CASE REPORT

Renal nutcracker syndrome in a young lady: unusual findings and endovascular management

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SUMMARY

Nutcracker syndrome (NCS) is caused by compression of left renal vein (LRV), usually between the aorta and the superior mesenteric artery (SMA). This can lead to obstruction of flow into the inferior vena cava and secondary left renal venous hypertension. Despite potential serious consequences, diagnosing NCS is often challenging, circuitous and commonly delayed. We report an extremely unique case of NCS. A 34-year-old woman presented with left flank pain and discomfort. On investigation, it was found that high pressure in the LRV, due to compression by the SMA, had led to a large venous aneurysm that had caused pelviureteric junction obstruction and hydronephrosis. Management was with stenting of the LRV and coil embolisation of the venous aneurysm with excellent clinical outcome.

BACKGROUND

Nutcracker syndrome (NCS) is caused by left renal vein (LRV) entrapment, most commonly between the aorta and superior mesenteric artery (SMA). This can obstruct venous return into the inferior vena cava (IVC) and lead to hypertension in the LRV. We report an unusual case where LRV hypertension due to NCS resulted in a venous aneurysm, pelviureteric junction obstruction and hydronephrosis.

CASE PRESENTATION

A 34-year-old woman without any significant medical history presented to us with left flank pain, progressively worsening over a 2-month period. The pain and discomfort were initially relieved by analgesics but subsequently increased in severity and frequency. The pain was worse in the evenings. There were no complaints of haematuria, dysmenorrhoea or dyspareunia. The patient was afebrile and haemodynamically stable. Physical examination was essentially unremarkable with no palpable abdominal mass. Urinalysis suggested mild urinary tract infection.

INVESTIGATIONS AND MANAGEMENT

Ultrasound was done which showed mild left-sided hydronephrosis. This was further evaluated with standard CT scan of abdomen and pelvis. The scan unexpectedly demonstrated a vascular aneurysm adjacent to the left collecting system (figure 1). There were multiple small tortuous vessels seen around the left renal hilum.

The overall impression at this time was that the patient had a large left renal artery aneurysm that was probably a flow-related aneurysm due to an underlying high flow renal arteriovenous malformation within the kidney.

Catheter angiogram was performed as the next step for further evaluation with a view to treat the vascular lesion in the same session through an endovascular approach. Angiogram unexpectedly demonstrated no abnormality in the arterial phase (figure 2). A careful review of the delayed images revealed an ill-defined abnormality in the venous outflow as the patient was unable to adequately hold her breath for a prolonged period of time.

Subsequently, left common femoral vein was then accessed, and a renal venogram was performed with the catheter in distal LRV. The venogram confirmed the presence of a venous aneurysm measuring 20.74×29.82 mm arising from the distal LRV, causing compression at the left pelviureteric junction. There were, in addition, multiple venous collateral vessels draining into the hemiazygos venous system and paravertebral venous plexus. There was suggestion of extrinsic mass effect on the mid LRV with appearance on renal venogram suggesting a significant anatomical and functional obstruction at this level (figure 3). Pressure gradient was measured between the LRV in its distal part and proximal renal vein and IVC. The gradient was 5 mm Hg confirming the diagnosis of NCS. The normal pressure gradient between the LRV and IVC is ≤1 mm Hg; LRV hypertension is defined as a gradient ≥3 mm Hg.^{1–3} The increase in venous pressure within the LRV was believed to be the cause of the large venous aneurysm.

Decision was made to treat the lesions during the same session through an endovascular approach: the obstruction was to be treated with a stent and the aneurysm was to be occluded with coils. We recognised that since it was a venous aneurysm causing an extrinsic mass effect, we would be able to thrombose the aneurysm by loosely packing it with limited number of coils. Left common femoral and right internal jugular venous accesses were obtained. Jailing technique was used with stent-assisted coil embolisation of the aneurysm.⁴ The aneurysm was easier to catheterise with microcatheter through the jugular approach while stent placement was easier through the femoral vein approach. A 2.2 F Progreat microcatheter (Terumo, Tokyo, Japan) was advanced into the aneurysmal sac.

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Figure 1 Contrast-enhanced CT image of the abdomen revealed large venous aneurysm (black arrow) with mass effect.

A 14×41 mm Epic (Boston Scientific, Natick, Massachusetts, USA) and 16×60 mm Smart stents (Cordis, Miami, Florida, USA) were placed in the left LRV across its entire length. Longer 14×80 mm stents were not available locally in the near foreseeable future, for which reason two stents were used. This was followed by embolisation of the venous aneurysm with non-fibred Cosmos coils (Microvention, Tustin, California, USA) and fibred coils Nestor coils (Cook, Bloomington, Indiana, USA).

