CASE REPORT

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A Rare Condition in Repeat Varicocele; Nutcracker Syndrome

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ABSTRACT This case report describes a rare occurrence of Nutcracker syndrome (NCS) in a 21-year-old male with recurrent varicocele. NCS is caused by compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery, leading to venous hypertension and a variety of symptoms, including testicular pain, hematuria, and pelvic congestion. The patient initially presented with left testicular pain and grade 3 varicocele, which persisted after varicocelectomy. Further diagnostic imaging with contrast-enhanced computed tomography revealed compression of the LRV, confirming NCS. Management of NCS depends on symptom severity, with conservative treatment preferred for mild cases and surgery considered for severe cases. In this instance, the patient was referred for further follow-up with cardiovascular surgery. NCS is a rare but significant cause of recurrent varicocele and should be considered in patients with unresolved or recurring symptoms after varicocele surgery.

Keywords: Nutcracker syndrome; varicocele; superior mesenteric artery

Nutcracker syndrome (NCS) is typically defined as left renal vein (LRV) stenosis due to compression of the LRV between the aorta and the superior mesenteric artery (SMA), leading to dilation in the renal vein before the compressed segment.[1] This condition is sometimes referred to as anterior NCS. Compression may also occur when the LRV follows a circumaortic or retroaortic path between the aorta and the vertebral body, referred to as posterior NCS.[2]

Possible causes include posterior renal ptosis, abnormally high exit of the LRV or an abnormal, narrow-angle aortic outlet of the SMA. LRV compression can also occur due to pancreatic masses or lymphadenopathy.[3] The clinical presentation varies, from asymptomatic hematuria to severe pelvic congestion. Patients may present with hematuria, orthostatic proteinuria, flank pain, abdominal pain, varicocele, dysmenorrhea, weakness, and fatigue.[4]

It is noted in the literature that more patients may suffer from NCS than previously thought, likely due to under-recognition of the disease. The key to diagnosis is suspicion of the syndrome. NCS can be diagnosed using imaging techniques such as Doppler ultrasonography, computed tomography (CT) angiography, magnetic resonance angiography, or retrograde left renal venography, which is the gold standard despite its invasiveness.[5] Doppler ultrasonography is often the 1st choice due to its non-invasive nature.

In this case report, I will present a patient who underwent varicocelectomy in our clinic but continued to experience pain and low sperm count post-operatively. The patient was diagnosed with NCS during angioembolization.

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The informed consent form was obtained from the patient participating in the study. A 21-year-old male patient, single, with no additional diseases, no history of surgery, and non-smoker, first applied to our clinic in August 2022 with complaints of pain and swelling

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in the left testicle. On physical examination, a grade 3 varicocele was detected on the left side. Scrotal Doppler ultrasound showed dilated left plexus pampiniformis veins, measured at 3.1 mm at the widest point, and reflux flow was observed with the Valsalva maneuver. A spermiogram was requested for the patient.

The patient's spermiogram results are shown in Table 1.

After evaluating the current ultrasound and spermiogram results, the risks were explained to the patient. Preoperative anesthesia consultation was done, and an Physical Status Classification System (ASA)-1 risk was given. In October 2022, left varicocelectomy was performed. However, the patient presented again with testicular pain in the 3rd month after the operation. Doppler ultrasound showed dilated left plexus pampiniformis veins, measured at 3.1 mm, and reflux flow was again observed with the Valsalva maneuver. A new spermiogram was requested.

The postoperative spermiogram results are shown in Table 2.

The patient was then referred to the interventional radiology clinic for angioembolization, where a decision for embolization was made. Contrast-enhanced CT before embolization revealed compression of the LRV between the aorta and the SMA, leading to the diagnosis of NCS.

TABLE 1: Spermiogram results (pre-op)

Examination	Patient's result	Normal range
Duration of sexual abstinence	3	3-5 days
Total mobility	290	38.000-42.000
Inactivity	71	0-1.000
Volume	2	2.000-5.000
Leukocytes	1	1-1.000
Sperm concentration	67	12.000-16.000
Mobile sperm concentration	2	0-1
Forward motile sperm	0	0-1
Average speed	<1	0-1.000
Total sperm concentration	134	33.000-46.000
Forward motion	1	31.000-34.000
Liquefaction	0	0-1.000
Viscosity	0	0-1.000
pH	7.8	7.2-7.201
In-place mobile	230	0-1
Total mobile sperm	30	0-1
Forward mobile sperm	0	0-1

TABLE 2: Spermiogram results (post-op)

Examination	Patient's result	Normal range
Duration of sexual abstinence	3	3-5 days
Total mobility	240	38.000-42.000
Inactivity	76	0-1.000
Volume	2	2.000-5.000
Leukocytes	0	1-1.000
Sperm concentration	64	12.000-16.000
Mobile sperm concentration	15	0-1
Forward motile sperm	0	0-1
Average speed	<1	0-1.000
Total sperm concentration	134	33.000-46.000
Forward motion	0	31.000-34.000
Liquefaction	0	0-1.000
Viscosity	0	0-1.000
pH	7.8	7.2-7.201
In-place mobile	290	0-1
Total mobile sperm	39	0-1
Forward mobile sperm	0	0-1

