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Case Report

Nutcracker syndrome treated by intravascular ultrasound-guided endovascular stenting

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Nutcracker syndrome is an uncommon disease and should be easily overlooked. It is characterized by left renal vein entrapment between the abdominal aorta and the superior mesenteric artery. Hematuria and abdominal or flank pain are the most common symptoms. We present a 36-year-old female with chronic lower abdominal pain for 2 months. Computed tomography revealed extrinsic compression of the left renal vein by the aorta and superior mesenteric artery. Venography with pressure measurement showed a 6-mmHg pressure gradient between the left renal vein and the inferior vena cava. Nutcracker syndrome was confirmed. Endovascular stenting was performed under intravascular ultrasound guidance. After procedure, the pressure gradient between the left renal vein and inferior vena cava fell to 0-mmHg and her symptoms resolved. Endovascular stenting is becoming popular among intervention therapies as it is less invasive with a satisfactory result. Intravascular ultrasound can provide more precise lesion information and is helpful in stent sizing and identifying appropriate landing zone.

Keywords: Nutcracker syndrome; endovascular stenting; intravascular ultrasound

1. Introduction

Nutcracker syndrome (NCS) is a rare clinical condition referring to the compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the aorta.[1] The extrinsic compression of the LRV causes a variety of symptoms, ranging from microscopic hematuria to macroscopic hematuria, proteinuria, abdominal or flank pain, dyspareunia, dysmenorrhea, and severe pelvic congestion.[1, 2] NCS is an easily missed diagnosis due to its rarity and wide variability of symptomatic presentations.[3] Consequently, its true prevalence is unknown.[1] Diagnosis is generally made after the exclusion of other more common causes, and is confirmed by imaging results, including Doppler ultrasonography (DUS), computed tomography (CT), magnetic resonance imaging (MRI) and venography.[1] The standard treatment of NCS is controversial. Options range from conservative treatment to nephrectomy, with many open survey techniques and endovascular procedures in between, depending on the severity of the clinical symptoms.[1, 4] We present herein the case of a 36-year-old female treated with intravascular ultrasound-guided endovascular stenting with satisfactory results and no reoccurrence of symptoms during a five-year follow-up period.

