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S. LeeAnn Denham MD, Felix A. Hester RDMS RVT, Therese M. Weber MD (Mentor)
The University of Alabama at Birmingham; Birmingham, Alabama

Case

A thin 27 year-old female presented to the emergency room with a hematocrit of 17, 3+ hematuria, and 3+ proteinuria. The patient reported a 6-month history of dark red/tea colored urine and intermittent flank pain. She described the pain as sharp and cramping, 10/10 in severity, intermittent (lasting for 10 to 15 minutes), and exacerbated by urination. The patient was admitted and had multiple subsequent admissions for anemia and unrelenting hematuria. A follow-up contrast-enhanced computed tomography (CT) scan demonstrated apparent 'pinching' of the left renal vein between the superior mesenteric artery (SMA) and aorta and suggested Doppler renal ultrasound to evaluate for nutcracker syndrome. On CT, the anterior-posterior (AP) diameter of the dilated and narrowed portions of the left renal vein measured 16 mm and 3.5 mm, respectively, resulting in an AP diameter ratio of 4.6 (dilated/narrowed).

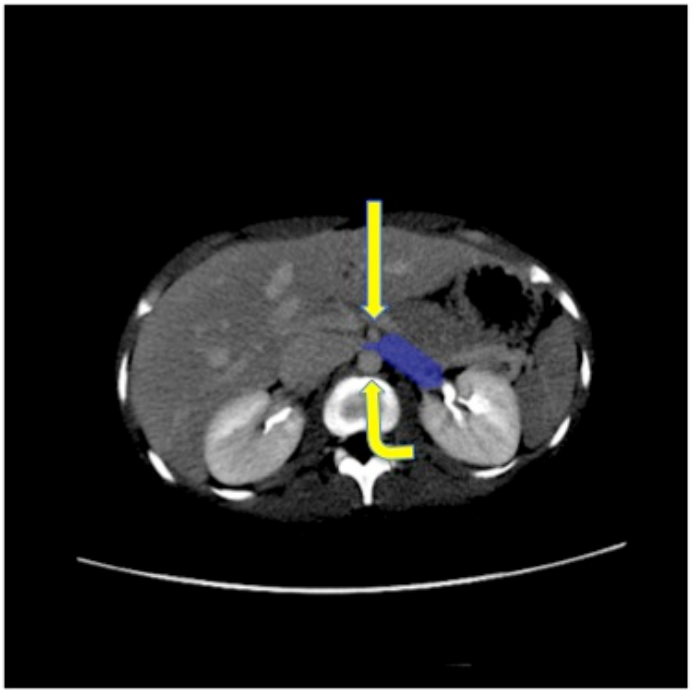

FIGURE 1. CT showing the left renal vein being 'pinched' between the aorta (curved arrow) and SMA (straight arrow). Note the proximal dilation of the left renal vein and narrowing between the aorta and SMA (blue highlight).

Doppler ultrasound imaging confirmed nutcracker anatomy and peak velocities consistent with nutcracker syndrome. On ultrasound, the left renal vein was dilated proximally and narrowed as it passed between the aorta and SMA. Peak velocity within the dilated and narrowed portions of the left renal vein was 8 cm/sec and 171 cm/sec, respectively, resulting in a peak velocity ratio of 21 (narrowed/dilated).