Poststent placement and coil embolisation venogram showed complete occlusion of the venous aneurysm with normal flow of blood across the LRV and complete disappearance of the retroperitoneal venous collaterals (figure 4). There was complete disappearance of pressure gradient across the LRV on completion.

Figure 2 Catheter angiogram showed no abnormalities in the arterial phase.

Figure 3 Digital subtraction angiogram in venous phase confirms the presence of a venous aneurysm (black arrow) arising from the distal left renal vein, causing compression on the left pelviureteric junction (*). High back pressure is noted to cause opacification of the retroperitoneal venous system (1).

OUTCOME AND FOLLOW-UP

Postintervention period was uneventful, and the patient was discharged 2 days later, well and asymptomatic. The patient continues to stay asymptomatic after 6 months of follow-up.

Figure 4 Post embolisation venogram shows complete occlusion of the venous aneurysmal sac (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. LRV, left renal vein.

DISCUSSION

NCS is characterised by symptomatic LRV compression that leads to outflow impedance into the IVC and left renal venous hypertension. It is most common in females in their fourth decade of life.⁵ Compression of the LRV between the aorta and SMA, known as anterior NCS, is the most common subtype. Less commonly, a retroaortic or circumaortic renal vein may be compressed between the aorta and adjacent vertebral body, giving rise to posterior NCS.³ Other reported, although rare, variations include compression of the LRV between the SMA and the right renal artery⁶; a posterior NCS with entrapment of a retroaortic duplicated LRV⁷ and compression of a left-sided IVC,⁸ to name a few. Occasionally, the third part of the duodenum courses anterior to the LRV. Hence, anterior NCS may coexist with entrapment of the duodenum by SMA, known as superior mesenteric artery syndrome (Wilkie syndrome).³

The main presenting symptom, for around 80% of cases, is haematuria, as renal venous hypertension leads to the rupture of thin-walled varices into the collecting system.^{5,9} Haematuria may even occasionally lead to anaemia, and this has been found in about 13% of NCS cases.⁹ Patients may also present with left flank pain, gonadal vein syndrome and varicocele.^{3,5} Gonadal vein syndrome, caused by pelvic venous congestion, leads to abdominal and flank pain that can be aggravated by sitting, standing or walking.³ Left flank pain may also be due to blood clots in the left ureter causing ureteral colic.³ In addition to the symptoms above, the initial presentation of the disease may also include proteinuria.⁹ Dysmenorrhoea, dyspareunia, dysuria, chronic fatigue, left iliac fossa or loin pain and lower limb varicosities have also been associated with this disease.⁹

Varying imaging methods have been used in the diagnosis of NCS. Renal angiography demonstrates delayed venous washout from the left kidney while intravenous urogram may show notching of the left renal pelvis and ureter from adjacent varices and collaterals. Although standard CT is insufficient to clinch the diagnosis of NCS, it is particularly useful in excluding other causes of haematuria such as tumours and vascular malformation. It also has the ability to clearly delineate spatial relations between vessels. Doppler ultrasonography is an easily accessible and non-invasive modality for evaluating suspected NCS. It was, however, not performed in our patient as the initial CT shows no evidence of hilar dilatation and mesoaortic narrowing of the LRV to suggest the presence of NCS.

Management options for NCS range from expectant management to nephrectomy, depending on the severity of symptoms. Those with serious impairment or severe symptoms may benefit from surgical or endovascular intervention, as in our patient. Most interventions aim to reduce venous pressure within the LRV. Surgical management includes options such as nephropexy,¹⁰ LRV bypass,¹¹ renal-to-IVC shunt¹² and nephrectomy.¹³ Intravascular stenting, as used in our patient, is a relatively new technique. Good outcomes with endovascular stents have been described but this technique is not exempt from complications such as stent migration, restenosis, thrombosis and embolism.^{3,14,15} However, overall, the endovascular approach, with its minimally invasive nature, is fast gaining traction in recent times.

To our knowledge, there are no previous reports of a renal vein aneurysm as a result of NCS in an adult patient (only one such case exists in the context of a paediatric patient¹⁶). Renal vein aneurysm causing symptomatic pelviureteric junction obstruction and hydronephrosis is also unique in our patient. Our treatment approach was successful in alleviating her symptoms, and she continues to do well.

Learning points

- ▶ Nutcracker syndrome (NCS) may present with just left-sided flank pain.
- ▶ The renal venous hypertension caused by NCS can be severe enough to lead to formation of an aneurysm and development of hydronephrosis.
- ▶ Management options can range from conservative to surgical, with the endovascular approach becoming an increasingly common one.

Contributors MT and BSYC were involved in the direct care of the patient. MT, BSYC and KD were involved in writing of this paper.

Competing interests None declared.

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