyeloplasty for right ureteropelvic obstruction, it was laparoscopic initially but due to difficulty of stenting it is changed to open. She had smooth post-op course then discharged on 3rd post-op day. Symptoms of nutcracker syndrome are not that much bothersome for the patient so we planned to follow her conservatively.

Keywords: nutcracker syndrome, ureteropelvic junction obstruction, pyeloplasty, UPJO

Introduction

When the left renal vein (LRV) is compressed by the superior mesenteric artery (SMA) and abdominal aorta (AO), renal venous hypertension and accompanying symptoms ensue. This condition is known as left nutcracker syndrome. When urine flow from the renal pelvis into the ureter is functionally or anatomically obstructed at their anatomical connection, this condition is known as ureteropelvic junction obstruction (UPJO).¹ The presentation of concomitant left nutcracker syndrome and right UPJO is very rare. To our knowledge, there is only one similar published report from china in 2013 on female patient managed by LRV decompression and right Anderson-Hynes pyeloplasty.

Case Report

A 50-year-old female patient visit our outpatient clinic with complaint of mild intermittent left flank pain of 10 years and 1 year history of right flank pain otherwise she denies hematuria, decrease urine output, fever and trauma. No history of previous surgery. She newly diagnosed for hypertension and on amlodipine 5mg po daily, no other chronic medical illness. For this she was investigated with CT urogram (Figures 1–4) which showed, right ureteropelvic obstruction with moderate hydronephrosis, 3×3 cm Bosniak I left renal cortical cyst and the LRV is compressed between SMA and aorta with reduced aortic-SMA angle about 18° (normal angle is greater than 45°) with LRV compression ratio (CR) of 4. Also left gonadal vein is enlarged measuring 9 mm and multiple bilateral tortuous veins which are encompassing the uterus are noted with maximum diameter of around 6 mm. No associated lower urinary tract anomalies seen. She also investigated with complete blood count and urine analysis all normal including renal function test, creatinine is 0.56 mg/dl. Nuclear study is not available in our country. The patient explored laparoscopically and the renal pelvis was dilated with abrupt tapering at ureteropelvic junction area otherwise no crossing vessel identified, for this laparoscopic Anderson-Hynes (dismembered) pyeloplasty tried but after suturing posterior layer stenting was difficult then procedure converted to open. Since symptoms of nutcracker syndromes are not bothersome for the patient pre-operative plan was to follow her

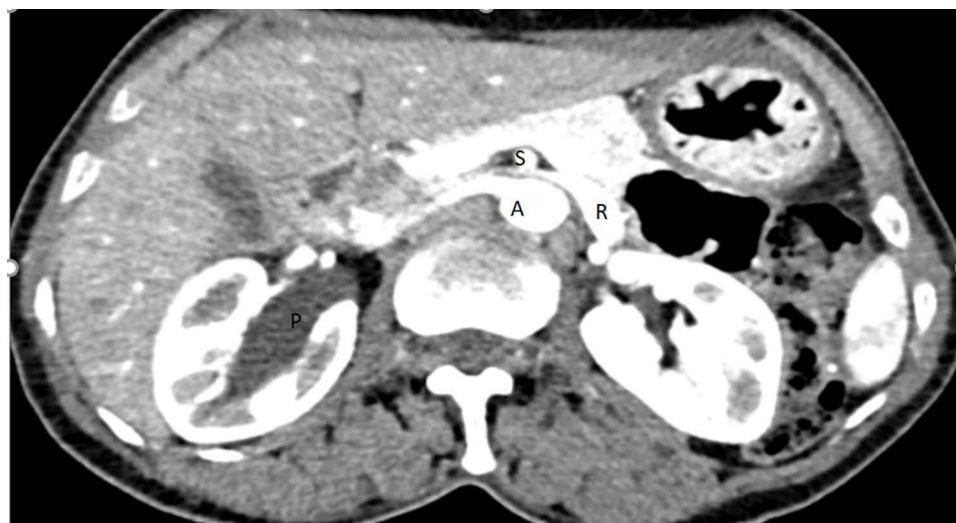


Figure 1 Cross-sectional image of patient showed left renal vein (R) compressed between SMA (S) and aorta (A) with dilatation of left renal vein. Dilated right renal pelvis (p) can also be seen.



