

Aortomegaly Causing Nutcracker Syndrome and Pelvic Congestion Syndrome

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Abstract

Keywords

- ▶ pelvic congestion syndrome
- ▶ nutcracker syndrome
- ▶ ovarian vein embolization
- ▶ renal vein stenting
- ▶ aortomegaly

The authors present an uncommon cause of pelvic congestion syndrome (PCS) secondary to anterior nutcracker syndrome, which was caused by aortomegaly. Positional flank and pelvic pain was the only presenting feature with no renal dysfunction. Early and significant decompression of left renal vein (LRV) via left ovarian vein resulted in preserved renal function with symptomatic pelvic varices. Endovascular management by left ovarian vein coiling and LRV stenting was done. They briefly review the etiopathology, imaging, treatment rationale, and management options for nutcracker and PCS.

Introduction

Pelvic congestion syndrome (PCS) presents as noncyclical chronic pelvic pain that lasts over 6 months' duration.¹ PCS is characterized by positional pelvic pain that is worse in the upright position and is associated with pelvic and vulvar varicosities as well as symptoms of dyspareunia and postcoital pain.

Nutcracker syndrome (NCS) due to compression of the left renal vein (LRV) may occasionally manifest as PCS owing to decompression of LRV via left ovarian vein. Retrograde flow of blood in the gonadal and internal iliac veins results in venous varicosities producing symptoms of PCS.²

We present an unusual case of PCS secondary to an uncommon cause of anterior NCS in which a diffusely enlarged aorta (aortomegaly) was causing compression of the LRV between the aorta and the superior mesenteric artery (SMA). Endovascular treatment was done by left ovarian vein coil embolization and LRV stent placement.

Case Report

A 65-year-old postmenopausal female presented with pain in lower abdomen and flank for 8 months, which was aggravated in upright position and was relieved in supine position. Past medical history was significant for chronic obstructive airway disease, diabetes, and recent myocardial infarction.

Her gynecologic and surgical evaluation failed to reveal anything specific. Blood investigations and stool and urine examinations were inconclusive.

Contrast-enhanced computed tomography (CT) scan of the abdomen revealed diffuse enlargement of several aortic segments greater than 50% suggesting aortomegaly (▶Fig. 1A). The dilated aorta was seen to displace and cause compression upon the LRV as it traversed between the SMA and aorta (▶Fig. 1B) with resultant dilated left ovarian vein and pelvic varices.

Endovascular management in the form of left ovarian vein coiling and LRV stenting was contemplated in view of her symptomatology and high surgical risk factors. Initial left renal arterial angiogram in the venous phase showed compressed LRV with reflux of contrast into the left ovarian vein and pelvic varices (▶Fig. 2A). LRV was cannulated via right transfemoral venous access using 5F Headhunter H1 catheter (Cordis). Selective left ovarian venogram showed dilated pelvic varices, which drained through right ovarian vein into inferior vena cava (IVC) (▶Fig. 2B). Venogram after coil embolization of the left ovarian vein showed obliteration of the left ovarian vein with non-opacification of the pelvic varices (▶Fig. 3A). A self-expanding nitinol stent 14 × 60 mm (Smart Control, Cordis) was deployed across the stenotic segment of LRV via right transjugular venous access. Poststenting venogram showed good antegrade flow across the stent into

