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ORIGINAL ARTICLE

Endovascular treatment of the nutcracker syndrome: report of two cases

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ABSTRACT

Background: Nutcracker syndrome (NCS), also known as left renal vein (LRV) entrapment causing dilatation of gonadal vein. Endovascular treatment has evolved as a potential minimally invasive therapy for relief symptom.
Objective: To present the early result of endovascular treatment in patient with NCS.
Material and Methods: We report two cases of endovascular treatment for NCS with different clinical manifestation. Both of the patients was diagnosed by CT scan. Endovascular treatment was choosing for both cases and started by measure reno-caval pressure gradient. We used 2 different options, such as ovarian vein embolization only and left renal vein stent only.
Results: The treatment of NCS remains a controversy. Endovascular treatment is alternative treatment and less invasive option but had diversity of decision making. We offered successful treatment options, which relieve symptoms of our patients.
Conclusions: Endovascular approach is an alternative treatment of NCS. There were more evidence of this option and revealed good results. Based on our experience, LRV stenting or ovarian vein embolization were good result for NCS. However, further follow-up will be request for determine durability of this approach.

Keywords : nutcracker syndrome, left renal vein compression, pelvic congestion syndrome, endovascular treatment, Thailand

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Introduction

Nutcracker syndrome (NCS), also known as left renal vein (LRV) entrapment causing dilatation of gonadal vein, was first described in 1950.[1] The clinical signs and symptoms were hematuria, proteinuria, flank pain, pelvic congestion in females, and varicocele in male patients.[2] Endovascular treatment has evolved as a potential minimally invasive therapy for relief symptom. We report two cases of successful endovascular treatment for NCS.

Case presentation 1

A healthy 31-year-old female presented with severe left pelvic pain and flank pain for 2 hours and multiple episodes of left lower quadrant pain for 1 year. The patient went to gynecological clinic and transvaginal ultrasound found cystic lesion at left side. The doctor scheduled for MRI for further investigation. The patient denied postprandial pain, nausea or vomiting. There was no history of urinary symptom. The patients had body mass index (20 kg/m^2). Physical examination revealed marked tenderness of left lower quadrant and no rebound tenderness. There was no palpable mass. Laboratory findings including hematological investigation, amylase level, lipase level, liver function test and urinalysis were normal. Computed Tomography (CT) of abdomen revealed compression of the left renal vein between superior mesenteric artery and aorta with mildly dilated left ovarian vein and engorgement of left-sided uterine plexus, aortomesenteric angle 15 degree and LRV hilar aortomesenteric cutoff ratio 5.87 as Figure 1

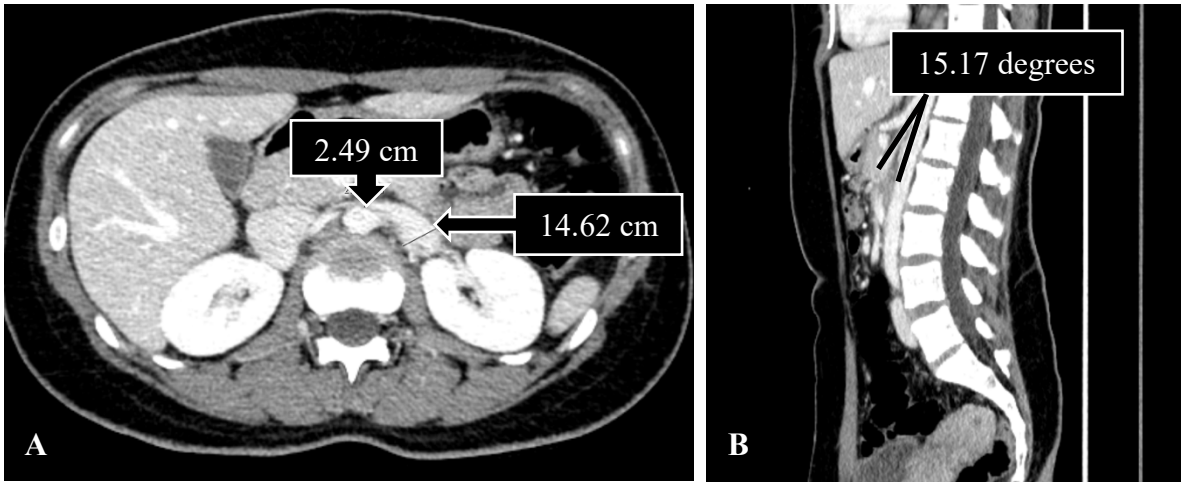


Figure 1: CT scan of abdomen axial view revealed compression of the left renal vein between superior mesenteric artery and aorta (A) and sagittal view revealed 15 degrees of aortomesenteric angle (B)

Initial presentation and physical examination finding raised the possibility of gynecological disease or renal disease. However, following CT finding were most likely nutcracker syndrome.



