1. Introduction

Anterior Nutcracker syndrome (NCS) was first described by Grant in 1937. Also known as left renal vein (LRV) entrapment syndrome, NCS most commonly occurs anteriorly when the LRV is compressed between the aorta and the superior mesenteric artery (SMA). Compression of the LRV reduces venous outflow, causes increased pressure within the venous circulation, and develops renal hilar varices. Presentations vary widely, but the most common initial symptom is hematuria. Innovations in imaging have increased the diagnosis of LRV compression in asymptomatic patients. The “nutcracker phenomenon,” this is a standard anatomical variant seen at frequencies of 8–30.5% in asymptomatic individuals. The syndrome affects a higher proportion of female patients and has been recorded from infancy to the seventh decade of life.

Presentations vary widely, but the most common initial symptom is hematuria. Innovations in imaging have increased the diagnosis of LRV compression in asymptomatic patients. The “nutcracker phenomenon,” this is a standard anatomical variant seen at frequencies of 8–30.5% in asymptomatic individuals. The syndrome affects a higher proportion of female patients and has been recorded from infancy to the seventh decade of life. Treatment modalities include surgical management via transposition of the LRV or gonadal vein transposition (GVT). GVT resolves renal vein compression and pelvic congestion. GVT may not always effectively treat symptomatic flank pain, specifically when there is undiagnosed concurrent Loin Pain Hematuria Syndrome (LPHS). Renal autotransplant (RAT) can be considered if GVT fails to relieve congestion and kidney pain. RAT involves removing the affected kidney and placing it in either the right or left iliac fossa. This placement relieves compression as well as de-innervating the kidney, simultaneously resolving the LPHS.

Treatment approaches vary, with a lack of uniform guidelines leaving decision-making to individual providers and institutions. Therefore, decision-making recommendations and objective scoring criteria are needed to guide the treatment of this complex and rare condition.

2. Case Presentation

We present a thirty-five-year-old female with a history of pelvic congestion and left flank pain secondary to NCS. The patient received significant workup for the diagnosis of anterior NSC by vascular surgery. The patient’s first line of treatment for NCS included a GVT to relieve congestion. The GVT was performed similarly in fashion to that presented by Miller et al. (Fig. 1). While the GVT successfully resolved the significant vascular congestion, it failed to remedy the flank kidney pain. Three months after the GVT, the patient reported worsening left flank pain. She was referred to the multidisciplinary team (MDT) at the University of Utah for kidney pain evaluation that included imaging (Fig. 2). The workup included a renal hilar block (RHB). This temporally simulates the potential decrease in pain and predicts the success of nephrectomy. The patient produced positive results from the RHB, and after consultation with urology and transplant surgery, they were identified as a candidate for RAT. With positive results from the RHB, there was adequate confidence that the patient’s flank kidney pain was likely LPHS unaddressed by the
previous GVT. The patient underwent a RAT performed in a similar fashion to that presented by Sood. The RAT was successful in reducing the left flank kidney pain.

3. Discussion

NCS can be a debilitating syndrome that is associated with a variety of symptoms. This patient had anterior nutcracker syndrome with typical imaging findings showing left renal vein compression. The GVT did resolve the vascular congestion but not the chronic flank pain. GVT is the first line of treatment for NCS. If it fails to resolve kidney pain, LPHS should be considered. We theorize that NSC is an insult to the kidney. Even when this insult is resolved, the patient may have persisting kidney pain. To stop this pain, the kidney must be denervated. It should be noted that nephrectomy is the most successful line of treatment.

LPHS is a chronic pain syndrome with associated microscopic or gross hematuria that is not fully explained by glomerular or other kidney diseases. It is theorized that NCS causes an initial vascular insult to a kidney. This initial injury creates an eventual sequela of chronic pain that can persist even after the vascular obstruction is resolved. Multiple medical management strategies for LPHS have been attempted with limited success, including angiotensin receptor blockers, beta-blockers, chronic pain control, and anticoagulation. In addition, surgical or other procedure-based therapies, including RAT, range widely in reported effectiveness in pain reduction.

Recent research has determined methods of better-selecting patients who are most likely to benefit from surgery. Therefore, we recommend that LPHS secondary to NCS treatment should only proceed after assessment by a multidisciplinary team. This collaboration should include clinicians from transplant, urology, interventional radiology, and vascular surgery. This model allows for increased accuracy where pain contracts can be addressed, especially in younger patient populations. Furthermore, the best chance of RAT success is predicted by a response to percutaneous renal hilar blockade (RHB).

This case report includes an algorithm that incorporates previous strategies to identify patients most likely to benefit from GVT vs. RAT. This algorithm incorporates imaging findings, evaluation from an MDT, and an RHB to assess the benefits of a nephrectomy. While not all listed conditions must be present for patients to be a candidate for RAT, the algorithm should act as a guide for teams in reducing pain. Previous research has shown the success of RAT in appropriate patients. Implementing this algorithm could help reduce unnecessary risk from surgery by accurately identifying ideal surgical candidates. This case report was scored demonstrated the successful use of the algorithm. The RAT was successful in removing pain.

4. Conclusion

We propose the University of Utah Nutcracker Scoring & Treatment Algorithm (Table 1) for identifying patients with combined NCS and...
LPHS as a safeguard to prevent unnecessary surgical procedures and further standardization of care. RAT should be considered the last line therapy, as it will only be successful on correctly diagnosed patients.

Credit author statement

Jeffrey Campsen: Supervision, Conceptualization, Writing – review & editing. Alyssa Thorman: Investigation, Writing – original draft, Writing – review & editing. Nicholas Baker: Investigation, Project administration, Writing – original draft preparation, Writing – review & editing. Blake Hamilton: Supervision.

Declaration of competing interest

All authors have no conflict of interest to declare.

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Table 1

Nutcracker syndrome severity & treatment algorithm.

| Level 1 | Level 2 | Level 3 | Level 4 |
|---------|---------|---------|---------|
| CT Imaging | + | + | + | + |
| Renal Venogram | - | - | Pressures > 2 | And/or gonadal vein enlargement w/ collateralization | And/or gonadal vein or lumbar enlargement w/ collateralization |
| Pelvic pain and/or congestion | - | +/- | + | + |
| Prior surgeries or insults to kidney | - | - | - | stent, coil, venous thrombus, hostile abdomen |
| Alternative anatomy** | - | - | - | retroaortic vein, multiple arteries |
| Left Kidney Pain | - | - | - | + |
| Renal Hilar Block* | - | - | - | + |
| Treatment | Observation | Observation and pain management | Left Gonadal Vein Transposition (LGVT) | Renal Autotransplant |

*Prior injuries/insults to kidney may include: stent/coil placement, previous venous thrombus, prior venous transposition, kidney pexy, PCNU, lithotripsy, ureter revision, denervation.

**Retroaortic vein, multiple arteries, acute-SMA angle < 35°

*Should the renal hilar block produce significant reductions in flank pain, consider eliminating the gonadal vein transposition.

Level 4 Nutcracker Syndrome*

| Type A | Type B |
|--------|--------|
| Diagnosis | Nutcracker Syndrome with Loin Pain Hematuria Syndrome | Nutcracker Syndrome with retroaortic vein |
| Additional workup | Renal Hilar Block reduces pain by > 50% | Vascular consult agrees with alternate-native anatomy, where gonadal vein transposition is not possible |
| Treatment | Left Renal Autotransplant | Left Renal Autotransplant |