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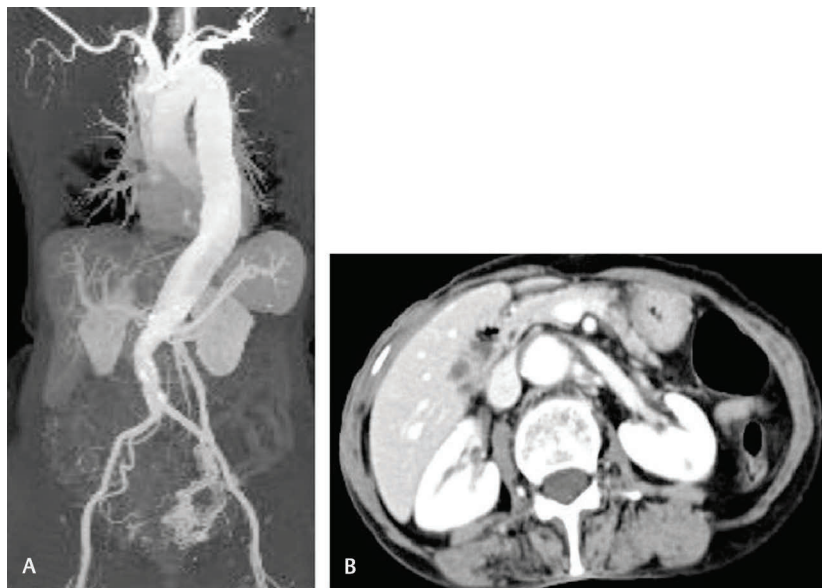


Fig. 1 (A) CT angiogram with coronal MIP reformation showing aortomegaly with dilated left ovarian vein and left pelvic varices. (B) Axial CT angiogram shows the left renal vein compressed between aorta and superior mesenteric artery as it drains into IVC. CT, computed tomography; IVC, inferior vena cava; MIP, maximum intensity projection.

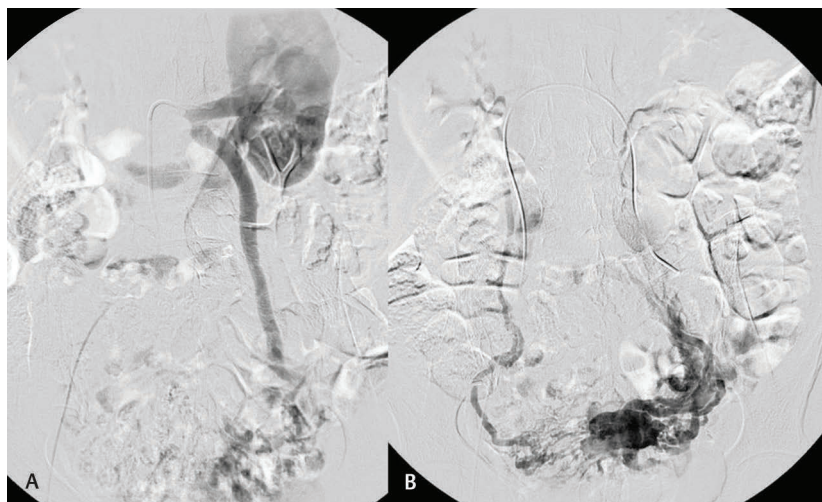


Fig. 2 (A) Selective left renal artery angiogram in venous phase shows reflux of contrast into dilated left ovarian vein and pelvic varices. Note near complete absence of flow across compressed portion of left renal vein into IVC. (B) Selective left ovarian venogram shows dilated left ovarian vein with pelvic varices and reflux of contrast onto the contralateral side.

the IVC with no further reflux (► **Fig. 3B**). At 1-month follow up, clinically, the patient had symptomatic relief by 80%. Follow-up angiography showed good flow across the stent with no stent migration or in-stent stenosis.

Discussion

Non-cyclical chronic pelvic pain due to PCS remains a diagnosis of exclusion; thus, all other potential causes of pain must be evaluated and excluded beforehand. Chronic pelvic pain differentials include a long list viz-a-viz. bowel pathologies, cancer/metastases, pelvic inflammatory disorder, porphyria, urologic, orthopedic and neurologic pathologies, endometriosis, fibroids, fibromyalgia, ovarian cyst, uterine prolapse, and PCS.¹

Retrograde flow of blood in the gonadal and internal iliac veins results in increased venous pressure causing venous varicosities, decreased clearance of blood, local hypoxia with subsequent acidemia, and compromised microcirculatory function producing symptoms of PCS.²

Pelvic congestion syndrome could be primary or secondary. The ovarian veins generally contain valves permitting unidirectional flow, however, incompetent or absent valves result in reflux. Valves maybe absent on the right and the left side in 6 and 15%, respectively, and are incompetent in 35 to 41% cases when they are present.³