Figure 2 Cross-sectional image shows dilated left gonadal vein (g) going along side the aorta (a) and dilated right renal pelvis (p).

conservatively. She had smooth post-op course and discharged home on 3rd post-operation day. On 3rd post-operation month pain on right side subsides and conventional abdominopelvic ultrasound shows normal kidneys without dilatation of the system. There was no worsening of pain on left side.

Discussion

The phrase “nutcracker syndrome” was first coined by De Schepper in 1972 after he observed two patients with recurrent hematuria brought on by compression of the LRV between the aorta and SMA. This condition was initially documented by el Sadr and Mina in 1950. The term for this condition is anterior or traditional nutcracker syndrome. Later, a variation known as “posterior” nutcracker syndrome described compression of a LRV that runs between the spine and the posterior wall of the aorta. A patient with duplication of the LRV and compression of either the anterior tributary between the aorta and SMA or the posterior tributary between the aorta and spine was also reported to have dual (anterior and posterior) nutcracker syndrome.²



Figure 3 (A) coronal images showed dilated left gonadal vein (G). (B) right UPJO on delayed film with no passage of contrast to right ureter (P).

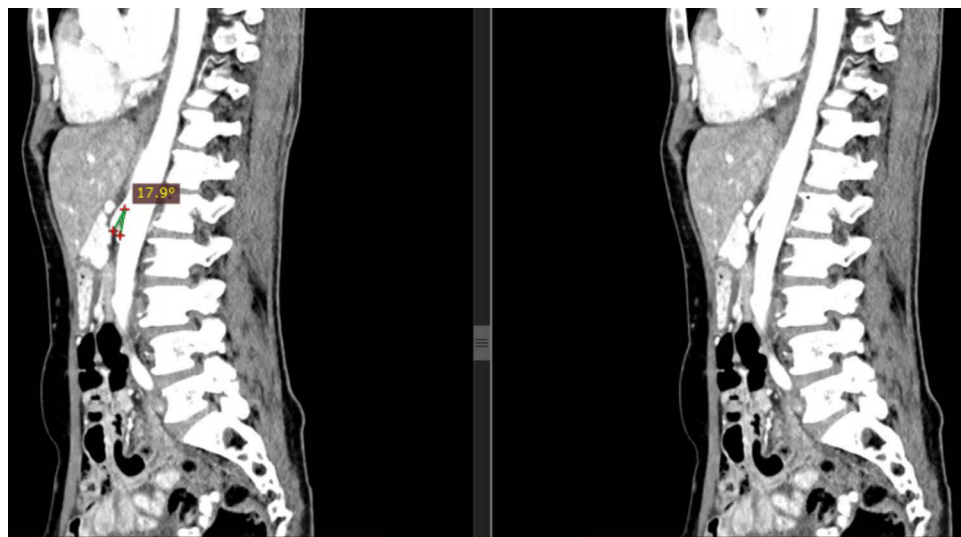


Figure 4 Sagittal view shows narrow angle between aorta and SMA, around 18° also slit of renal vein seen while passing between SMA and aorta.

A disorder known as pelvic-ureteric junction obstruction (PUJO) causes disruption of urine flow between the proximal ureter and kidney pelvis. A male to female ratio of 2:1 and a live birth rate of 1:500 are used to identify PUJO. The most frequent cause of congenital hydronephrosis is thought to be PUJO. However, it may also appear later in adulthood and evolve (acquired).³ Pelviureteric junction obstruction (PUJO) is a collection of obstructive processes that develop as a result of several circumstances, which can be roughly categorized as intrinsic or extrinsic. PUJO is not a singular physical defect. Internal or ureteric causes arise from stenotic or aperistaltic segments, however intraluminal lesions or the presence of ureteric valves may also occur less commonly. An accessory crossing vascular, typically an accessory lower-pole renal artery, is the most frequent extrinsic cause and results in external compression of the PUJ. There is still debate over how frequently crossing vessels are the underlying source of blockage in individuals presenting with PUJO, despite the fact that they are a common finding in the unobstructed population.⁴