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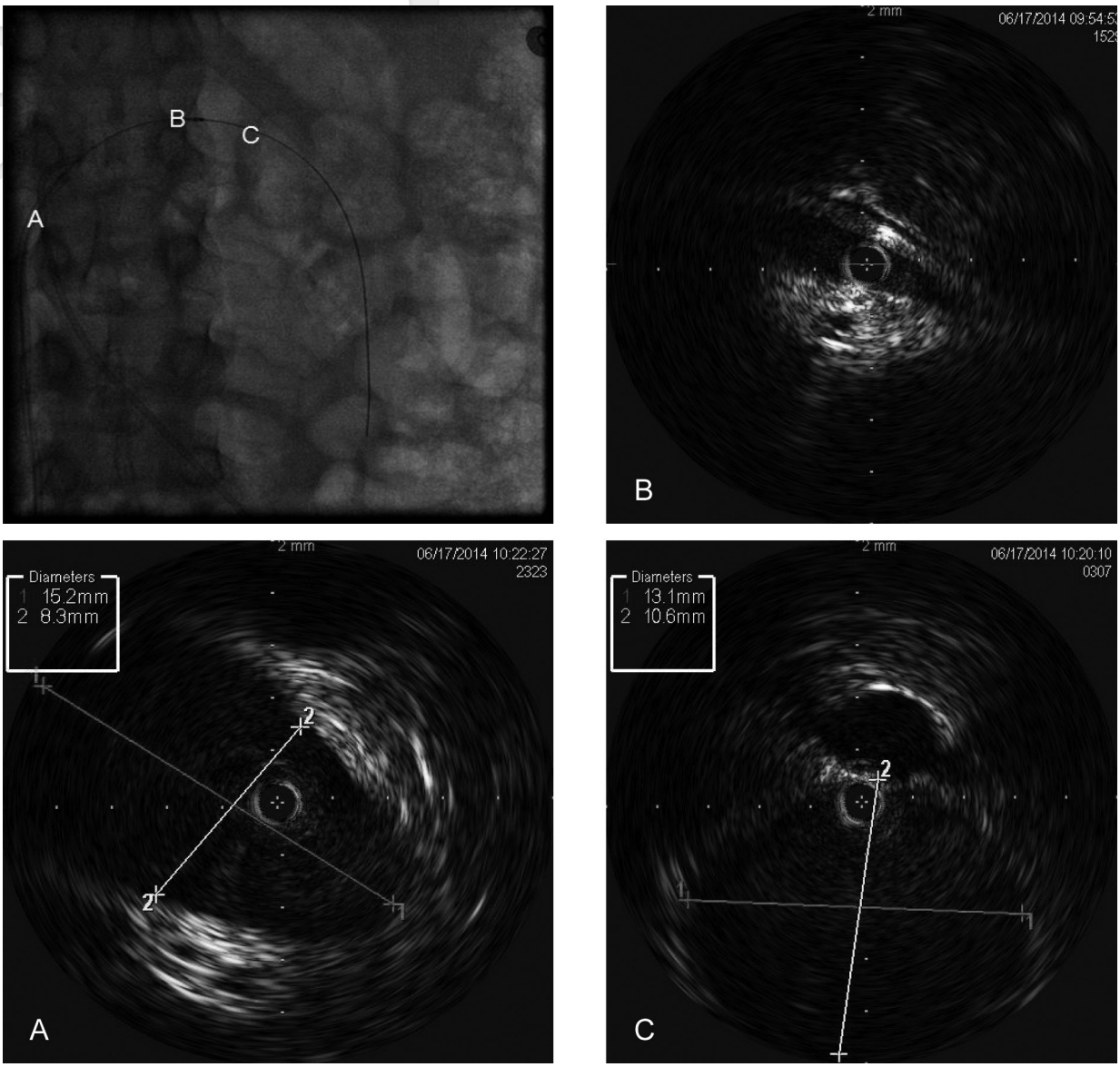

Fig. 1. Accurate IVUS detection of LRV diameter at different sites (A, B, C). (A) Proximal renal vein. (B) Narrowest site of renal vein. (C) Distal renal vein.

2. Case Report

A 36-year-old female with no remarkable history presented with lower abdominal fullness and intermittent cramping pain for 2 months. She had visited a local medical clinic for an initial evaluation, and upper gastrointestinal endoscopy had revealed a shallow peptic ulcer. Her abdominal pain persisted after medical therapy and hence she sought further assessment in the colorectal clinic of the hospital outpatient department. Other than intermittent abdominal pain, she reported no other gastrointestinal symptoms. A physical examination found no abdominal tenderness or flank knocking pain. Furthermore, her blood biochemistry data were all within normal limits and a urine analysis showed no hematuria or pyuria.

Abdomen and pelvic contrast-enhanced CT revealed no gastrointestinal abnormalities. However, extrinsic compression of the LRV by the aorta and SMA was noted, with proximal venous dilatation and an engorged ipsilateral gonadal vein. The diameter of