Nutcracker phenomenon is the compression of LRV and impaired blood outflow with its clinical equivalent being NCS characterized by a complex of symptoms. *Anterior nutcracker*

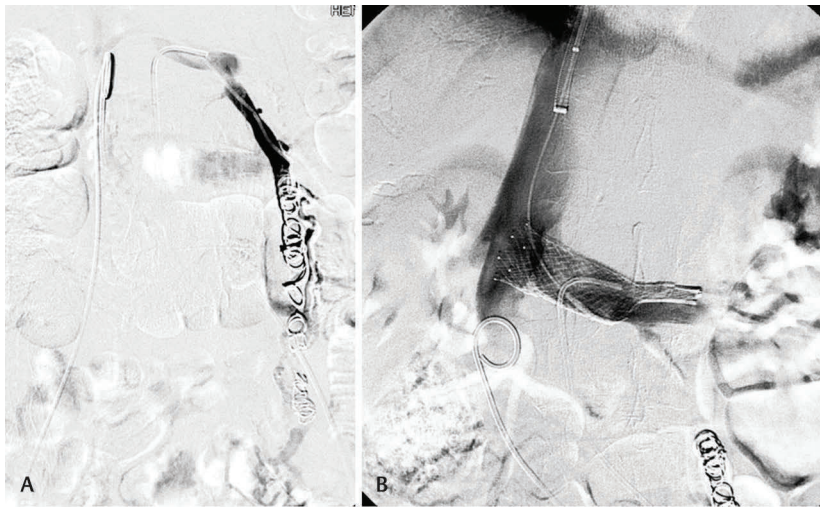


Fig. 3 (A) Selective left ovarian venogram after coiling shows occlusion with no further opacification of the pelvic varices. (B) Venogram after deploying self-expanding nitinol stent across compressed left renal vein shows good caliber and no extrinsic compression. There is preferential free flow of contrast into the IVC and no reflux into the left ovarian vein.

implies compression of the LRV between the aorta and the SMA. The retroaortic or circumaortic renal vein may be compressed between the aorta and the vertebral body, which is termed as *posterior nutcracker*. 'Overflow' situation can also appear in portal hypertension where splenorenal shunts may develop causing increased LRV flow, which decompresses via left ovarian vein. May–Thurner syndrome may also result in pelvic varices secondary to iliofemoral deep vein thrombosis.³ Hematuria and orthostatic proteinuria are among the most common reported symptoms in NCS. Decompression of LRV via left ovarian vein leads to pelvic varices and PCS. d'Archaubeau et al reported an 83% incidence of the NCS in patients referred due to symptoms of pelvic venous congestion.⁴

A majority of pelvic varices go undetected by laparoscopy owing to technical limitations, which include compression of these varices from peritoneal carbon dioxide insufflation and Trendelenburg position causing decompression. Despite advances in cross-sectional imaging, catheter-directed venography still remains the gold standard. Cross-sectional imaging may be employed to exclude a concurrent pelvic pathologic process.

Venographic evaluation includes assessment of the IVC, LRV, gonadal veins, and common and internal iliac veins. Proposed diagnostic criteria on venography are as follows (→ Fig. 2):

- Valvular incompetence with reflux into the ovarian vein
- Ectatic veins—at least 5 mm diameter of the ovarian, uterine, and utero-ovarian arcade veins
- Reflux of contrast medium onto contralateral side
- Vulvar or thigh varices opacification
- Stagnation of contrast medium in pelvic veins.

