Management and diagnosis of Nutcracker syndrome-a case report

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Introduction

Nutcracker syndrome (NCS), also known as left renal vein (LRV) entrapment syndrome is a condition that leads to stenosis of the aorto-mesenteric region of the LRV, with dilatation of the distal portion of the vessel. The most common characteristic clinical signs and symptoms are intermittent hematuria, proteinuria, flank pain, pelvic congestion in females, and varicocele in male patients, alongside with diagnostic imaging of the anatomy associated with the syndrome.

Case presentation

In June 2019, a 43-year-old woman was admitted to the Urology department with symptoms of intermittent painless hematuria and mild to moderate left lumbar pain for the last three months. The patient has no other symptoms like dysuria, constipation and etc. On physical examination the patient had a regular pulse of 74 beats/min, a temperature of 36.7 °C, and a respiratory rate of 17 breaths/min, also the abdomen was palpable painless. On the ultrasonography no pathological finding were observed. Patient’s routine laboratory results such as complete blood cell count, renal function tests, liver function tests were in normal ranges. Urine analysis showed 15 proteinuria.

The patient underwent cystoscopy under general anesthesia. Bloody urine was noticed to appear from the left ureter ostium and an intravenous contrast CT of the abdomen was performed. The final diagnosis was anterior Nutcracker syndrome.

The patient refuse suggested operative treatment and due to that reason a conservative treatment with angiotensin converting enzyme inhibitor- Lisinopril 5 mg and Aspirin 100 mg daily were commenced, to improve renal perfusion. Also the patient was advised to gain weight and expecting to increase in retroperitoneal adipose tissue, and hence reduction in LRV tension. Furthermore the patient was advised to be followed up every 6 months with a control examinations and if needed a intra venous CT scan.

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Discussion

The exact prevalence of NCS is unknown, partly because of an absence of definitive diagnostic criteria and partly because of the variability in symptomatic presentation. The prevalence of this condition has been reported as higher in females, but some recent studies have shown that it is equally prevalent among both genders. Hematuria is the most commonly reported symptom and is attributed to rupture of thin-walled varices, due to elevated venous pressure, into the collecting system. It varies from microhematuria to macrohematuria, occasionally with resultant anemia that requires blood transfusions. Cystoscopy may identify a left ureteral origin. The causes of isolated hematuria could not be identified by routine methods in 69% of pediatric cases. Of those, 40% were found to have NCP by renal Doppler ultrasonography (DUS); although microhematuria in these patients was 4 times more common than macrohematuria, there were no differences in peak renal vein systolic velocities. In our case it was a 43-year old woman with complaints of 3 months.

Both CT and MRI can demonstrate compression of the LRV in the fork formed by the SMA and abdominal aorta, gonadal vein distension, and pelvic congestion. However, despite the accuracy with which they demonstrate the anatomy between the LRV and the surrounding structures, neither CT nor MRI are dynamic modalities and therefore do not accurately measure flow velocity and direction. The most specific finding with CT for NCS was a LRV diameter ratio (hilar to aortomesenteric ratio) $\geq 4.9$ (specificity 100%). However, the highest diagnostic accuracy observed in axial CT images is “the beak sign.” These two main findings on CT scan were presented in our case too, with aortomesenteric ratio $> 4.9$ and also presence of “Beak sign.”

The treatment of NCS remains a controversial topic, both in terms of good indications for treatment as different diagnostic criteria are used, and in terms of the best treatment modality for specific individuals. In cases presenting with mild hematuria or with mild and tolerable symptoms, conservative management is recommended. However, surgery may be considered for gross hematuria (especially if recurrent); for severe symptoms including flank or abdominal pain, anaemia, autonomic dysfunction, impairment of renal function including persistent orthostatic proteinuria, varicocele formation; and for ineffective conservative measures after 24 months in patients aged less than 18 years and after 6 months in adults. In our case, because the symptoms of the patient were mild to moderate and the hematuria was not prolonged, she refused to undergo suggested surgery and a conservative therapy and lifestyle changes were recommended.

The natural history and the exact prognosis of NCS is not clear and it remains an under-diagnosed condition because of its non-specific presentation. However, without treatment it can predispose to left renal vein thrombosis, followed by a kidney damage. Increasingly, the use of a combination of techniques is being recognised such that the use of open surgery combined with endovascular stenting has shown a resolution of symptoms, specifically hematuria, proteinuria, and varicoceles.
Despite the non specific presentation of NCS, it is important that it is not missed and recognised and treated, not only to prevent damage to the kidneys from chronic renal vein thrombosis, but also to improve symptomatology and relieve pain for patients. Treatment is dictated by the severity of symptomatology and different treatment approaches from conservative, medical, and surgical and all, have to be chosen patient individually and all have evidence bases.

Declaration of competing interest

The authors declare that they have no competing interests.

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Fig. 3. Sagittal MPR in arterial phase shows decreases angle between SMA and Aorta/red arrow/- at range 29°.