Case Reports

Two cases of nutcracker syndrome

L N Senevirathna¹, N D Perera², M Wijayarathne³

Sri Lanka Journal of Child Health, 2010; **39**: 112-114

(Key words: Nutcracker syndrome, left renal vein, LRV, ureteroscopy, haematuria)

Introduction

Entrapment of the left renal vein (LRV) between the abdominal aorta (AA) and the superior mesenteric artery (SMA) was first described in 1950 by El Sadr and Mina and given the name nutcracker syndrome (NS) by De Schepper and Chait. These two cases highlight a rare cause of haematuria viz. nutcracker syndrome. Due to its relative rarity and non availability of advanced imaging, it is rarely diagnosed and is under-reported in developing countries.

Case reports

Two boys aged 9 & 7 years presented with 4 and 3 episodes respectively of intermittent gross

macroscopic painless haematuria appearing nearly once a year since the age of five and four respectively. Examination was unremarkable and both were normotensive. Initial investigations, including urine for red cell casts, renal function tests, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibodies, renal biopsy and cystoscopy were normal excluding medical and surgical causes such as urolithiasis and tumours. Ureteroscopy performed during an episode revealed left ureteric haematuria and Doppler ultrasonography and spiral CT scan demonstrated entrapment of left renal vein between abdominal aorta and the superior mesenteric artery (Figures 1 & 2). Since the patients had minimal symptoms both were treated by close observation and follow-up.

Figure 1: 3D reconstruction of CT scan - Left renal vein (LRV) entrapment between the superior mesenteric artery (SMA) and abdominal aorta (AA) resulting in dilated proximal LRV and distal LRV of normal calibre

¹Senior Registrar in Urological & Transplant Surgery, ²Consultant Urologist, ³Consultant Vascular & Transplant Surgeon

Figure 2: Contrast CT scan of the abdomen at the renal hilar level
 AA aorta, SMA superior mesenteric artery, P dilated proximal left renal vein, D compressed distal left renal vein

Discussion

Nutcracker phenomenon, which is asymptomatic dilatation of the left renal vein, should be differentiated from nutcracker syndrome (NS) where patients with LRV hypertension are symptomatic with macroscopic or microscopic haematuria, orthostatic proteinuria, varicocele and hypertension¹. However, nutcracker syndrome can exist even in entrapped non-distended LRV and normal flow can also exist in distended LRV. Therefore, nutcracker phenomenon or syndrome should be defined only when the clinical signs are present along with compatible radiological findings.

The pathophysiology of the NS is not fully understood. Although passage of the renal vein in the fork formed by the aorta and SMA is a normal anatomical finding, it is not known why compression of the vein occurs in only a few patients. Wendel proposed that posterior renal ptosis with stretching of the LRV over the aorta leads to venous hypertension. Hoffen-fellner suggested that abnormal branching of the SMA and the resulting compression is the cause of elevated pressure gradients between the proximal segment of the LRV and vena cava in such patients². LRV

hypertension leads to increased pressure in the venous system resulting in rupture of the thin walled septum separating the veins from the collecting system in the renal fornix resulting in haematuria. Furthermore, stagnating venous pool in the gonadal veins will give rise to pelvic congestion. This theory was further highlighted by Shokeir who performed CT scans to compare the anatomical relations of the LRV with the SMA and aorta in patients with the NS and in healthy control patients³.

Clinical features include macroscopic and microscopic haematuria which is unilateral, uniquely from the left side, left abdominal pain, flank pain, pelvic or scrotal discomfort due to varicocele or ovarian/gonadal vein syndrome. The gonadal vein syndrome is characterized by abdominal and flank pain exacerbated by standing, sitting or walking. On the other hand, 'pelvic congestion syndrome' is characterized by symptoms of dysmenorrhoea, dyspareunia, post-coital ache, lower abdominal pain, dysuria, pelvic, vulval, gluteal or thigh varices and emotional disturbances.

Diagnosis is difficult. At panendoscopy haematuria from the left ureteric orifice in the absence of any

other detectable pathology should raise the suspicion of the nutcracker phenomenon⁴. The differential diagnoses one must consider are other rare causes of haematuria such as Henoch-Schonlein purpura, lupus erythematosus, renal endometriosis, haemangioma, cysts of the renal papillae, renal papillary angiomas, venocalyceal fistula, panarteritis nodosa and retrocaval ureter. Eventual diagnosis is from imaging which includes

ultrasound scan with combined renal Doppler studies, CT angiograms, MRI and venography. Renal Doppler ultrasound revealed a significant difference in the diameter and peak velocity between the hilar and aorto-mesenteric portions of the LRV.⁵ (Figure 3)

Figure 3: Doppler ultrasound study comparing the venous pressure of the left renal vein distal to the compression (left) and proximal to the compression (right)

Treatment of the NS is controversial. Therapy should be dictated by the clinical symptoms. Conservative therapy has been proposed for patients with mild haematuria where spontaneous resolution of haematuria has been reported. Surgery is indicated for patients with persistent, severe, life threatening haematuria and significant pain. Wendel has performed medial nephropexy with excision of the renal varicosities². LRV bypass or transposition of the LRV or rarely SMA has been done⁶. Autotransplantation is another alternative that allows better protection of the kidney against ischaemia by proper cooling and irrigation. Lately, minimally invasive techniques to place endovascular stents across the LRV have given encouraging results.⁷ Silver nitrate solution instilled directly on to the renal pelvic membrane via an ureteroscope to stop the haemorrhage has been tried without convincing long term results.

In summary, the nutcracker phenomenon is a recognized but unusual cause of haematuria. The diagnosis should be borne in mind in patients with unexplained unilateral haematuria which requires careful investigation to avoid delay in treatment.

References:

1. Robert L. Nutcracker phenomenon or nutcracker syndrome. *Nephrology Dialysis Transplantation* 2005; **20**(9):2015

2. Hersey N et al. The Nutcracker phenomenon – Case report and review of literature. *Current Urology* 2007; **1**:110-2.
3. Fu WJ et al. Nutcracker phenomenon: a new diagnostic method of multistatic computed tomography angiography. *International Journal of Urology* 2006; **13**:870-3.
4. A Wang L et al. Diagnosis and surgical treatment of Nutcracker Syndrome. *Urology Today* 2009; **10**:1016.
5. Jung-Eun C. et al. Nutcracker syndrome in children with gross haematuria. Doppler sonographic evaluation of the left renal vein. *Pediatric Radiology* 2006; **36** (7): 682-6.
6. Ahmed K, Sampath R, Khan MS. Current trends in the diagnosis and management of renal nutcracker syndrome; a review. *European Journal of Vascular & Endovascular Surgery* 2006; **31**:410-6.
7. Segawa N et al. Expandable metallic stent placement for nutcracker phenomenon. *Urology* 1999; **53**:631-3.