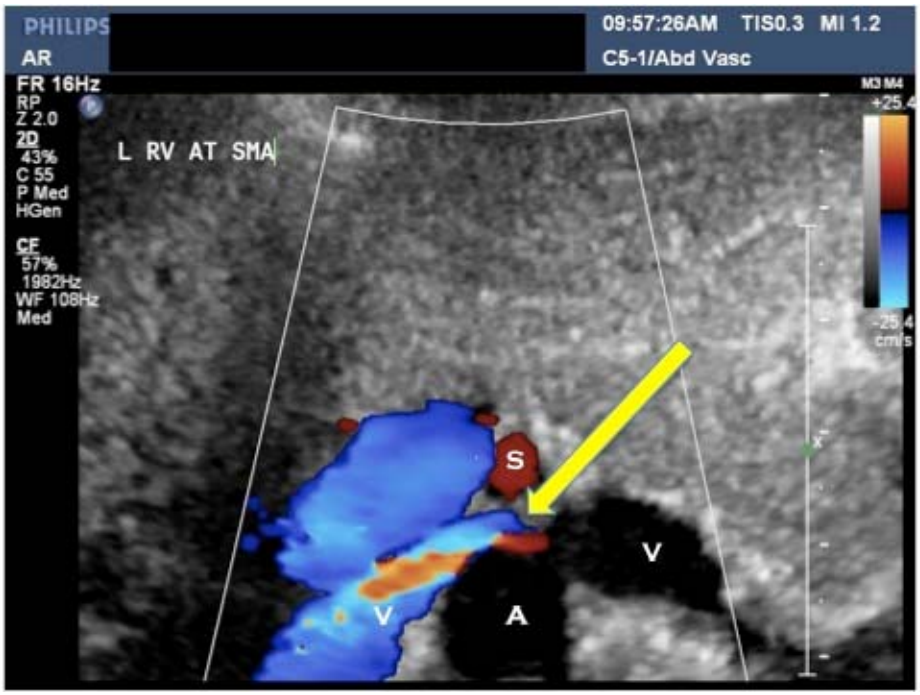

FIGURE 2. Left renal vein (V) coursing between the aorta (A) and SMA (S). Note the pre-stenotic dilation and narrowing of the vein between the aorta and SMA (arrow).

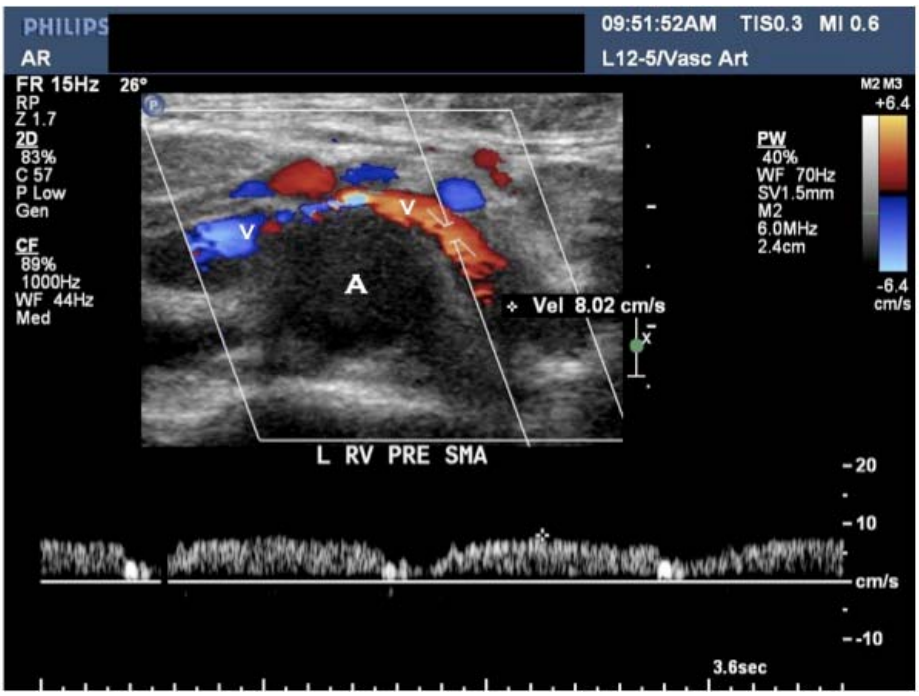

FIGURE 3. Left renal vein (V) stretching across the aorta (A). The peak velocity of the blood within the pre-stenotic vein is 8 cm/sec.

