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CASE REPORT

Resolution of Posterior Nutcracker Syndrome through Left Renal Vein Ligation: A Case Report

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Abstract: This case report presents the clinical course of a 54-year-old male patient diagnosed with posterior Nutcracker Syndrome (NCS). The patient experienced recurrent episodes of macroscopic hematuria and severe left flank pain. He was diagnosed with retro-aortic left renal vein (RLRV), an unusual congenital abnormality, which can result in the compression of the left renal vein between the aorta and the vertebra. Initial attempts at conservative management and endovascular interventions were unsuccessful. An open surgical intervention was ultimately performed. The surgery involved ligation of the left renal vein. Postoperatively, the patient experienced marked improvement in symptoms, with complete resolution of hematuria and pain. Furthermore, renal function remained stable, and no kidney-related complications were reported. This case suggests that, in certain situations where endovascular intervention and transposition of LRV or re-implantation of the left gonadal vein are not feasible, ligation of the LRV may provide a safe and practical treatment option for patients with posterior NCS. Future research is needed to validate these findings.

Keywords: Posterior Nutcracker Syndrome, Hematuria, Abdominal pain, Retro-aortic renal vein, Left renal vein ligation, Surgical intervention

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1. Introduction

Retro-aortic left renal vein (RLRV) is a relatively uncommon congenital anomaly. It is one of the most important anomalies regarding inferior vena cava (IVC) and its branches occurring in 1.8-2.4 of the population [1]. RLRV most commonly drains into the IVC at the same level as the normal left renal vein [2]. This anatomical abnormality is usually asymptomatic and diagnosed in a routinely performed abdominal imaging called the posterior Nutcracker phenomenon [3]. In some patients, mechanical compression of the RLRV between the aorta and the vertebra can cause increased pressure in the left renal vein and thus the left renal venous system, which causes varicosities in the renal pelvis. These varicosities may further communicate with the urinary tract, thereby causing hematuria. The symptoms that may be experienced include microscopic or gross hematuria, secondary anemia, orthostatic proteinuria, venous thrombosis, blood clots in the urinary system, ureteropelvic junction obstruction, left flank pain, varicocele, and inguinal pain. Symptomatic cases are called posterior nutcracker syndrome (pNCS) [4,5]. It is important to note that the fundamental underlying cause for all of these symptoms is the reno-caval pressure gradient [6].

RLRV can be easily diagnosed using ultrasound, computed tomography, or magnetic resonance imaging. The treatment options for pNCS include conservative management, surgical interventions, and endovascular stenting. Conservative management is recommended for younger patients with mild symptoms, as there is a high rate of spontaneous remission [7]. Endovascular stenting is also an option and is considered the preferred treatment by some, although there is limited data. Surgical methods, such as transposition of the renal vein, autotransplantation, and bypass, can be used to relieve symptoms. Open surgical repair is considered the gold standard for treatment for its long-term efficacy. The choice of treatment depends on the severity of symptoms, the patient's age, and individual factors. This report presents a case of pNCS where the management approach was liga-

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tion of the left renal vein by open surgery.

2. Case presentation

A 54-year-old man was referred with left flank pain and gross hematuria by the urology clinic to the vascular surgery department after an investigation for gross hematuria showing only RLRV. The multidetector computed tomography of the abdomen revealed RLRV compressed between the abdominal aorta and vertebral column.

The patient had experienced intermittent episodes of macroscopic hematuria and moderate to severe left flank pain for the past decade, which had become increasingly frequent over the last six months. Initially, the pain was sporadic and related to physical exertion. However, it gradually intensified in frequency and severity, occurring suddenly even when the patient was at rest.

No other symptoms or family history of nephropathy or hematuria were reported. The physical examination, including vital signs, was within normal limits. A CT venography of the abdomen and pelvic region revealed compression of the LRV between the aorta and spinal column. The scan also showed prominent left adrenal veins and a relatively normal left gonadal vein. No signs of pelvic varicose veins were observed.

The diagnosis of pNCS was made. An ascending venogram of the left renal vein from right femoral vein access showed dilated origin of the left renal vein but we could not pass the catheter or guide wire to the hilum of the left kidney to measure reno-caval gradient pressure (Figure 1). This also precluded the endovascular stenting option.

We decided to proceed with a surgical intervention for venous revascularization. At first, we chose to transpose the left gonadal vein which involves transecting it distally and reimplanting it into the inferior vena cava. Despite performing this procedure, the gonadal vein did not seem adequate to drain the kidney and the left adrenal veins were larger and more prominent; So, we decided to transpose the LRV to the anterior of the aorta but there were multiple lumbar veins posterior to this vein. Therefore, after the transaction of these veins, the renal vein was short and its quality did not seem proper for anastomosis. We considered venous bypass with graft or ligation of the LRV while preserving the adrenal veins for drainage. Since our center had a lot of experience in ligating the LRV during open abdominal aortic aneurysm repair with none or minimal adverse effects and we observed the kidney for 30 minutes and there was no obvious congestion in the kidney, we decided to proceed with ligation of the LRV without bypass.

