Posterior nutcracker syndrome with left renal vein duplication as a cause of gross hematuria and recurrent left varicocele in an eight-year-old boy

Malek Barka, Faouzi Mallat, Wissem Hmida, Sidiya Ould Chavey, Khaled Ben Ahmed, Amel Ben Abdallah, Kalthoum Tlili

ABSTRACT

Introduction: The posterior nutcracker syndrome with duplication of the left renal vein is extremely rare, especially in childhood, and often misdiagnosed because it embraces an extended non-pathognomonic spectrum of symptoms which imply a difficult diagnosis.

Case Report: An eight-year-old boy with a history of left varicocele treated by Palomo technique one year ago, presented with intermittent painless hematuria. Systemic examination revealed high degree of left varicocele. An abdominal computed tomography scan revealed left renal vein duplication. The retroaortic branch was compressed between aorta and the vertebral column, suggesting posterior nutcracker syndrome with left renal vein duplication. Transposition of the posterior branch of left renal vein or autotransplantation of the left kidney were decided, but refused by parents of the patient and only the varicocele was managed. The child is currently asymptomatic and proposed to clinical and analytical assessment.

Conclusion: Although posterior nutcracker syndrome with duplication of the left renal vein is a rare entity, it should be considered in the differential diagnosis of hematuria and recurrent varicocele.

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CASE REPORT

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Keywords: Posterior nutcracker syndrome, Renal vein duplication, Hematuria, Recurrent varicocele

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INTRODUCTION

Nutcracker syndrome also known as left renal vein entrapment is an uncommon cause of hematuria [1–3]. Most usually, the left renal vein (LRV) is entrapped between aorta and superior mesenteric artery and it is called anterior Nutcracker syndrome. Posterior nutcracker syndrome is due to a retroaortic position of the LRV compressed between the aorta and the vertebral column [4]. This entity affects women more than men and in most cases present in the third or fourth decades of life [5].

Herein, we present a case report of a posterior nutcracker syndrome with LRV duplication observed in our institution in an eight-year-old boy.

CASE REPORT

An eight-year-old boy, with a history of left varicocele treated by Palomo technique one year ago, was admitted

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to the pediatric department with a gross hematuria. On physical examination, the patient was pale, his vital signs were within normal limits, and systemic examination revealed only high degree of left varicocele. His hemoglobin on presentation was 9.2 g/dL, the urine analysis confirmed gross hematuria and the other biological analyses were normal.

X-ray and ultrasonography of the abdomen were normal. When we asked him again, he complained of abdominal pain spatially in the left flank. He denied any other urological signs or symptoms, namely, dysuria, fever, asthenia, or fatigue.

The abdominopelvic computed tomography angiography (CTA) revealed LRV duplication with a retroaortic branch entrapped between aorta and vertebral column, promoting a posterior nutcracker syndrome (Figures 1 and 2). Based on the clinical and radiological presentation, early recurrent left varicocele, left flank pain and gross hematuria and imageries finding; a posterior nutcracker syndrome with duplication of the LRV was confirmed. Blood transfusion was not needed.

For the management of nutcracker syndrome, transposition of the posterior branch of LRV or autotransplantation of the left kidney were decided, but refused by parents of the patient.

A surgical cure for the recurrent left varicocele was performed by inguinal approach. Thereafter, he was counseled to restrict intense physical exercise and take oral iron supplements. This therapy showed decrease in episodes of hematuria and substantial improvement of anemia (actual hemoglobin: 11 g/dL). The patient is currently asymptomatic and proposed to clinical and analytical assessment. At the last follow-up, he

Figure 2: Computed tomographic angiography showing compression of the posterior branch of the left renal vein (red arrow) between abdominal aorta and vertebral column.

was asymptomatic without recurrence of hematuria or varicocele.

DISCUSSION

The first case of nutcracker syndrome was described in 1950 but this term must be distinguished from the nutcracker phenomenon which is a relatively common anatomical variance, in which the patient stays asymptomatic [4, 6]. The degree and the stage of syndrome depend on the renal venous pressure. Well developed collateral veins or the presence of the preaortic branch of duplicated renal vein as in our case could dissipate the high pressure gradient and diminish the blood flow volume of the LRV, resulting in the absence of distended LRV and LRV hypertension [7].

The clinical manifestation of nutcracker syndrome ranges a wide variety from asymptomatic microscopic hematuria to severe pelvic congestion syndrome characterized by an array of signs and symptoms such as dyspareunia, dysmenorrhea, lower abdominal pain and pelvic, perineal and lower limb varices [8]. The classic presentation include symptoms which are often aggravated by physical activity such as hematuria, varicocele as seen in our case, pain or gonadal vein syndrome, orthostatic proteinuria and orthostatic intolerance [9–12]. Hematuria is the most commonly reported symptom and is attributed to the communication between dilated venous sinuses and adjacent renal calices [13]. Varicoceles almost always occur on the left side and affect up to 9.5% of cases and it is admitted that LRV hypertension is the usual cause of varicoceles [14, 15]. The ultrasonography is a good first radiographic study as it is safe, cost effective and screens possible renal and bladder abnormalities and identifies the turbulent vessel renal flow [16–18]. On the other hand, computed tomography

Figure 1: Computed tomographic angiography (reconstructive CTA) showing duplication of the left renal vein with compression of the posterior branch (red arrow) between abdominal aorta and vertebral column and a normal anterior branch (yellow arrow).

(CT) scan is the most appropriate and single imaging modality in a patient with multiple abnormalities like ours [19].

In this case, the computed tomographic angiography (CTA) led to this diagnosis by showing a dilated posterior LRV branch being compressed between aorta and vertebral column.

The management of nutcracker syndrome evolved in the last four decades depends upon the clinical presentation, and the severity of the left renal vein hypertension, including close expectant surveillance, endoscopic proceedings such as external or internal stenting and more complex open surgical procedures LRV transposition, renal autotransplantation of the left kidney [20–23]. Patients in prepubertal age like our case should be offered less aggressive forms of treatment since the likelihood of spontaneous remission due to normal physical development and despite the initial presentation with hospitalization, our patient was managed conservatively with good evolution [4].

CONCLUSION

Posterior nutcracker syndrome with duplication of the left renal vein is a rare vascular anomaly, which may be a possible cause of hematuria and recurrent varicocele. Computed tomography angiography is the optimum choice of imaging modality to diagnose posterior nutcracker syndrome as well as to find out associated other abnormalities. Conservative treatment with strict follow-up is recommended for patients with microscopic hematuria or intermittent short periods of painless gross hematuria, insignificant pain. Surgical reconstruction can be indicated in case of severe symptoms or complications.

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Author Contributions

Malek Barka – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Kalthoum Tlili – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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