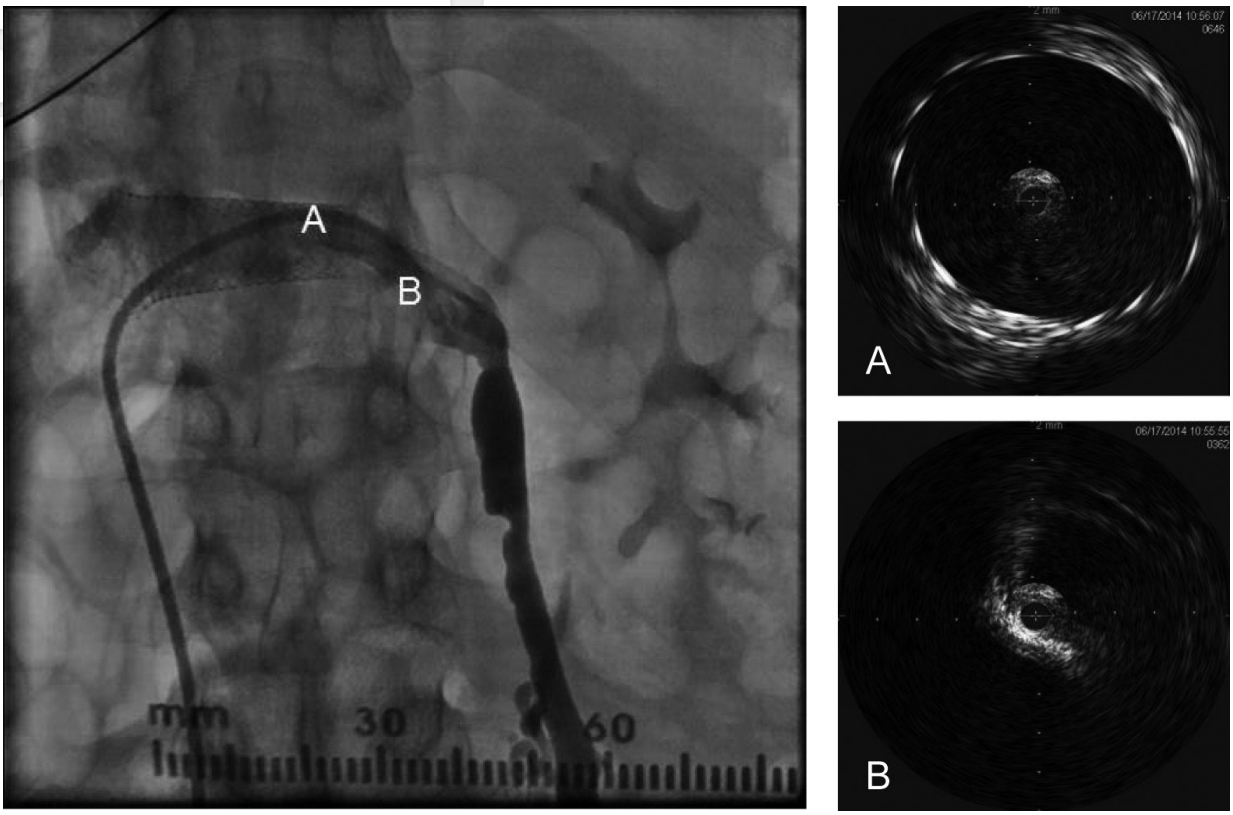

Fig. 2. Successful expansion of compression site following stent placement. IVUS images show increased vessel diameter with final pressure gradient of 0 mmHg. (A) Stent full expansion. (B) Enlarged diameter of narrowest segment.

the LRV at its most compressed level was 4.34 × 4.09 mm, while that at its most dilated level was 20.9 × 15.35 mm. In addition, the diameter of the ipsilateral proximal gonadal vein was 12 mm. NCS was thus highly suspected. The patient's abdominal pain persisted after 2 months of conservative treatment. Therefore, renal venography was performed and showed significant eccentric stenosis of the LRV with a 6-mmHg pressure gradient between the LRV and the inferior vena cava (IVC). NCS was confirmed. Following thorough consideration of the risks and benefits of different procedures, the patient chose endovascular treatment.

During the procedure, the LRV was cannulated with a 7F renal double curve (RDC) guiding catheter. A V-18™ guidewire (Boston Scientific Corp.) was advanced into the left gonadal vein and intravascular ultrasound (IVUS) (Volcano, Visions PV .018) was used to identify and size the stenotic segment of the LRV (Fig. 1). An Amplatz Super Stiff™ guidewire (Boston Scientific Corp.) was deployed for better support and a 14 × 40-mm Wallstent™ (Boston Scientific Corp.) was then deployed under IVUS guidance. The stent shortened after expansion and the stenotic segment was not fully covered. However, IVUS showed that the diameter of the LRV increased in the narrowest segment (Fig. 2). Furthermore, the pressure gradient between the LRV and the IVC fell to 0 mmHg after stent deployment. Therefore, an additional stent was not used.