The postoperative period was uneventful, and the patient reported improvement in his pain and gross hematuria which resolved completely by day 12 after surgery. His flank pain completely resolved within 15 days. His preoperative creatinine level was 0.8 mg/dL, and there was no change in the postoperative period. His urine analysis showed no microscopic hematuria one month after surgery and at 6 months, one year, and one and half years later. He has been followed up for 2 years and 3 months, and no kidney-related complications have been observed.

Nine months after the operation the patient underwent an abdominopelvic computed tomography with IV and oral contrast to rule out partial intestinal obstruction at another hospital which showed normal kidneys with no congestion of the left kidney and drainage from the adrenal veins (Figure 2).

To report the course of the disease, informed consent was obtained from the patient and the conditions were reported anonymously.

3. Discussion

Diagnosing pNCS can be challenging, and it should be considered in patients with hematuria originating from the left kidney and an anatomic variant of the left renal vein, after ruling out other potential causes. In adults, computed tomography is the preferred method for investigating renal vein anatomy and the degree of compression, despite the significant radiation exposure involved. Most patients discover the anomaly incidentally, and it is important to be aware of it before undergoing surgery.

There have been only a few case reports of pNCS in the literature. The severity and stage of symptoms, as well as the patient's age, are crucial factors in determining the appropriate treatment to reduce hypertension in the LRV. For patients under 18 years old or those with mild hematuria, conservative treatment with at least 2 years of follow-up is recommended [7]. Several open surgery techniques have been described including left renal vein bypass, left renal vein transposition with Polytetrafluoroethylene (PTFE), Dacron or spiral saphenous vein graft, medial nephropexy with excision of renal varicosities, renal autotransplant, gonadocaval bypass and nephrectomy [8-9] with transposition of the left renal vein being the most common procedure. However, it may lead to thrombosis or anastomotic stenosis. One study reported 11 cases of renal vein for anterior NCS. Intraoperative LRV thrombectomy was performed in two patients before transposition. both cases resulted in thrombosis of the transposed renal vein at 4 weeks and 4 months postoperatively [10].

LRV compression between the aorta and vertebral body would result in a rise in the venous pressure in the distal venous portion. This would cause a decrease in flow and also temporary retention of venous blood. This will initiate a cascading effect resulting in venous congestion, and increased