The patients receive analgesic drug and pain resolved. Treatment modality was discussed with this patient, the patient decided for endovascular treatment due to suffering recurrent pelvic pain after diagnosis. We scheduled for endovascular treatment. Right femoral vein was accessed under ultrasound guidance. After 6Fr sheath was inserted, Hydrophilic wire 0.0035" (GLIDECATH® TERUMO) with selective catheter (IMPRESS® Cobra 2 catheter, Merit medical) was used for passing wire to left renal vein and renocaval gradient was evaluated. Pressure gradient between IVC and left renal vein was 2 mmHg. Ovarian vein embolization was done by detachable coil (Concerto™ PGLA helical, Medtronic) size 12 mm and 10 mm. After procedure, the patients had no postoperative complication. At 3-month follow-up, the patient had resolution of symptom.

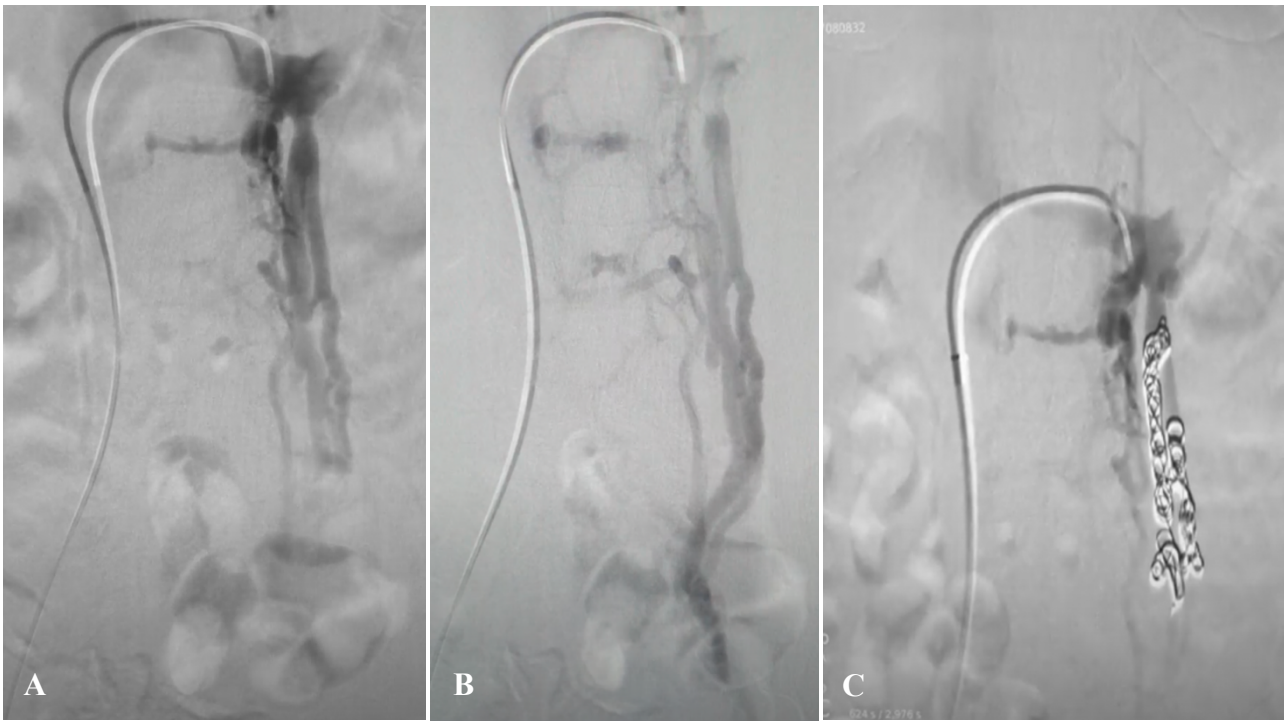


Figure 2: Intraoperative venogram demonstrated selective venography of left renal vein (A), reflux of left ovarian vein (B) and post ovarian vein embolization by detachable coil (C)

