

Unusual Clinical Presentation of Nutcracker Phenomenon

Dear Editor,

A 34-year-old female presented with progressively worsening left flank pain. The pain was initially relieved by analgesics but subsequently increased in severity and frequency. There were no complaints of haematuria, dysmenorrhoea or dyspareunia. Physical examination was essentially unremarkable with no palpable abdominal mass or pelvic varicose veins. Urinalysis suggested mild urinary tract infection.

Ultrasonography revealed mild left-sided hydronephrosis. This was further evaluated with computed tomography (CT) which demonstrated a vascular aneurysm compressing upon the left pelvi-ureteric junction (PUJ) (Fig. 1A). The initial impression was that of a large left renal artery aneurysm.

Catheter angiogram was performed with a view to treat the vascular lesion via endovascular approach. Diagnostic angiogram demonstrated no arterial abnormality. A renal venogram was acquired. It showed a venous aneurysm arising from the left renal vein (LRV), causing compression upon the left PUJ (Fig. 1B). The pressure gradient between the LRV and inferior vena cava measured approximately 5 mmHg, clinching the diagnosis of nutcracker syndrome (NCS). LRV hypertension is defined as a gradient equal or greater than 3.0 mm Hg.^{1,2} However, in advanced cases where collateral circulation has formed, the pressure gradient may be normal. The increase in venous pressure within the LRV is believed to be the cause of the large venous aneurysm.

The decision was made to proceed with endovascular treatment of the venous aneurysm. “Jailing” technique was used with stent-assisted coil embolisation of the venous aneurysm. At the same time, overlapping stents were deployed into the LRV. Longer stents were not available locally and hence 2 overlapping stents were used. Post-treatment venogram showed complete occlusion of the venous aneurysm (Fig. 1C).

The patient was discharged well 2 days later and remained asymptomatic after 6 months of follow-up.

Discussion

Nutcracker phenomenon (NCP) is a result of LRV compression, leading to outflow impedance from the LRV into the inferior vena cava and left renal venous hypertension.³ NCS is the clinical equivalent of NCP,

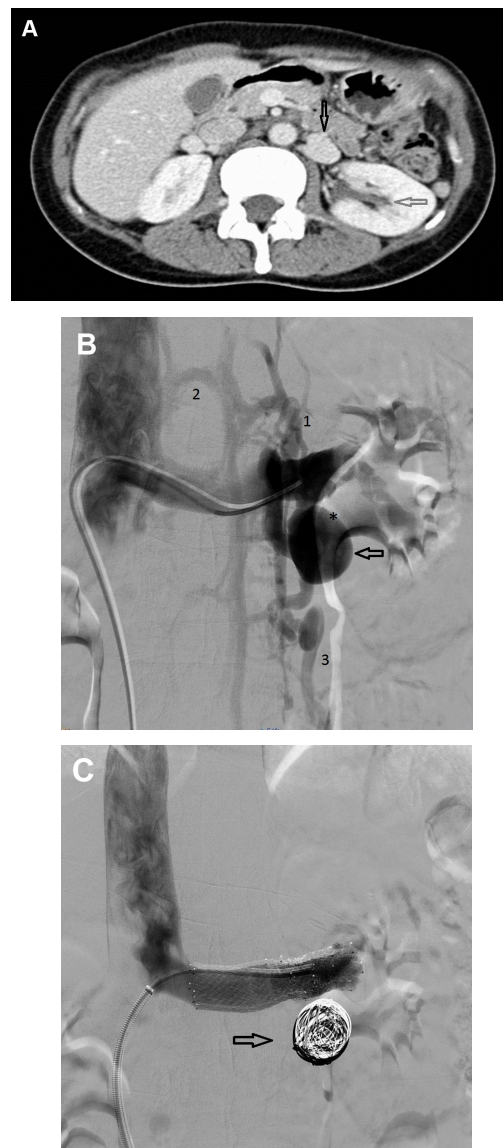


Fig 1. In A, contrast-enhanced computed tomography (CT) of the abdomen reveals a vascular aneurysm (black arrow), which compresses on the left pelvi-ureteric junction (PUJ), causing mild left hydronephrosis (grey arrow). In B, renal venogram shows a venous aneurysm (black arrow) arising from the distal left renal vein (LRV), causing compression on the left PUJ. Backflow into the hemiazygous system (1), left paravertebral venous plexus (2) and left gonadal vein (3) are noted, objective evidence of increased venous pressure (IVC). The pressure gradient between the LRV and IVC measured approximately 5 mmHg. In C, post-treatment venogram shows complete occlusion of the venous aneurysm (black arrow). In addition, satisfactory flow across the stented LRV is seen. Of note is also the absence of backflow into the LRV venous tributaries, indicative of reduced left renal venous pressure.

characterised by complex symptoms with substantial variations.⁴ Compression of the LRV between the aorta and superior mesenteric artery (SMA), known as anterior NCP, is the most common subtype. Less commonly, a retroaortic or circumaortic renal vein may be compressed between the aorta and adjacent vertebral body, known as posterior NCP. Occasionally, the third part of the duodenum courses anterior to the LRV. Hence, anterior NCP may coexist with compression of the duodenum by SMA, known as SMA syndrome (Wilkie syndrome).

The main presenting symptom is that of haematuria, believed to be due to rupture of thin-walled varices into the collecting system, presumably induced by renal venous hypertension.⁵ Patients may also present with left-sided flank pain, gonadal vein syndrome and varicocele.⁵ Gonadal vein syndrome is characterised by abdominal and flank pain aggravated by sitting, standing or walking, due to pelvic venous congestion. Left flank pain may also be a consequent of left ureteric colic, from passage of blood clots into the left ureter.

Management options for NCS range from expectant management to nephrectomy, depending on the severity of symptoms. Those with severe symptoms may benefit from surgical or endovascular intervention, as in our patient. Most interventions aim to reduce venous pressure within the LRV. Intravascular stenting, as applied in our patient, is a relatively new technique, extrapolated from stenting experience in May-Thurner and superior vena cava syndromes.⁶ Until stent endothelialisation occurs, anticoagulation therapy is hence recommended.

To our knowledge, although there has been previous report of PUJ obstruction associated with nutcracker syndrome, none was secondary to the presence of a venous aneurysm which was subsequently treated with coil embolisation.

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