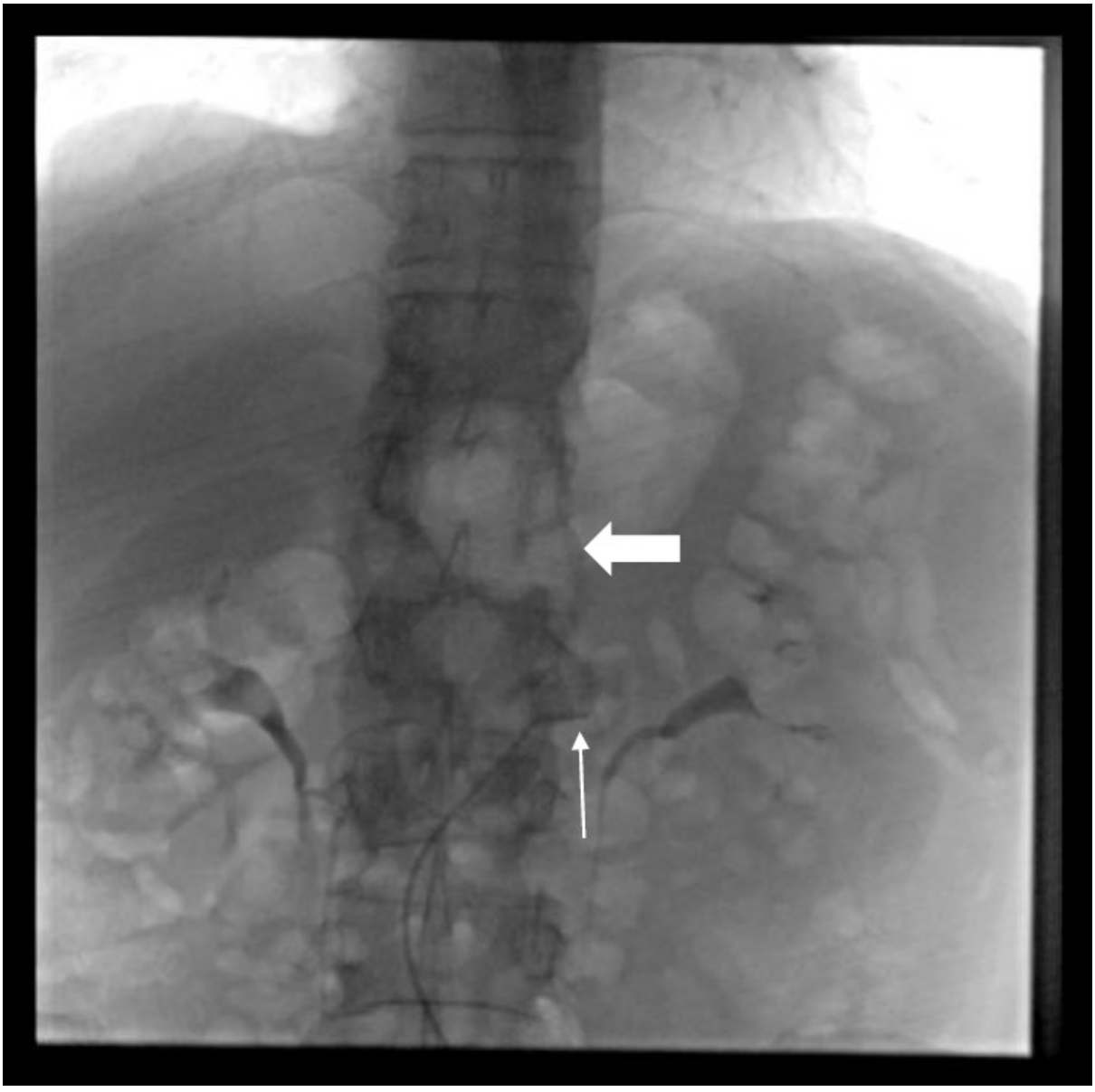

Figure 1: Ascending venography image highlighting the drainage via the left adrenal vein (indicated by block arrow) and distal left renal vein (indicated by the arrow). Moreover, the catheter's inability to pass to the renal hilum is notable, indicating compression of the left renal vein.

resistance to arterial blood flow in the kidney, leading to collateral blood flow. The collateral blood flow would act as a 'pop off' mechanism, resulting in a reduction in arterial resistance and an increase in renal perfusion. An upright erect posture further worsens the angulations, leading to an exacerbated hemodynamic response, which is responsible for the various clinical manifestations [3]. This compression translates into increased pressure on the left renal vein system, which causes varicosities in the renal pelvis and ureter, which can in turn communicate with the urinary tract and cause hematuria.

Ligation of the left renal vein during AAA surgery appears to be safe and feasible [11]. The initial drop in renal function improved over 2-6 weeks, and at each stage, the renal function was similar to patients in whom the left renal vein was not ligated also changes in Estimated Glomerular Filtration Rate (eGFR) are not different in the LRV ligation group at 90 days [6-15]. In a report of open repair of the concomitant abdominal aorta and right common iliac arteries aneurysm, an RLRV was encountered and ligated patients did not experi-

Figure 2: CT scan showing both kidneys with normal cortical parenchyma. The block arrow indicates the stump of the ligated left renal vein.

ence changes in renal function postoperatively [4]. Left renal vein ligation has also been suggested as a technique to improve portal venous flow and mitigate the risk of portal vein thrombosis after orthotopic liver transplant, with minimal and transient effects on renal function [12,13,14].

We propose that as LRV compression between the aorta and vertebral body is a positional phenomenon, retention of venous blood temporarily causes collateral flow and hilar venous varicosities. While the pressure rises varicosities in the renal pelvis open to the urinary tract and cause hematuria.

Ligation of the LRV avoids sudden, positional changes in this pressure, and after a while drainage via collateral flow becomes adequate and avoids the symptoms of venous hypertension.

Although this one case cannot be the guide for every posterior nutcracker syndrome treatment; but shows in situations the same as our case with no endovascular option and no potent gonadal vein in the re-implant and with a lot of side branches on the posterior aspect of RLRV or short RLRV ligation could be a safe and practical option avoiding bypass graft.

4. Appendix

4.1. Acknowledgment

None.

4.2. Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

4.3. Funding support

None.

4.4. Author's contributions

All the authors had the same contribution.

References

bakhtiar, hesam, shariati., Hamed, Shoorei. (2018). Retro Aortic Left Renal Vein: A Case Report and Brief Literature of Review. Open Access Journal, 8(4) doi: 10.19080/OAJS.2018.08.555744.

2. Nam, J. K., Park, S. W., Lee, S. D., & Chung, M. K. (2010). The clinical significance of a retroaortic left renal vein. Korean journal of urology, 51(4), 276–280. https://doi.org/10.4111/kju.2010.51.4.276.