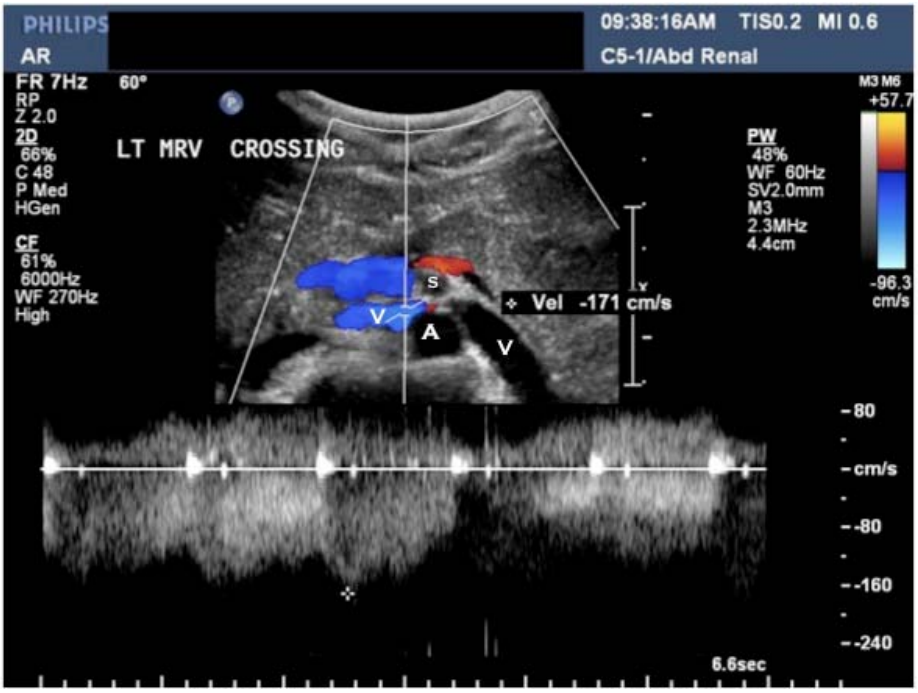

FIGURE 4. Left renal vein (V) being narrowed between the aorta (A) and the SMA (S). The peak velocity in the narrowed portion of the left renal vein is 171 cm/sec.

Subsequent cystoscopy showed bloody efflux from the left ureteral orifice. Left renal vein selective venography was then performed with concomitant intravascular ultrasound which demonstrated 50-60% narrowing of the left renal vein at its confluence with the inferior vena

cava (IVC) secondary to compression by the SMA. The diameter was reported to be 4 mm in the narrowed portion of the left renal vein while the dilated portion measured 8-9 mm. Angioplasty, without stenting, was performed at this time. However, the patient returned to the emergency room in approximately one month with continued complaints of left flank pain and gross hematuria.

The patient then underwent left renal vein mobilization and decompression performed through a midline approach. Nutcracker compression of the left renal vein by the aorta and SMA was seen intra-operatively. Compression was relieved upon complete mobilization of the left renal vein.

Diagnosis

Nutcracker syndrome – extrinsic compression of the left renal vein between the aorta and superior mesenteric artery resulting in characteristic symptoms.

Discussion

The exact prevalence and incidence of Nutcracker syndrome (NCS) is unknown, likely secondary to the variability of symptoms and lack of consensus diagnostic criteria [1][2]. However, as evidenced by the small number of reported cases, it is rare. Based on these cases, a slight female predominance and symptomatic incidence in the 2nd-4th decades is typically seen [1][2]. NCS often goes underdiagnosed, especially in patients who are asymptomatic [1]. Understanding the clinical manifestations and diagnostic criteria for NCS is important to effectively diagnose patients with a variety of urological or gynecological symptoms for which a vascular etiology is less often considered.

