Nutcracker Syndrome: A Rare Etiology of Pelvic Congestion Syndrome

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Abstract: Nutcracker syndrome, also known as left renal vein (LRV) entrapment syndrome, is the result of extrinsic compression of the LRV by the superior mesenteric artery, which courses anterior to the vein and causes a dilated proximal portion of the LRV and an occlusion of the mid to distal renal vein. This results in collateral drainage through dilated gonadal, perirenal, and periureteral vessels. Variceal collateral veins, along with hypertension in the LRV, results in left flank pain, orthostatic proteinuria, microscopic or macroscopic hematuria, pelvic congestion, lower limb varices, and varicoceles. In this case report, we describe the case of 34-year-old woman with pelvic congestion syndrome who was found to have nutcracker syndrome and was successfully treated with intravascular ultrasound-guided percutaneous intervention with a stent placement.

VASCULAR DISEASE MANAGEMENT 2021;18(11):E204-E205.
Key words: left renal vein entrapment, nutcracker syndrome, pelvic congestion syndrome

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Case Report

A 34-year-old woman with a history of hypotension, anemia, and asthma was referred to the cardiology clinic for evaluation of pelvic congestion syndrome (PCS). She reported longstanding constant, diffuse, progressively worsening, dull, aching pelvic and lower abdominal pain, exacerbated by exertion and sexual intercourse, for almost 2 years. She was amenorrheic and taking depot medroxyprogesterone for the past 2 years. Physical exam was significant for normal vital signs and tenderness over the lower abdominal quadrants, bilateral levator, and lumbar spine. Labs were significant for hemoglobin (10.7 mg/dL), hematocrit (33.4%), and microscopic hematuria. Computed tomography (CT) scan of the abdomen and pelvis with contrast was significant for mild right-sided hydronephrosis and a 3 mm right distal ureteral calculus. Transabdominal and transvaginal pelvic ultrasound showed prominent pelvic vasculature with a small amount of free pelvic fluid, suggestive of PCS. Inferior vena cava (IVC) venography and renal vein angiography with

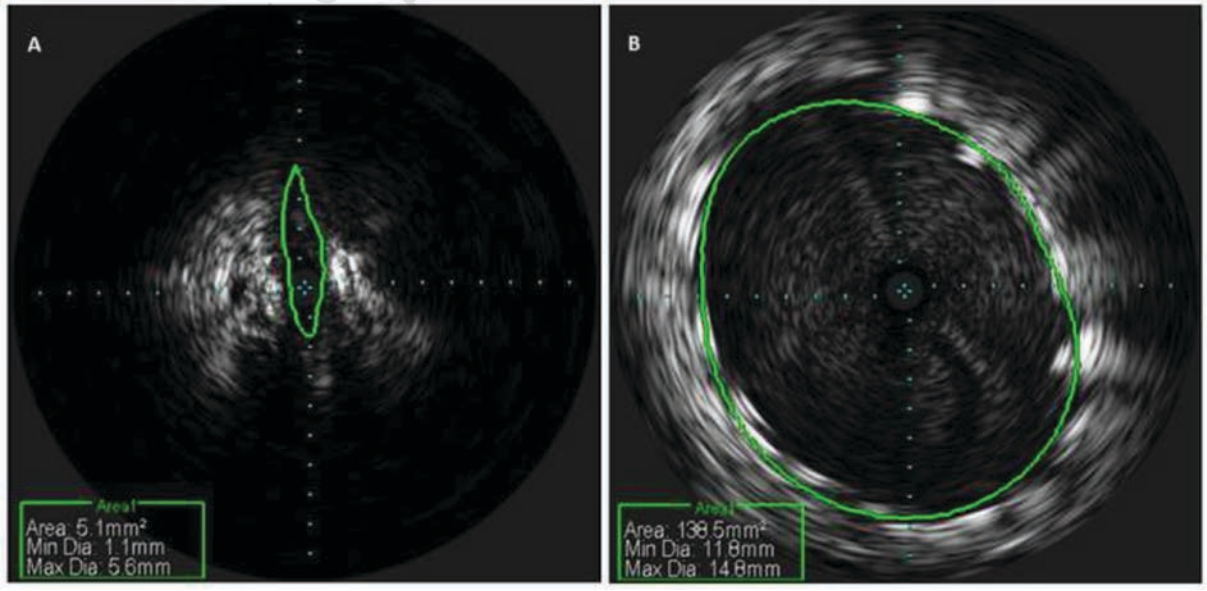

Figure 1. Intravascular ultrasound images of the distal left renal vein pre **(A)** and post **(B)** percutaneous intervention. Note the increase in lumen diameter after the stent placement.

intravascular ultrasound (IVUS) was performed with a high suspicion for nutcracker syndrome. It revealed a normal IVC, a dilated proximal left renal vein (LRV) and gonadal vein with varicose veins, and a complete obliteration of the lumen from the mid to distal LRV adjacent to the superior mesenteric artery (SMA). Balloon angioplasty with an 8.0 balloon and self-expanding 14 x 4 cm stent was placed in the LRV, overlapping with another 6 x 4 cm stent. IVUS after stent placement revealed the dimension of the LRV to be 40 mm, with no compression at the end of the procedure. The patient was started on aspirin and clopidogrel and was followed up in clinic.