After the procedure, the patient's abdominal pain resolved. Antithrombotic agents with warfarin were given for 2 months, followed by aspirin (100 mg/day) alone. Follow-up CT imaging 2 months later showed that there was no stent migration (Fig. 3) and the engorged left gonadal vein was the normal size (Fig. 4). Furthermore, at 5-year follow-up, the patient was still symptom free and chest X-rays showed no stent migration.

3. Discussion

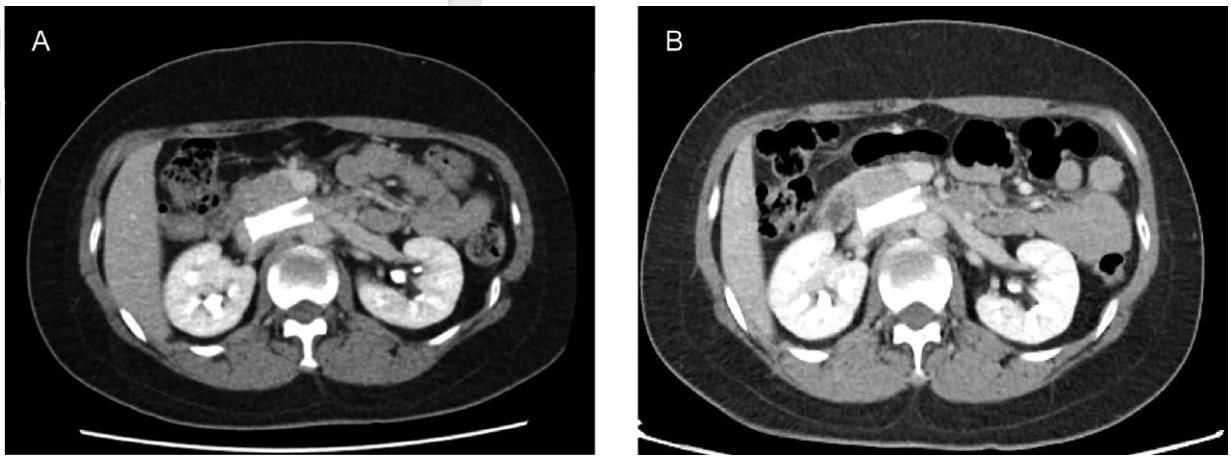

Fig. 3. CT images following stent placement (transverse view). (A) 1 day after stent placement. (B) 2 months after stent placement (no stent migration).

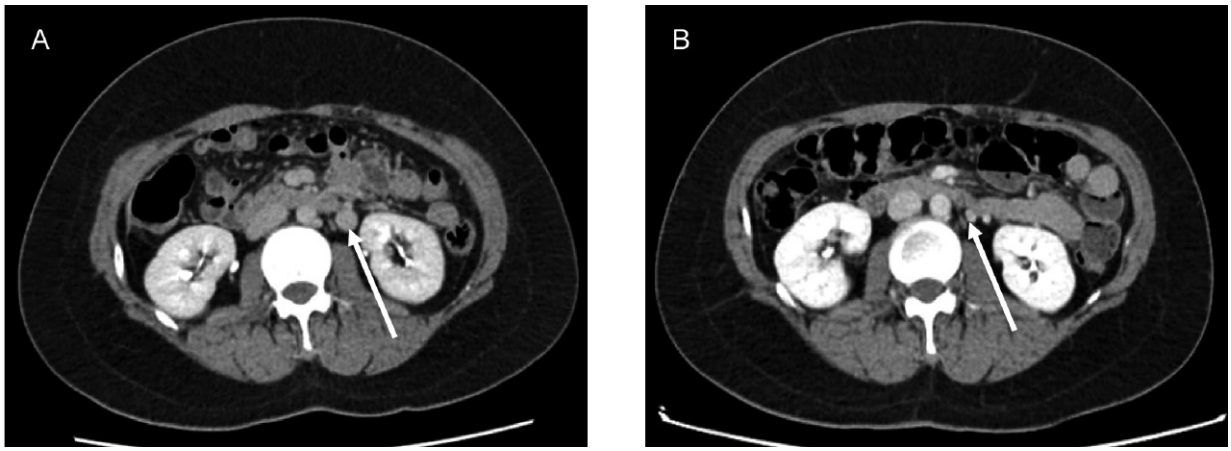

Fig. 4. CT image of left gonadal vein (arrowed). (A) Before stent placement. (B) 2 months after stent placement. The engorged gonadal vein returned to normal size after stent placement.