Case presentation 2

A healthy 22-year-old male presented with hematuria for 2 weeks and urinary incontinence. The patients had body mass index (24 kg/m^2). Physical examination reveals no palpable mass or tenderness of abdomen. Laboratory finding were all within normal limit while urinalysis showed macroscopic hematuria without evidence of urinary tract infection. Urologic evaluation of this symptom led to CT scan of abdomen. The CT scan

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revealed compression of the left renal vein between superior mesenteric artery and aorta with mildly dilated left testicular vein and engorgement of left-sided pampiniform venous plexus, Aortomesenteric angle 28.3 degree and LRV hilar aortomesenteric cutoff ratio 2.87. The patient was sent to vascular clinic for evaluation. Most likely cause of gross hematuria of this patient was nutcracker syndrome. Treatment modality was discussed with this patient and the patient decided for endovascular treatment. We scheduled for endovascular treatment. Right femoral vein was accessed under ultrasound guidance. After 6Fr sheath was inserted, Hydrophilic wire 0.0035"(GLIDECATH® TERUMO) with selective catheter (IMPRESS® Cobra 2 catheter, Merit medical) was used for passing wire to left renal vein and renocaval gradient was evaluated. Pressure gradient between IVC and left renal vein was 5 mmHg. We exchange to Introducer sheath 12 Fr with long sheath 9Fr and administration of systemic heparinization. A 14x60 mm bare self-expandable stent (VICI venous stent™, Boston) was deployed at left renal vein. Complete venogram show proper position of stent. After procedure, the patient was started on low molecular weight heparin (1mg/kg for 3 days) and clopidogrel (75mg) for 3 months. At 6-month follow-up, the patient had not recurrent gross hematuria.

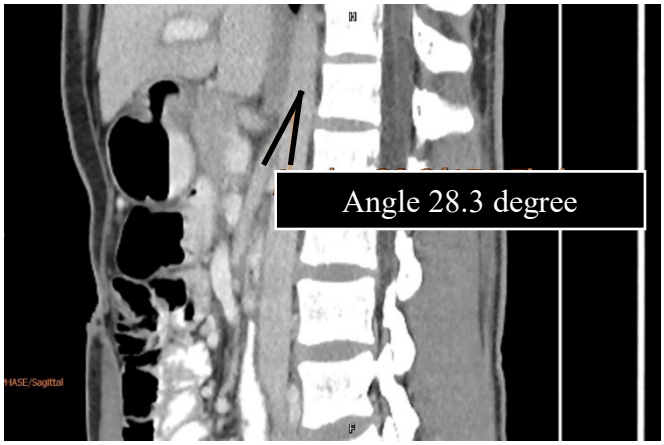


Figure 3: CT scan of abdomen sagittal view revealed 28.3 degree of aortomesenteric angle

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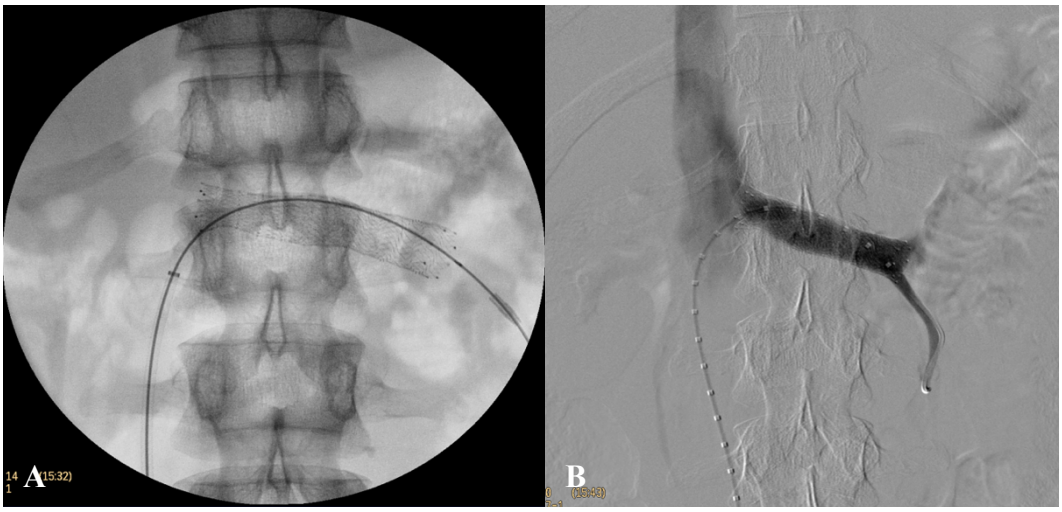


