Interventional Radiology

Nutcracker syndrome in adolescent with perineal pain
An interesting case of an adolescent with perineal pain due to pelvic congestion from nutcracker syndrome with relief after balloon venoplasty and sclerotherapy

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ABSTRACT

Nutcracker phenomenon is the descriptor for a patient’s anatomy whenever the left renal vein becomes compressed between the abdominal aorta and the superior mesenteric artery. Nutcracker syndrome is the terminology used when the nutcracker phenomenon is accompanied by symptoms including pain (abdominal, flank, pelvic), hematuria, and orthostatic proteinuria. Diagnosis can be made with Doppler ultrasound, venography, computed tomography, or magnetic resonance imaging. This case demonstrates some of the typical findings of nutcracker syndrome. The limited clinical features and interesting imaging findings, in addition to the young age of the patient, make this a notable case.

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Case report

A 13-year-old girl presented with a chief complaint of perineal pain of 5 weeks duration. Two weeks before, the patient had complained of coccygeal pain, thought to be secondary to a fall during cheerleading practice, although no incident was reported. The patient’s pain at baseline rated at 2 of 10, which increased when bearing down while stooling or sneezing. Initial workup included a sacrococcygeal radiograph to rule out...
fracture, which only demonstrated a moderate stool burden. The patient was subsequently started on Mirilax therapy twice daily, resulting in regular bowel habits. Her pain persisted, becoming so severe that she stayed at home from school for 2 weeks.

The patient’s medical history was remarkable for Helicobacter pylori gastritis, chronic constipation, exercise-induced asthma, multiple allergies, and anaphylaxis to nuts. Surgical history included tonsillectomy, adenoidectomy, and colonoscopy. The patient was adopted and lived with her adoptive mother, father, and younger sister. She was active in cheerleading and soccer. She denied alcohol, drugs, tobacco, and sexual activity. She did well in school, although had missed multiple days due to pain, which included the inability to sit at her desk for more than a few minutes secondary to perineal pain.

Review of systems revealed no saddle paresthesia, numbness of the extremities, or pain radiation. She denied lower limb weakness, changes in gait, loss of balance, or bladder and bowel incontinence. Lastly, she denied fever, decreased appetite, emesis, and diarrhea. The patient reported pain localized at her perineal and sacral regions, which worsened with any action that involved bearing-down (sneezing), sitting on hard surfaces, and activities such as running. Finally, the patient endorsed a moderate weight gain, which she attributed to the considerable decrease in physical activity from the pain.

On physical examination, the patient’s vitals were normal. She had mild lower abdominal tenderness to palpation. There was no edema. She also had perineal tenderness at the right and left lower regions between the vagina and anus. A speculum examination was performed under anesthesia, and no visible perineal varicosities were seen at the vaginal introitus. The patient’s laboratory values at the time (including urinalysis and culture) demonstrated normal values. Following this examination, the patient left the office without clear etiology for her symptoms. Over the next 2 weeks, the patient’s pain gradually became more severe with radiation up her back and around her hips. The patient’s symptoms were not relieved by positional changes and became recalcitrant to ibuprofen (initially dosed at 400 mg as needed and then increased to 800 mg as needed), which led her to visit the emergency room.

Because of the unclear etiology of the patient’s pain, multiple imaging studies were ordered including pelvic magnetic resonance imaging (MRI) and ultrasound. MRI with and without contrast of the patient’s pelvis demonstrated dilated enhancing serpentine vessels extending to the left lateral aspect of the uterus (Figs. 1A-E). These findings were most consistent with pelvic varicosities or plexiform neurofibroma. Ultrasound evaluation was then performed. Dynamic transperineal ultrasound, with Valsalva maneuvers and Doppler, demonstrated multiple dilated perivaginal, left adnexal, and uterine veins (Figs. 2A-D). These sonographic findings further supported pelvic venous congestion vs vascular malformation.

Computed tomography (CT) of the abdomen and pelvis with contrast was performed next and it demonstrated a compressed left renal vein (LRV) at the level of the superior mesenteric artery (SMA) with a “beak sign” (rapid tapering of the vessel to a point) [1] (Figs. 3A and B). Reconstructed sagittal CT imaging demonstrates the angle of the SMA from the abdominal aorta (AA) acute at 24 degrees (Fig. 3B), less than...
the normal 60-90 degree angle, suggestive of nutcracker syndrome (NCS).

The left gonadal vein was noted to be of normal caliber. At this point, the patient was referred to vascular surgery and interventional radiology for further care.