As described above, NCS is associated with a variety of symptoms. However, not all of these symptoms are specific to the disease. There is no consensus regarding the diagnostic criteria for NCS. Hence, diagnosis is generally made after the exclusion of other more common causes.[1] Real-time DUS is generally recommended as the first diagnostic test in patients with suspected NCS due to its high sensitivity (~78%) and specificity (as much as 100%).[5, 6] However, other studies have suggested that the renocaval pressure gradient cannot be properly predicted by the Doppler flow velocity since the flow rate depends on the degree of compensatory collateral vein formation.[3] According to a previous review article, the diagnostic criteria for NCS include:

a) A venous pressure gradient between the LRV and the IVC ≥ 3 mmHg;
b) A five-times increase in the maximum flow velocity in the LRV as it passes the SMA relative to that in the renal hilum;
c) Computed tomography angiography or magnetic resonance angiography showing an angle between the aorta and the SMA of less than 45o.[1]

The treatment options for NCS range from simple observation to nephrectomy, depending on the severity of the clinical symptoms.[3, 4] However, interventions should only be considered when the symptoms are severe or persistent, including severe, unrelenting pain, severe hematuria, renal insufficiency, and failure to respond to conservative treatment after 24 months.[3] In a recent study, Hangge et al.[7] reported that a higher compression of the LRV is correlated

to more hematuria, abdominal pain and more proteinuria. In the case of the patient considered herein, the abdominal pain persisted after 2 months of conservative treatment and CT imaging revealed a high degree of LRV compression with an engorged ipsilateral gonadal vein. Consequently, invasive diagnostic venography was performed.

Intervention therapies for NCS include open surgery and endovascular stenting. Open surgery, specifically LRV transposition, is the mainstay of treatment. However, it has several key disadvantages, including a prolonged period of renal congestion, the need for additional anastomosis, and extensive dissection.[1] By contrast, endovascular stenting is less invasive and generally achieves satisfactory results. For example, a previous study compared 15 patients treated using endovascular stenting with 5 patients treated using open surgery, and found that all of the patients treated with stents were asymptomatic during the follow-up period with the exception of a single case, in which the stent migrated to the right atrium and required surgical intervention to effect its removal.[8] Another study on a cohort of 61 patients with a mean follow-up period of 64 months also showed that endovascular stenting for LRV was safe and effective. [9] However, a later study of the same cohort reported a 6.7% stent migration rate attributed to inappropriate LRV diameter measurement or stent sizing during the original intervention.[10] Therefore, stent migration may not in fact be as rare as originally thought. The authors thus suggested that the preoperative anatomic parameters of the LRV must be more accurately measured. IVUS is beneficial in this regard due to the ability it provides to identify the true compressed lesion and vessel size. For example, in a recent published cohort of 18 patients, IVUS was used in 11 patients (61%), and no stent migration occurred.

In the present case, endovascular stenting was chosen as the intervention therapy after extensive discussions between the patient and the surgeon. In performing the intervention, difficulties were encountered in accurately identifying the stenotic segment by venography since the stenosis was eccentric. Accordingly, IVUS was used to recognize the accurate lesion location and vessel size. Based on the IVUS observations, a 14 x 40 mm Wallstent® was chosen for implantation. The stent shortened after expansion and did not position ideally. Thus, the implantation of a second stent was considered. However, the pressure gradient between the LRV and the IVC resolved and IVUS showed a reduced extrinsic compression of the LRV. Consequently, the second stent was considered to be unnecessary and was therefore not implanted. CT imaging performed 2 months after the procedure revealed that no stent migration occurred, and the engorgement of the left gonadal vein was significantly reduced. In other words, the stent functioned well despite its shortening in the implantation procedure. Furthermore, the patient was still symptom-free 5 years after the procedure. According to a previous study, WallStent™ is the most frequently used stent, but may exhibit more than 30% shrinkage in length on implantation.[9] Consequently, Wang et al recommended that stents with a length of 6 or 8 centimeters should be employed.[12] In addition, to avoid migration of the stent, its diameter should be specified around 20% larger than the venous diameter at the renal hilum. Thus, the basic size is considered to be 14 mm in diameter for patients of the Eastern population.[9]