3. Barad, B.B., Krishnamoorthy, S., & Kumaresan, N. (2021). Posterior nut-cracker syndrome: Urologist's perspective – A mini review. Biomedicine, 41(2), DOI: 10.51248/.v41i2.780.

4. Zubair, Khanzada., Greta, Saggu., S., C., Whitaker., Ganesh, Kuhan. (2016). Retro Aortic Left Renal Vein in Open Abdominal Aortic Aneurysm Surgery.. Jcpsp-journal of The College of Physicians and Surgeons Pakistan, 26(10):874-874.

5. Park, J. H., Lee, G. H., Lee, S. M., Eisenhut, M., Kronbichler, A., Lee, K. H., & Shin, J. I.. (2017, November 22). Posterior nutcracker syndrome – a systematic review. https://doi.org/10.1024/0301-1526/a000670.

6. Tapan, Mehta., Ryckie, G., Wade., J.M.F., Clarke. (2010). Is it Safe to Ligate the Left Renal Vein During Open Abdominal Aortic Aneurysm Repair. Annals of Vascular Surgery, 24(6):758-761. doi: 10.1016/J.AVSG.2010.02.008.

7. Jakub, Tomasz, Kramek., Zbigniew, Krasiński., Hubert, Stępak. (2021). Nutcracker syndrome - a mini review on current knowledge. Journal of the Medical Sciences, 90(3)

doi: 10.20883/MEDICAL.E527.
8. Rudloff U, Holmes RJ, Prem JT, Faust GR, Moldwin R, Siegel D. Mesoaortic compression of the left renal vein (nutcracker syndrome): case reports and review of the literature. Ann Vasc Surg 2006;20:120.
9. Tabiei, A., Cifuentes, S., & DeMartino, R. R. (2022). Spiral saphenous vein graft for left renal vein reconstruction after endovascular failure in the treatment of nutcracker syndrome. Journal of vascular surgery cases and innovative techniques, 9(1), 101085. https://doi.org/10.1016/j.jvscit.2022.101085.
10. Nanette R. Reed and Manju Kalra and Thomas C. Bower and Terri J. Vrtiska and Joseph J. Ricotta and Peter Gloviczki . "Left renal vein transposition for nutcracker syndrome". Journal of Vascular Surgery volume = 49, number = 2, pages = 386-394, year = 2009, issn = 0741-5214,doi = https://doi.org/10.1016/j.jvs.2008.09.051,
11. Kimihiro, Komori., Tadashi, Furuyama., Yoshihiko, Maehara. (2004). Renal Artery Clamping and Left Renal Vein Division during Abdominal Aortic Aneurysm Repair. European Journal of Vascular and Endovascular Surgery, 27(1):80-83. doi: 10.1016/S1078-5884(03)00408-8.
12. Michelle, C., Nguyen., Latifah, Sage, Silski., Musab, Alebrahim., Sylvester, M., Black., Elmahdi, A., Elkhammas., Kenneth, Washburn., Ashraf, El-Hinnawi. (2021). Left Renal Vein Ligation for Spontaneous Splenorenal Shunts During Deceased-Donor Orthotopic Liver Transplant Is Safe and Can Mitigate Complications from Portal Steal: A Case Series.Experimental and clinical transplantation : official journal of the Middle East Society for Organ Transplantation, 19(4):374-377. doi: 10.6002/ECT.2018.0096.
13. Martino RB, Júnior ER, Manuel V, Rocha-Santos V, D'Albuquerque LAC, Andraus W. A Case of Left Renal Vein Ligation in a Patient with Solitary Left Kidney Undergoing Liver Transplantation to Control Splenorenal Shunt and Improve Portal Venous Flow. Am J Case Rep. 2017;18:1086-1089. Published 2017 Oct 11. doi:10.12659/ajcr.905719.
14. Rui, Tang., Dongdong, Han., Modan, Li., Shan, Shen., Xin, Huang., Wenping, Zhao., Jiahong, Dong. (2017). Left renal vein ligation for large splenorenal shunt during liver transplantation.. Anz Journal of Surgery, 87(10):767-772. doi: 10.1111/ANS.14044.
15. Kamashi, Pandirajan., Emmanuel, Katsogridakis., Emmanuel, Katsogridakis., David, Sidloff., David, Sidloff., Robert, D., Sayers., Robert, D., Sayers., Matthew, J., Bown., Matthew, J., Bown., Athanasios, Saratzis., Athanasios, Saratzis. (2020). Effects of Left Renal Vein Ligation During Open Abdominal Aortic Aneurysm Repair on Renal Function.. European Journal of Vascular and Endovascular Surgery, 60(6):829-835. doi: 10.1016/J.EJVS.2020.08.003.