Nutcracker syndrome is also called 'left renal vein entrapment' and describes extrinsic compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA) resulting in obstructed blood flow from the LRV to the inferior vena cava (IVC) [1]. This typically occurs when the SMA branches from the aorta at an acute angle; this angle is usually less than half the normal 90° angle. An inverse relationship between body mass index and NCS has been observed, and is likely secondary to a paucity of retroperitoneal fat resulting in a decreased branching angle of the SMA [1]. However, fibrosis around the SMA, posterior left renal location, and vascular anomalies can also lead to NCS. Such anomalies include abnormal configuration, branching, or origin of the SMA [2]. Compression of a circumaortic or retro-aortic left renal vein between the aorta and vertebral column has also been reported to cause NCS, though this entity is more appropriately called 'posterior nutcracker syndrome' [2][3].

The most common symptom of NCS is hematuria, which can be gross or microscopic; however, hematuria is not necessary for a diagnosis of NCS. Hematuria is attributed to the rupture of thin-walled veins into the collecting system secondary to LRV hypertension [1][3]. Patients with NCS also often present with abdominal pain and left flank pain. The pain is postulated to either be secondary to venous congestion or as a result of renal colic secondary to the passing of blood clots [1][2]. When increased venous pressures result in the formation of collaterals, pelvic congestion syndrome can also occur. This results in lower abdominal pain, dysuria, dyspareunia, dysmenorrhea, and pelvic

variceal formation [1][2]. Mild to moderate proteinuria has also been seen in approximately half of patients with NCS [3][4].

Diagnosis is made by careful history and a variety of diagnostic tests. Diagnostic algorithms typically include urine microscopy and culture, Doppler ultrasound (DUS), cystoscopy, CT or Magnetic Resonance (MR) angiography, and venography [1][2]. Although usually only performed in severe cases, the gold standard for diagnosing NCS is venography with reno-caval pressure gradient determination. A greater than 3-mmHg pressure gradient between the IVC and the LRV is diagnostic of NCS; the normal gradient ranges from 0 to 1 mmHg [1][2].

Doppler ultrasound is non-invasive, does not expose the patient to radiation, and is capable of evaluating the velocity of blood within the LRV. DUS, therefore, should be utilized as the first-line imaging modality for diagnosis of NCS in both children and adults. The sensitivity and specificity of DUS ranges from 69-90% and 89-100%, respectively [1].

Nutcracker syndrome DUS criteria is based on both the ratio of the peak velocity in the narrowed and dilated portions of the LRV and the ratio of the AP diameter of the LRV in the dilated and narrowed portions. The peak velocity ratio cutoff for NCS ranges from 4.1 to 5.0 (narrowed/dilated) and the AP diameter ratio cutoff ranges from 4.0 to 5.0 (dilated/narrowed) [3][4][5].

Cystoscopy may show left-sided unilateral hematuria [1]. CT and MR angiography also have a high diagnostic yield with MR having the advantage of being radiation free [2][6].

Treatment for NCS varies with symptom severity and ranges from surveillance to open surgical intervention [1][2]. Observation is the treatment of choice in patients younger than 18 years old because as many as 75% have resolution of hematuria within 2 years [1]. External and intravascular stenting have recently been employed, however, long-term follow-up data is not available. Surgical intervention is considered in patients with unrelenting, severe hematuria, pain, or renal insufficiency as well as those who have not responded to 24 months of conservative treatment. Surgical interventions include LRV transposition (with or without a Dacron wedge placed between the SMA and aorta), LRV bypass, SMA transposition, renal-to-IVC shunt, gonadocaval bypass, medial nephropexy with excision of renal varicosities, renal autotransplant, and even nephrectomy [1].

NCS is a rare, underdiagnosed phenomenon that has substantial morbidity. A working knowledge of this entity and its diagnostic criteria is important for physicians in many specialties. Doppler ultrasound is non-invasive, radiation-free, and has accurate diagnostic parameters for NCS, making it the imaging modality of choice to readily and effectively evaluate and diagnose NCS.

References

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