

Nutcracker Syndrome Managed by Endovascular Approach

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Abstract

Nutcracker syndrome is a clinical syndrome, presenting with various symptoms caused by compression of left renal vein, between aorta and superior mesenteric artery, due to narrow angle between these vessels. Patient may present with abdominal pain, hematuria, or symptoms of urinary tract infection. We present a case of Nutcracker syndrome presenting with chronic abdominal pain, managed by percutaneous intervention.

Keywords: nutcracker; hematuria; stent

Case

44 years female, presented with pain in abdomen for the last one and half years. Pain was present in left lumbar region and left iliac fossa and radiated to back. Pain initially was mild in intensity, but for the last 6 months, it was moderate in intensity and increased after meals and was aggravated during menstruation. Patient took treatment in various hospitals, without any relief. Patient also gave history of intermittent hematuria and also intermittent burning micturition. 3 months back, her ultrasound was done in some outside hospital, which reported mildly dilated venous channels seen in the region of left renal vein-possibility of compression of left renal vein was kept.

CECT of abdomen was done in same hospital which reported- Grossly dilated venous channels are seen in the pelvic cavity-likely pelvic congestion, with maximum diameter of venous channels approximately 12 mm. Left ovarian vein is dilated with opacification of left ovarian vein in corticomedullary phase from left renal vein and measures up to 11 mm in diameter. There is abrupt narrowing of left renal vein as it courses posterior to SMA due to compression between aorta and SMA with approx. 90% luminal narrowing. SMA and aorta angle is approximately 5 degrees. With space between Aorta and SMA at the level of left renal vein is approx. 3.3 mm.- likely anatomical variant/ nutcracker syndrome.

Blood investigations:

Hemoglobin-11.3 gm%

TLC-7.7x1000/cumm

Polys-65%

Platelet count-99x1000/cumm

ESR-15 mm in 1st hour.

Urea-21.8 mg/dl

Creatinine-0.49mg/dl

Keeping in view the possibility of Nutcracker syndrome (NKS). patient was taken up for percutaneous treatment. Right femoral vein puncture was done. Femoral vein angiography was done to see inferior vena cava (IVC), (Fig.1) to have idea of origin of left renal vein. Femoral artery access was also taken and left renal arteriography was done to trace renal vein in venous phase, but it could not be visualised. Since renal vein was not visualised, blind entry was done with terumo wire passed through right Judkins (JR)catheter. Catheter was then negotiated into LRV and venography was done (Fig. 2). It showed markedly dilated left gonadal vein (LGV), and left ovarian vein (Fig 3). Hardly any contrast was entering into IVC, due to compression by superior mesenteric artery (SMA). It was decided to put a stent in LRV across the compressed segment. To properly define the site of obstruction and stent placement, a JR catheter was placed in SMA (Fig 4), from left brachial artery approach. Self-expanding venous stent was then deployed, size 14x 60 mm. (Fig 5). LRV angio was then done which showed free flow of contrast into IVC and minimal reflux into LGV (Fig. 6). If, large reflux persisted, then coiling of LGV would have been required.



Figure 1: Inferior vena-cavography



Figure 2: Left renal venography shows hugely dilated gonadal vein, with very little flow into inferior vena cava (Due to renal vein compression and reflux of blood into gonadal vein)



Figure 3: Marked dilatation of left ovarian vein, due to reflux from gonadal vein.



Figure 4: Catheter in superior mesenteric vein, from left brachial artery approach, as marker to guide the position of stent.



Figure 5: Self expanding stent placed in left renal vein across the superior mesenteric artery crossing.



Figure 6: Left renal venography shows free flow into inferior vena cava, with hardly any reflux into gonadal vein.

Discussion:

NKS is a rare condition caused by compression of LRV between aorta and SMA [1]. Patient has non-specific symptoms and so condition remains undiagnosed for long time. Symptoms include flank pain, intermittent hematuria, orthostatic proteinuria and autonomic dysfunction [2]. Women may present with pelvic congestion syndrome,

dysmenorrhoea or dyspareunia. Men may present with left sided varicocele. NKS may cause infertility in both sexes. NKS refers when anatomy coincides with symptoms, while asymptomatic persons with anatomy are labelled as nutcracker phenomenon.

There are 2 varieties of NKS:

1. Anterior NKS- compression of LRV between aorta and SMA.

2. Posterior NKS- compression of LRV between aorta and spine.

NKS occurs when angle between aorta and SMA is reduced to less than 40degree. Normal 4-5 mm space between the vessels is maintained by retroperitoneal adipose tissue and 3rd part of duodenum. So, thinner individuals are more affected. In some cases, relief of symptoms have been reported with weight gain [3].

- NKS is difficult to diagnose and requires a high index of suspicion. Patients suspected to have NKS on the basis of intermittent hematuria, flank pain, features of PCS and left sided varicocele should undergo renal Doppler ultrasonography. It has sensitivity of 69-90% and specificity of 89-100% to diagnose NKS [4].
- In adults, the ratio of the anteroposterior diameters of the hilar portion of the left renal vein to the aortomesenteric portion is greater than four and the peak flow velocity ratio of the left renal vein at the site of narrowing compared to the flow proximal to the narrowing is greater than five [5]. Additional features include venous collaterals and pelvic varicosities.
- CT angiography is good option in doubtful cases. It has a sensitivity of 92% and specificity of 89% [6]. It is based upon ratio of LRV diameter at the hilum and at the compressed segment more than 4.9, ratio of lateral uncompressed segment to compressed segment more than 2.25, as also on the angle between SMA and aorta less than 35 degree.
- MRI has sensitivity and specificity similar to CT but has better soft tissue visualization [7].

Management: Initial management, especially in children should be conservative, as there are chances of spontaneous relief with weight gain. Around 75% of all children with NKS will have complete resolution of symptoms within two years [4].

Surgical treatment is recommended in patients with severe hematuria, severe proteinuria, varicocele, renal failure and severe pain. Endovascular stenting was first described as therapy for NKS in 1996 and results are good [8,9]. 6-8 Cm long stent with diameter of 12-14mm is placed near

the first division of left renal vein to prevent stent migration. If significant reflux persists into the LGV after stent placement, then embolization of LGV may be required. Stent migration, erosion into adjacent structures, and kinking are the most common complications of endovascular procedures.

Our patient remained undiagnosed for one and half years, she was lucky to have been subjected to CT from outside, after suspicion on ultrasound. We implanted a long stent. Patient became symptom free within 3 days and is doing well. She will be subjected to ultrasound after 3 months and then annually for 3 years.

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