

## Anterior Nutcracker syndrome as a rare cause of hematuria in an 8 year old boy: A case report

Prashant S Patil, Gupta Abhaya, Kothari Paras L, Kekre Geeta, Dikshit Vishesh K, Shahaji Deshmukh, Apoorva Kulkarni

*Department of Pediatric Surgery, L.T.M.G. Hospital, Sion, Mumbai, Maharashtra. India*

### Abstract

Nutcracker syndrome is caused by a compression of the left renal vein between the aorta and the superior mesenteric artery. It results in left renal venous hypertension, and the subsequent development of venous varicosities of the renal pelvis, ureter, and gonadal vein. We report a rare case in an 8-year-old boy who presented with a history of intermittent episodes of hematuria leading to anemia and left flank pain. Ultrasound examination of abdomen revealed bulky left kidney and features of subacute medical renal disease. Computed tomography of abdomen showed compressed left renal vein between aorta and superior mesenteric artery. Authors report the successful operative management of this rare syndrome in a pediatric patient.

### Keywords

Nutcracker syndrome; hematuria; renal vein transposition; left renal vein; children.

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**Corresponding Author:** Dr. Prashant S Patil

*Ward 1A, Pediatric Surgery ward, L.T.M.G. Hospital, Sion, Mumbai*

*E-mail: [docprashant2010@gmail.com](mailto:docprashant2010@gmail.com)*

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### Introduction

Nutcracker syndrome (NCS) is quite rare condition and characterized by the compression of the left renal vein (LRV) between aorta and superior mesenteric artery (SMA) [1]. It is also called renal vein

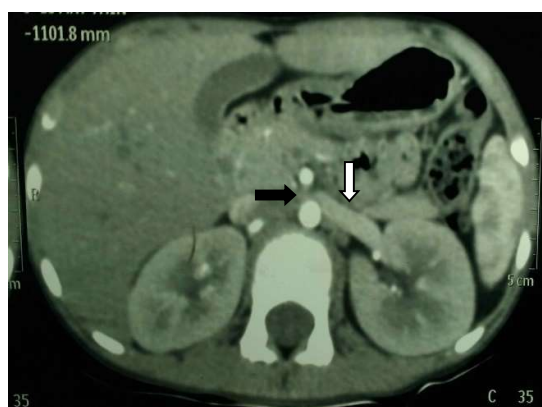
entrapment syndrome [2]. Clinical symptoms are usually caused by venous hypertension within the left kidney: left-flank pain that can radiate to the buttock and non-glomerular hematuria [3]. El Sadr reported the first case in 1950 [4], but De Schepper named it [5]. In normal anatomical development, the left renal vein (LRV) passes anteriorly across the abdominal aorta. But, in rare cases, the left renal vein becomes located in a retro-aortic position,

compressed between the aorta and the vertebral column. The condition that LRV is anterior to the abdominal aorta is called anterior nutcracker syndrome. The other is called posterior nutcracker syndrome [6]. Doppler ultrasound measurements of the anterior-posterior (A-P) diameter and peak velocities of the LRV may be helpful in diagnosis of NCS [7]. Computed tomography (CT) and CT angiography, Magnetic resonance imaging (MRI) and MR angiography are other noninvasive modalities that can demonstrate compression of the LRV in the aorto-mesenteric angle and collateral veins [8]. Here, we report our experience treating a young boy with NCS.

### Case Reports

An 8 years old boy was admitted with a two months history of hematuria and left flank pain. Urine analysis revealed that the patient had gross hematuria and pyuria. His renal function tests were normal. Hemogram revealed low hemoglobin. He was given intravenous antibiotics. USG abdomen showed bright echotexture in bilateral kidneys with decreased cortico-medullary differentiation. Left kidney was bulkier than right one. He was discharged from hospital on oral antibiotics but after two weeks the patient's symptoms recurred. He was again admitted. CT abdomen was done which

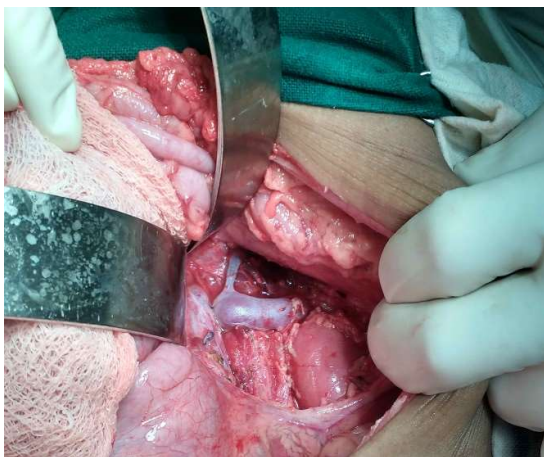
showed significant decrease in aorto-mesenteric angle measuring 22 degrees and aorto-mesenteric distance measuring 5 mm [Fig. 1] causing compression of LRV between SMA and aorta. Dilatation of renal vein was seen before compression with small hypodense filling defect within suggestive of thrombus.