Left femoral vein injection demonstrated mild compression of the left common iliac vein by the right common iliac artery with minimal reflux into the left internal iliac vein (Fig. 4A). LRV injection demonstrated reflux into the gonadal, renal capsular, and paralumbar veins. There was also evidence of

Fig. 2 – (A-D) Superficial longitudinal view of patient’s left labial area demonstrating filling of paralabial vessels with valsalva maneuver (A) without valsalva, (B) with valsalva. Similar portrayal with transverse views (C) without valsalva, (D) with valsalva.

Fig. 3 – (A, B) Axial computed tomography (CT) images with contrast demonstrating beak sign the left renal vein (A), sagittal CT with contrast demonstrating acute angle of superior mesenteric artery and abdominal aorta at roughly 39 degrees (B).
diminished contrast drainage of the LRV into the inferior vena cava (IVC), suggesting compression of the LRV (Fig. 4B).

Venous pressure measurements demonstrated a LRV to IVC cava gradient of 3 mm Hg (normal < 1 mm Hg) [2]. After balloon angioplasty, there was no considerable angiographic change. Measurement of IVC to left common iliac vein after PTA demonstrated reduction of the gradient by 1 mm Hg. In summary, the left iliac venogram with venoplasty demonstrated mild left common iliac vein compression syndrome with minimal response to venoplasty and LRV compression from the MSA without response to venoplasty.

LRV stent was considered; however, this more definitive therapy was deferred until growth was complete. Conservative management was followed to include lifestyle changes and analgesia use. After 2 months of persistent pain, symptoms were readaddressed by vascular surgery and interventional radiology, with sclerotherapy of the pelvic varicosities. Left renal venogram demonstrated prominent reflux into the retroperitoneal, lumbar, and gonadal veins (Fig. 5A). When the catheter was advanced into the gonadal vein, venography demonstrated multiple tortuous gonadal veins (Fig. 5B) and grade III reflux in the periuterine varices with filling into perineal or vulvar varices was discovered (Fig. 5C). Sclerotherapy was performed in the distal left gonadal vein, resulting in decreased reflux (Figs. 6A and B). Next, the vulvar or peroneal varicosities were directly accessed percutaneously, and sclerotherapy was performed using 98% dehydrated ethanol (Figs. 6C and D).

Immediately after the procedure, pain was significantly decreased. At 4 months, the patient had a second sclerotherapy procedure using a slurry made with Gelfoam (Pfizer Inc., NY, NY) and 2 mL of 3% Sotradecol administered to the left gonadal and deep pelvic presacral veins, with similar positive results and continued to be followed by vascular surgery and interventional radiology until many years later.

**Discussion**

The patient above demonstrates pelvic congestion syndrome secondary to NCS. Chait et al. described the anatomic positioning of the LRV sandwiched between the SMA and AA as the LRV is positioned between the 2 arms of a nutcracker [3]. Compression of the LRV by the SMA can cause impeded outflow from the LRV to the IVC [1,2]. Venous hypertension at the LRV develops and collateral vessels form around the renal capsule. Initially, there are no signs of reflux in the left ovarian vein due to competent venous valves and patients remain asymptomatic. This asymptomatic, yet anatomically significant finding is termed nutcracker phenomenon [1,2]. At later stages, the heightened pressure leads to valvular incompetence and reflux into ovarian and lumbar veins, ultimately causing the development of pelvic or vulvar varices [4]. These varices are typically what cause pelvic congestion symptoms such as abdominal or pelvic pain, dysuria, dysmenorrhea, dyspareunia, or other clinical signs such as hematuria and orthostatic proteinuria [5]. The development of symptoms is typically observed in

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**Fig. 4 – (A, B) Images from the venogram demonstrating impression on the left common iliac vein by the right common iliac artery with minimal reflux into the left internal iliac vein (A). Left renal venogram shows left renal vein compression by the superior mesenteric artery (orange arrow) with evidence of collateral vessels (blue arrow) (B).**

**Fig. 5 – (A-C) Images from left renal and gonadal venograms before sclerotherapy demonstrating reflux into retroperitoneal, lumbar, and gonadal veins (A), gonadal veins (B), and parauterine, perineal, and vulvar varices (C).**
middle-aged women and it is at this point that they are diagnosed with NCS [1,2,5].