Hematuria is the most common symptom of NCS, along with or without left flank pain. Young people make up the bulk of the affected patients. In young guys, varicocele is a common finding. Older white men see a second rise in the incidence of NCS. These individuals typically exhibit pelvic venous congestion symptoms as a result of severe pelvic varices and gonadal vein reflux. Ninety percent of the time, the left side is affected, and in 4% of cases, bilateral varices have been documented. These patients frequently have microscopic hematuria, and only one-third of them report left flank pain.⁵

On CT images obtained on arterial or portal venous phases, the preaortic or retroaortic course of the LRV and the compression between the AO and the SMA can be clearly seen. The “beak sign”, or sudden narrowing of the LRV with an acute angle below the aortomesenteric junction, is typically visible on axial CT imaging of the LRV. The aortomesenteric angle (AMA), which typically ranges from 38° to 56° between the SMA and AA, can be calculated using sagittal CT images. The LRV may experience external compression if the AMA drops to 9–35°. In fact, when AMA is less than 35°, anterior NCS may be diagnosed. The pelvic varices and dilated gonadal veins can also be seen on CT.⁶ Retrograde venography, which can measure the renocaval pressure gradient and contrast map the dilated gonadal vein and the perihilar, periureteral, and pelvic collateral network, is still the gold standard for diagnosing nutcracker syndrome.⁷ A LRV diameter ratio (hilar to- aorto-mesenteric ratio) ≥ 4.9 was the most specific finding with CT for NCS (specificity 100%).⁷

The diagnosis of UPJO is aided by the precise anatomic and functional information provided by contrast-enhanced CT scans. Because it offers quantitative information about varying renal function and blockage, even in hydronephrotic renal units, diuretic renography is still an effective test for identifying UPJ and ureteral obstruction.⁸ Renal scintigraphy measures both the total and split renal function.⁹ But renal scintigraphy is not available in our country.

Laparoscopic pyeloplasty has replaced open pyeloplasty as the preferred method of treating PUJO. Despite its high cost, robotic-assisted laparoscopic pyeloplasty may play a role in the future. Deterioration of renal function or symptoms such flank discomfort or infection are reasons to consider surgery.⁴ Within the proper follow-up protocols, successful conservative therapy of asymptomatic cases in youngsters has been recorded.¹⁰ The robotic approach seems to add further technical advantages when compared to conventional LP but sustains a higher costs. Currently, the choice to adopt one of the different minimally invasive approaches depends on the surgeon's preference or experience, and on institutional availability.¹¹

Conservative therapy is advised for patients with NCS who present with modest hematuria or mild and manageable symptoms. However, in cases of severe symptoms such as flank or abdominal pain, anemia, autonomic dysfunction, impairment of renal function, including persistent orthostatic proteinuria, varicocele formation, and ineffective conservative measures after 24 months in patients under the age of 18 and after 6 months in adults, surgery may be considered.¹²

Conclusion

Nutcracker syndrome is rare cause of flank pain and hematuria. Concomitant left NCS and right PUJO obstruction is rare case scenario. Based on clinical manifestation, patient conditions and multiple factors both conservative management and surgical intervention are an option, either open or laparoscopic. For our patient we tried laparoscopic pyeloplasty but after applying posterior sutures stenting was difficult then procedure converted to open. Patient had smooth post-operative course. Physician need to draw attention on possibility of this rare association of the two syndromes.

Consent for Publication

Written and signed informed consent was obtained from the patient for publication of this case report and accompanying images. No need of ethical clearance from institution for publication of the case report.

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Disclosure

The authors report no conflicts of interest in this work.

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