Figure 4: Fluoroscopic view demonstrated LRV stent placement by 14x60 mm bare self-expandable stent (A) and intraoperative venogram after LRV stent placement (B)

Discussion

NCS is a rare disease caused by compression of LRV by superior mesenteric artery (SMA). The prevalence was peaking in second or third decades and higher in females.[2,3] The most common clinical presentation of NCS include pelvic pain, flank pain, hematuria and gonadal varices (varicocele or ovarian vein syndrome),[2,3] caused by pelvic and renal congestion. The diagnosis of NCS is confirmed on imaging results, including doppler ultrasound, CT, venogram, intravascular ultrasound.[4,5,6] Axial imaging should be required, they demonstrated anatomy and surrounding structure. According to one study,[6] Beak sign (narrowing of LRV at aortomesenteric portion) demonstrated the highest diagnosis accuracy. Another diagnostic cutoff used an LRV hilar aortomesenteric cutoff ratio of 4.9 and a less than 41-degree angle between the SMA and aorta.[7] However, the gold standard in diagnosis of NCS is venogram and measurement of the venous pressure gradient. Bernert et al demonstrated that in the majority of healthy patients to renocaval pressure gradient was 0-1 mmHg,[8] so validating the use of a more than 3 mmHg gradient to diagnose NCS. Although the renocaval pressure gradients even below 3 mmHg can be explained by efforts to compensate for the renal venous hypertension by the formation of collateral formation.[9,10] Furthermore, it has been shown that IVUS has a higher specificity of 90% compared with 62% with venogram.[11,12]



The treatment of NCS remains a controversy especially the indication of treatment and treatment modality. Mild hematuria or mild symptoms was recommended conservative treatment.[11] The development of venous collateral help resolved LRV hypertension.[5] Furthermore, Weight gain resulting reduction of LRV tension, which resolved symptom in 30% of patients.[12] A conservative approach is preferred in patients < 18 years because of an increase in intraabdominal fibrous tissue at SMA origin resulting released LRV entrapment and resolution of symptoms.[11] However, Patients with recurrent gross hematuria, severe symptoms of pain, anemia, impairment renal function, varicocele formation and failure conservative treatment should be considered for surgery.[9]

There were several modalities for treatment such as open surgery, laparoscopic surgery and endovascular surgery. Endovascular stent placement is an alternative treatment. The largest case series with 61 patients undergoing LRV stenting with median follow up of 66 months reported excellent improvement for 59 patients[13].Complication included maldeployment of stent and stent migration.[13] Self-expanding stent (Wallstent) are the most frequently used and recommend device with 14 or 16 mm in diameter by 60 mm in length because of oversize 20% of renal hilum venous diameter for avoid migration. The stent migration rate was 6.7% and the others complication were in-stent thrombosis and fracture.[14,15]

Another endovascular option was left gonadal vein (LGV) embolization, which had been indicated in patients with varicocele in men and pelvic congestion syndrome in woman. There could relief symptoms in 56-98% of patients.[16] LGV embolization has become gold standard for treatment pelvic congestion syndrome.[17] Embolic agent had been used such as sclerosing foam, glue, vascular plug and coils.[17]Coil embolizaton of ovarian vein demonstrated symptom improvement in 56-98 of patient.[16]

In this case report, both of patients had in range of most common age group. First case presented with pelvic pain with flank pain. Another case presented with gross hematuria. Both of them meet CT criteria of NCS. We offered endovascular treatment of both. We started with venogram and measured renocaval gradient before decision making. In first case had less than 3 mmHg pressure gradient and we decided for ovarian vein embolization for improving symptoms. Second case had more than 3 mmHg pressure gradient and we decided for LRV stent placement. We used self-expandable stent and sizing as the previous studies recommendation. The technical points for precise deployment were using long sheath to supporting and stiff wire advance distally to gonadal vein to facilitate stent tracking.



Conclusion

Endovascular approach is an alternative treatment of NCS. There were more evidence of this option and revealed good results. Based on our experience, LRV stenting or ovarian vein embolization were good result for NCS. However, further follow-up will be request for determine durability of this approach.

Consent informed

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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The clinical case is exempt from ethical approval.

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Guarantor

Nawaphan Taengsakul, MD

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Not applicable.

Credit authorship contribution statement

Nawaphan Taengsakul: performed the surgery, Investigation, Writing - Original draft, Writing - Review & editing; critical revision

Thatchawit Urasuk: performed the surgery, Investigation.

Declaration of competing interest

The authors report no declarations of interest.

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