Discussion

NCS, also known as LRV entrapment syndrome, is a condition characterized by extrinsic compression of the LRV by the SMA, which courses over the vein, resulting in an impedance to the flow of blood from the LRV into the IVC and distention of the distal portion of the vein.[1] Anatomically, NCS has two variants: anterior and posterior. The anterior variant is characterized by the compression of a normally situated LRV by the abdominal aorta and SMA. The posterior variant is characterized by retro-aortic or circum-aortic LRV compression, usually between the abdominal aorta and the vertebral column.[1,2] Other uncommon causes include pancreatic neoplasm, para-aortic lymphadenopathy, retroperitoneal tumors, overarching testicular artery, and strangulating fibro-lymphatic tissue between the SMA and the aorta. Compression of the right renal vein is rare and is usually associated with pregnancy or a left-sided IVC, hemiazygos continuation, and persistent left superior vena cava combination.[3]

The impedance to the outflow of LRV causes LRV hypertension with a measurable renocaval pressure gradient. Normal pressure gradient is < 1 mm Hg. A renocaval pressure gradient of > 2 mm Hg is suggestive of NCS. The pressure gradient may raise up to 3 mm Hg, leading to the rupture of the thin-walled septum between the small veins and collecting system in the renal fornix, leading to hematuria. LRV hypertension leads to the formation of varices and collateral drainage through dilated gonadal, perirenal, and periureteral vessels, resulting in left flank pain, orthostatic proteinuria, microscopic or macroscopic hematuria, pelvic congestion, lower limb varices, and varicocele.[4]

NCS is a clinical diagnosis; there is no common consensus or a definite diagnostic criterion. It is usually a diagnosis of exclusion in patients presenting with chronic abdominal pain, flank pain, and hematuria. Noninvasive imaging techniques such as ultrasound of the abdomen and pelvis, with and without Doppler; CT; magnetic resonance imaging (MRI); CT angiography; and MRI angiography can be used for initial screening and to aid in ruling out other common etiologies of the presenting complaint.[1] Transabdominal or transvaginal ultrasound is the cheapest and most readily available test. An aortomesenteric vein to renal vein hilar segment ratio > 4.16 along with a peak velocity of > 3.98 m/s is suggestive of NCS.[1] MRI and CT angiography help in delineating the anatomic structures and can demonstrate the NCS along with collaterals and varices. Invasive techniques, such as venography with or without IVUS, can be considered the gold standard for diagnosis. A renocaval pressure gradient of > 2mm Hg is diagnostic of NCS.[4]

Management strategies depend on the severity of the symptoms, anatomy, and extent of renal vein hypertension. A conservative approach with symptomatic management and surveillance with routine lab investigations are appropriate for patients with mild or atypical disease. It is hypothesized that collateralization occurring over time helps reduce renal vein hypertension and alleviates symptoms. An interventional approach includes surgical strategies such as nephrectomy, variceal ligation, nephropexy, and renocaval reimplantation.[1] Minimally invasive endovascular approaches, such as percutaneous transluminal angioplasty with stent placement of the compressed vessel, can be considered in select patients with good results.[4]

Conclusion

NCS is well-described in the literature but its heterogeneity in presentation often poses diagnostic challenges. Due to the variability in presentation and lack of common consensus or diagnostic criteria, a high index of suspicion is often warranted in patients with symptoms suggestive of NCS. ■

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Disclosure: *The authors have completed and returned the ICMJE Form for Disclosure of Potential Conflicts of Interest. The authors report no conflicts of interest regarding the content herein.*

Manuscript accepted September 28, 2021.

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REFERENCES

1. Gulleroglu K, Gulleroglu B, Baskin E. Nutcracker syndrome. *World J Nephrol.* 2014;3:277-281.
2. Shah D, Qiu X, Shah A, Cao D. Posterior nutcracker syndrome with left renal vein duplication: an uncommon cause of hematuria. *Int J Surg Case Rep.* 2013;4:1142-1144.
3. Yildiz AE, Cayci FS, Genc S, Cakar N, Fitoz S. Right nutcracker syndrome associated with left-sided inferior vena cava, hemiazygos continuation and persistent left superior vena cava: a rare combination. *Clin Imaging.* 2014;38:340-345.
4. Venkatachalam S, Bumpus K, Kapadia SR, Gray B, Lyden S, Shishehbor MH. The nutcracker syndrome. *Ann Vasc Surg.* 2011;25:1154-1164.