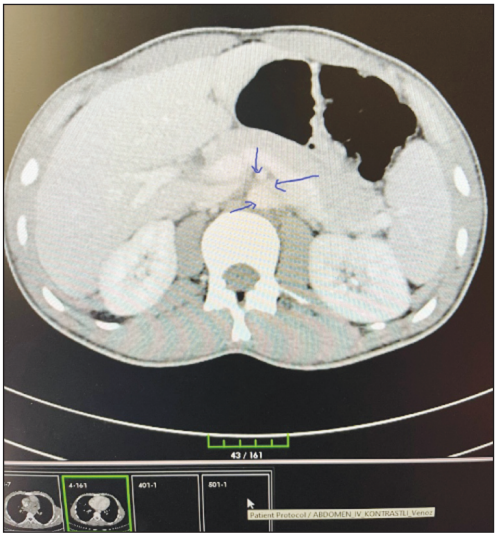

FIGURE 1: Nutcracker syndrome CT imaging

The contrast-enhanced CT image is shown in Figure 1.

Due to this situation, embolization could not be performed. In NCS, different treatments are suggested for adults, including observation, conservative management, or surgery. For children, conservative management is recommended due to spontaneous re-

missions. Medical treatment [e.g., medroxyprogesterone acetate (Provera®, Pfizer, USA), goserelin acetate (Zoladex®, AstraZeneca, UK), and anti-inflammatory drugs] may be given to patients with isolated pelvic congestion. Surgery can be considered for patients with anemia, chronic pelvic pain, and varicocele. Our patient was consulted with cardiovascular surgery, and follow-up was planned before discharge.

DISCUSSION

In conclusion, NCS is a rare condition that is difficult to diagnose. It usually causes clinical symptoms, including microscopic and occasionally macroscopic hematuria, less commonly orthostatic proteinuria, vague flank pain, pelvic congestion findings, and left varicocele. One of the main factors contributing to its rarity is its under-recognition, both in primary health care and in radiological advanced diagnostic studies. Therefore, the most critical step in diagnosing NCS is to consider it as a possibility in the differential diagnosis.

NCS is often suspected based on clinical findings, but confirming the diagnosis frequently requires invasive procedures such as retrograde left renal venography and pressure measurements.[6] The preferred non-invasive screening method for NCS is Doppler ultrasound, as it provides both anatomical and physiological evaluation of the LRV. Cross-sectional imaging with CT or magnetic resonance imaging can also be used, relying on vessel diameter and, to a lesser extent, the angle of the SMA. Normally, the aorto-SMA angle ranges between 45° and 90°. An angle of 35° in the sagittal plane suggests NCS.[7] When invasive surgical treatment is considered, precise diagnosis is often confirmed using invasive tests. Renal venography with direct pressure measurements is regarded as the "gold standard" for NCS diagnosis. A high-pressure gradient (≥3 mmHg) between the LRV and the inferior vena cava serves as the standard reference for diagnosing venous hypertension.[8]

In many cases, especially in pediatric patients, conservative management is sufficient. Pediatric NCS patients often experience spontaneous resolution, likely due to the increase in retroperitoneal and/or mesenteric fat tissue and fibrous tissue accumulation at the SMA origin during growth. The most widely accepted treatment in children is the use of angiotensin-converting enzyme inhibitors, with or without aspirin, to minimize orthostatic proteinuria and maximize renal perfusion.[9,10] For patients with persistent symptoms**,** renal vein transposition is the standard care. This procedure offers excellent immediate and long-term symptom relief and is the most common invasive intervention for anterior NCS. Left kidney autotransplantation**,** though more invasive, is also a relatively common approach. It involves moving the kidney to the iliac fossa, eliminating complications like nephroptosis, and is considered a more comprehensive treatment.[11] A newer, less invasive method involves the laparoscopic placement of an extravascular stent outside the LRV. This technique avoids the need for mobilization or clamping of the LRV, offering a minimally invasive solution for treating NCS.[12-14]

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Muhammed Fatih Keleş;* ***Design:*** *Muhammed Fatih Keleş;* ***Control/Supervision:*** *Kerem Taken;* ***Data Collection and/or Processing:*** *Muhammed Fatih Keleş;* ***Analysis and/or Interpretation:*** *Rahmi Aslan;* ***Literature Review:*** *Mazlum Özkan;* ***Writing the Article:*** *Muhammed Fatih Keleş;* ***Critical Review:*** *Rahmi Aslan;* ***References and Fundings:*** *Kerem Taken;* ***Materials:*** *Kerem Taken.*

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