Extrinsic impression, compression or complete cut-off of the LRV can be seen in NCS. Renocaval pressure gradients may be elevated (normal 0–1 mm Hg). Strict measurements though should not preclude treatment if the overall clinical picture favors PCS.⁵

Two important components need to be assessed and addressed when planning treatment in cases of NCS, i.e., LRV hypertension and pelvic venous reflux. The correlation between imaging evidence of LRV compression and clinical symptoms is strenuous; thus, interventions should be proposed only when symptoms are severe or persistent.⁶

Medical, surgical, and endovascular means have all been previously employed in managing PCS. Medical management comprises hormone analogues and analgesics, surgical treatment involves ligation of ovarian veins, hysterectomy with or without bilateral salpingo-oophorectomy, and endovascular treatment involves transcatheter embolization.⁵

Symptoms of PCS have to be addressed by eliminating reflux pathways that cause abnormal hydrostatic pressure on the ovary and adnexa. Endovascular therapy involves occluding the major refluxing channels and any branch vessels that may cause recurrence via collateral flow. The ovarian veins should be embolized as close to ovary as possible, as smaller branch vessels can be numerous and difficult to visualize. Coils should be oversized to optimize stability and prevent migration.

Few authors advocate evaluating and embolizing branches of internal iliac veins if found communicating with pelvic varices or with ovarian veins.⁷ Coaxial temporary balloon occlusion maybe employed to control flow during embolization.

Coil embolization of ovarian veins in patients with PCS provides symptomatic improvement in 56 to 98%. When PCS is secondary to NCS, occlusion of the collateral veins may increase renocaval pressure gradients; therefore, measures to relieve the renocaval pressure gradients should also be undertaken.⁶

Management of NCS could be endovascular (intravascular balloon angioplasty and stenting using self-expanding stent, balloon expandable stent, or self-expanding covered stents) or surgical (medial nephropexy with excision of renal varicosities, LRV bypass, LRV transposition with or without Dacron wedge insertion between SMA and aorta,

SMA transposition, renal-to-IVC shunt, renal autotransplant, gonadocaval bypass, external stenting, and nephrectomy).

The advantage of endovascular stenting over surgery is its relatively less invasive nature, which is preferred especially in patients with multiple co-morbidities, and it allows shorter hospitalization.

Complications could be those related to any other endovascular procedure such as puncture site hematoma, pseudoaneurysm, contrast reactions, etc. Risk of coil and/or glue migration during ovarian vein and internal iliac vein is pertinent and measures such as providing additional support to delivery catheter, stable positioning of the delivery catheter, selection of appropriately sized coils should be taken to avoid this. Stent migration during LRV stenting has also been previously reported.³

The technical success of this procedure is nearly 100% with clinical success being quite high. In a single-center prospective study recruiting 202 patients with PCS that had been followed up over 5-year period, Laborda et al reported clinical success in 93.85% cases with complete resolution of symptoms in 60 (33.52%) patients. Overall, there was a statistically highly significant reduction in pain score at the end of follow-up.⁸

Wang et al reported 30 patients with NCS who underwent endovascular treatment. All were treated with one self-expanding metallic stent in the LRV, and three patients with severe left-sided varicoceles received left gonadal vein embolization. Two stents migrated into IVC at 1-year follow-up, but with eventual uneventful follow-up (49 and 56 months, respectively). The clinical symptoms almost disappeared at 3 months following treatment. All stents were patent at the duplex scan examination, and patients were symptom free at the end of the follow-up (median 36 months).⁹

Antiplatelet therapy has been advocated until the time stent endothelialization occurs. Aspirin and Clopidogrel were initiated in our patient for a period of 6 months.

To our knowledge, so far no case has been reported with aortomegaly resulting in anterior NCS and eventual PCS that was managed by endovascular approach. However, a similar case with surgical management was reported recently by Lozuk et al,¹⁰ where an abdominal aortic aneurysm was resulting in NCS and engorged left ovarian vein. This was treated surgically by aneurysm resection, followed by left ovarian vein ligation and left adnexectomy.

Conclusion

Exact prevalence of NCS is unknown in view of variable symptoms, but can be seen in any age group with most symptomatic patients being in second or third decade and a second peak in middle-aged females. Hematuria, pain, pelvic varicosities, and varicoceles are the most common clinical signs that should raise suspicion for the diagnosis. Treatment for this should be directed depending on the clinical severity

of the condition rather than mere imaging abnormality. The rates of complication and symptomatic recurrence after endovascular therapy appear rather low; hence, it should be strongly considered over more invasive surgical procedures.

Note

Informed consent was obtained from the patient included in the study. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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Prior Publication

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Conflict of Interest

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