After stent placement, antithrombotic agents are necessary for preventing thrombosis. However, no standard antithrombotic therapy exists. Thus, the use of anticoagulant for 3 days to 3 months, followed by dual antiplatelet therapy (aspirin and clopidogrel) for 1 to 3 months, and then aspirin alone indefinitely is often suggested.[1, 3, 11]

In conclusion, endovascular stenting is a safe and effective therapy for NCS, and is gaining popularity among intervention therapies. However, to achieve proper stent apposition and avoid stent migration, the preoperative anatomic parameters of the LRV must be accurately measured and venography with corresponding IVUS performed to obtain more precise lesion information. Furthermore, a longer stent (6 to 8 centimeters) is recommended to compensate for stent shortening during implantation.

References

1. de Macedo, G.L., M.A. Dos Santos, A.B. Sarris, and R.Z. Gomes, *Diagnosis and treatment of the Nutcracker syndrome: a review of the last 10 years.*

J Vasc Bras, 2018. **17**(3): p. 220-228.
2. Nickavar, A., *Nutcracker syndrome; a rare cause of hematuria.* J Nephropathol, 2016. **5**(4): p. 144-145.
3. Kurklinsky, A.K. and T.W. Rooke, *Nutcracker phenomenon and nutcracker syndrome.* Mayo Clin Proc, 2010. **85**(6): p. 552-9.
4. He, Y., Z. Wu, S. Chen, L. Tian, D. Li, M. Li, W. Jin, and H. Zhang, *Nutcracker syndrome--how well do we know it?* Urology, 2014. **83**(1): p. 12-7.
5. Quevedo, H.C., S.A. Arain, and N. Abi Rafeh, *Systematic review of endovascular therapy for nutcracker syndrome and case presentation.* Cardiovasc Revasc Med, 2014. **15**(5): p. 305-7.
6. Takebayashi, S., T. Ueki, N. Ikeda, and A. Fujikawa, *Diagnosis of the nutcracker syndrome with color Doppler sonography: correlation with flow patterns on retrograde left renal venography.* AJR Am J Roentgenol, 1999. **172**: p. 39-43.
7. Hangge, P.T., N. Gupta, A. Khurana, K.B. Quencer, H. Albadawi, S.J. Alzubaidi, M.G. Knuttinen, S.G. Naidu, and R. Oklu, *Degree of Left Renal Vein Compression Predicts Nutcracker Syndrome.* J Clin Med, 2018. **7**(5).
8. Zhang, H., M. Li, W. Jin, P. San, P. Xu, and S. Pan, *The left renal entrapment syndrome: diagnosis and treatment.* Ann Vasc Surg, 2007. **21**(2): p. 198-203.
9. Chen, S., H. Zhang, H. Shi, L. Tian, W. Jin, and M. Li, *Endovascular Stenting for Treatment of Nutcracker Syndrome: Report of 61 Cases With Long-Term Followup.* Journal of Urology, 2011. **186**(2): p. 570-575.
10. Wu, Z., X. Zheng, Y. He, X. Fang, D. Li, L. Tian, and H. Zhang, *Stent migration after endovascular stenting in patients with nutcracker syndrome.* J Vasc Surg Venous Lymphat Disord, 2016. **4**(2): p. 193-9.
11. Avgerinos, E.D., Z. Saadeddin, R. Humar, K. Salem, M. Singh, E. Hager, M. Makaroun, and R.A. Chaer, *Outcomes of left renal vein stenting in patients with nutcracker syndrome.* Journal of Vascular Surgery: Venous and Lymphatic Disorders, 2019. **7**(6): p. 853-859.
12. Wang, X., Y. Zhang, C. Li, and H. Zhang, *Results of endovascular treatment for patients with nutcracker syndrome.* J Vasc Surg, 2012. **56**(1): p. 142-8.