**Fig. 1.** CT abdomen showing stenosis of the left renal vein between the superior mesenteric artery and the aorta (black arrow) with proximal dilatation of the left renal vein (white arrow).

LRV Doppler showed internal diameter of left renal vein at crossing of SMA and aorta was less than 4 mm and distal to it was 7 mm. There was no thrombus in LRV or inferior vena cava (IVC). The highest peak velocity was detected at this point. No pelvic or ureteral notching was identified on CT. Patient was operated in supine position under general anesthesia via midline

abdominal incision. Left side colon and small bowel was retracted to right side of midline and posterior peritoneum and Gerota's fascia over the left kidney opened. LRV was found to be dilated. LRV was traced up to its insertion into IVC. Left suprarenal vein, left gonadal vein and left lumbar vein draining into LRV identified and preserved. IVC delineated from the point of insertion of left renal vein to 4 cm distally. LRV and left renal artery looped up. IVC controlled with a side biting clamp. LRV clamped, divided and stump transfixed at IVC with prolene 7-0 RB. Opening made in the IVC 4 cm from its junction with the renal vein and end to side anastomosis done with prolene 7-0 RB [Fig. 2].



**Fig. 2.** End to side anastomosis between LRV and IVC.

IVC declamped, hemostasis achieved. From incision to final wound closure, the overall

time of the surgery was nearly 130 minutes. Total blood loss, as calculated from anesthesiology charts, was less than 40 ml.

Patient was given intravenous antibiotics till 10<sup>th</sup> post-op day. Low molecular weight heparin was given till 3<sup>rd</sup> post-op day and later patient was shifted to oral anticoagulants. Patient had persistent hematuria and developed hypoalbuminemia for which blood transfusion and intravenous albumin infusion was given. Renal vein doppler done on 15<sup>th</sup> post-op day showed patent renal vein. Patient was discharged on 17<sup>th</sup> post-op day and is on follow-up for 5 months. Patient is on Tb Ecosprin 75 mg OD and Tb. Envas 5 mg BD. His monthly renal function tests are normal. However he has persistent hematuria though to a lesser degree. His bilateral medullary disease could explain persistent hematuria. His abdominal pain has subsided completely.

### Discussion

Controversy also exists regarding treatment of nutcracker syndrome. Only symptomatic and disabling nutcracker syndrome should be treated. Conservative management with routine urinalysis is proposed for mild hematuria, since the development of collateral veins may resolve the hypertension in the left renal vein and alleviate symptoms [9]. Children should be

treated conservatively because cases of spontaneous remission have been reported during growth [10,11]. Different approaches can be used.

Medication can be proposed only in patients suffering from isolated pelvic congestion syndrome caused by nutcracker syndrome. In this case, symptoms can be improved by treatments effective in treating pelvic congestion syndrome, as medroxyprogesterone acetate, [12] goserelin acetate, [13] and micronized purified flavonoid fraction [14].

Indications for surgery include severe persistent or recurrent hematuria causing anemia, and blood clots causing abdominal or flank pain. Surgical options include left renal vein transposition [15], LRV bypass [16], SMA transposition [17] and autotransplantation [18]. External stenting, gonadocaval bypass, left gonadal vein transposition, patch angioplasty, LRV phlebolytic, renal fixation have also been reported as other surgical techniques [3]. Hohenfellner even reported one case which was radically treated by nephrectomy [19].

More recently, endovascular treatment options have been applied [3,7,9]. This approach is less invasive as it can be performed percutaneously. Endovascular treatment options include embolization, balloon angioplasty and stenting [3].

LRV transposition is nowadays regarded as the surgical method of choice, [20] and has been used to treat 28 patients, with excellent results in 27 cases and 1 recurrence. Left renal vein transposition involves dividing the left renal vein at its junction with the inferior vena cava, repair of the vena cava defect and the re-anastomosis of the left renal vein to the inferior vena cava at a lower level away from the superior mesenteric artery. A transposition of the superior mesenteric artery is based on similar surgical principles, but is more difficult compared to a left renal vein transposition. This procedure involves risks that include bleeding, thrombosis and a paralytic ileus [15]. A thrombosis of the superior mesenteric artery would be disastrous for patients undergoing this operation.

Our patient had persistent hematuria leading to anemia, recurrent episodes of flank pain affecting his daily activities hence conservative management was not attempted. Left renal vein transposition is technically easy to perform even in pediatric age group with preservation of left suprarenal vein, left gonadal vein and branches of left lumbar vein. To our knowledge there are very few reported cases of treated Nutcracker syndrome in pediatric patients and experience of this case suggests

that this syndrome is treatable in patients below 18 years in symptomatic cases.

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