The diagnosis of NCS is based on the patient’s clinical examination and radiographic findings. Hematuria is noted to be the most common clinical feature and often what drives a patient to the clinician [2,6]. Gulleroglu et al. noted that there are 2 classifications of patients presenting with features of NCS. The authors note patients to have a renal presentation (symptoms of hematuria, orthostatic proteinuria, and flank pain) or a urologic presentation (symptoms of abdominal pain, varicocele, dyspareunia, dysmenorrhea, fatigue, and orthostatic intolerance) [1]. In the case of the patient above, she presented solely with perineal and lower abdominal pain, placing her most likely within the urologic class; however, with just one symptom, a definitive diagnosis of NCS as the source for her pain would be challenging. Therefore, her radiographic findings were essential in solidifying her diagnosis.

Doppler ultrasound is the initial diagnostic modality used in patients suspected of having NCS [1,6,7]. With Doppler ultrasound, tortuous pelvic vessels with large diameters (5-6 mm), decreased or reversed flow velocities, can be visualized [7]. Retrograde venography (RV), however, is considered the gold standard modality for making a diagnosis [1,7]. RV can depict pressure differences between the area of compression in the LRV and the IVC. The normal change in pressure between these 2 vessels is less than or equal to 1 mm Hg [2]. Diagnosis of NCS is supported when this pressure difference is equal to or greater than 3 mm Hg [7]. Other modalities used to further demonstrate nutcracker anatomy include CT and MRI. Both modalities can demonstrate the angle between the SMA and the aorta. The tighter this angle becomes, the more compression applied to the LRV. The normal angle of these two arteries is 25-60 degrees. In patients with NCS, this angle is often less than 35 degrees, but anything less than 90 degrees is considered a significant finding [6]. CT and MRI can also demonstrate the “beak sign” of the LRV, which is simply narrowing of the LRV due to the SMA-AA compression [1,7].

Treatment for NCS varies based on the age of the patient and scope of symptoms. Typically, patient’s less than 18 years of age are managed conservatively. Often with physical development and growth, their condition and symptoms will resolve. Increased fat and fibrous tissue will encompass the origin of the SMA and relieve the compression on the LRV [6]. Supporting this theory, the authors noted that increases in body mass index have lead to symptom regression in affected individuals [8,9]. Until such relief is met, analgesics may be used for pain control, angiotensin converting enzyme inhibitors have been tried to improve proteinuria, and aspirin therapy has been initiated to improve left-to-right renal perfusion ratios [6]. This conservative approach is maintained for roughly 24 months.

If after this time, the patient continues to have life-altering symptoms of venous congestion, surgery becomes the therapy

Fig. 6 – (A-D) Pre- and postembolization of the distal left gonadal vein (A and B). Pre- and postpercutaneous sclerotherapy of the left vulvar varicosities (C and D) both demonstrating decreased aberrant and dilated vessels.
of choice [6,8]. Surgery is also the course of care for patients older than 18 years of age that are persistently symptomatic for greater than 6 months [6]. Multiple surgical recommendations have been made for the alleviation of symptoms caused by NCS. The initial therapy of choice is endovascular stenting of the renal vein and may be accompanied by embolization of the gonadal veins, in hopes for decreasing the flow through varicosities [4,7]. Sclerotherapy may also be used on pelvic varicosities [4]. If these procedures do not lead to resolution of patient symptoms, other surgical procedures such as nephropexy, transposition of the LRV or SMA, gonadocaval bypass, renal autotransplantation, or even nephrectomy have been implemented as treatment [8].

This patient did demonstrate mild findings of May-Thurner anatomy. However, the lack of robust reflux during pelvic angiography and no findings for May-Thurner syndrome led us to suspect NCS as the primary cause of her pelvic congestion. May-Thurner syndrome has been reported as a contributing factor in pelvic congestion syndrome [10,11].

In conclusion, the patient presented in the case above elicited many exciting findings atypical for pediatric patients with nutcracker anatomy. First, most pediatric patients with LRV compression are asymptomatic or present with asymptomatic hematuria, and the nutcracker anatomy is simply an incidental finding on imaging studies. Our patient’s main symptom was perineal and lower abdominal pain that became debilitating. She also never demonstrated signs or had laboratory values for hematuria. Second, the degree to which the patient developed varicosities and collateral veins, with her ultrasound demonstrating signs similar to varicocele, is not commonly seen in female patients with NCS. Collateral vessels and varicosities may form, but the extent to which they are seen on our young patient’s imaging is remarkable. Third, symptoms of NCS will often arise following a sudden weight loss. Our patient, however, had complaints of weight gain. Lastly, this case further exemplifies that in young female patients with chronic pelvic pain of unknown etiology, imaging with transvaginal dynamic Doppler ultrasound and CT or MRI is essential for diagnosis and planning for treatment.

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