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.
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.

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

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 LVR 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...
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 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...
(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.

ACKNOWLEDGEMENTS

We are grateful to Dr. Faouzi Mallat for his comments on the manuscript.

**********

Author Contributions
Malek Barka – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Khaled Ben Ahmed – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Amel Ben Abdallah – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Kalthoum Tlili – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Wissem Hmida – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Faouzi Mallat – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Malek Barka – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

REFERENCES

1. Cope C, Isard HJ. Left renal vein entrapment: A new diagnostic finding in retroperitoneal disease. Radiology 1969;92(4):867–2.
2. Barnes RW, Fleisher HL 3rd, Redman JF, Smith JW, Harshfield DL, Ferris EJ. Mesoaortic compression of the left renal vein (the so-called nutcracker syndrome): Repair by a new stenting procedure. J vasc surg 1986;8(4):415–21.
3. Radloff U, Holmes RJ, Prem JT, Faust GR, Moldwin R, Siegel D. Mesoaortic compression of the left renal vein (nutcracker syndrome): Case reports and review of the literature. Ann vasc surg 2006;20(1):120–9.
4. Shin JI, Park JM, Lee SM, et al. Factors affecting spontaneous resolution of hematuria in childhood nutcracker syndrome. Pediatr Nephrol 2005;20(5):609–13.
5. Shin JI, Lee JS, Kim MJ. The prevalence, physical characteristics and diagnosis of nutcracker syndrome. Eur J Vasc Endovasc Surg 2006;32(3):335–6.
6. EL-Sadr AR, Mina E. Anatomical and surgical aspects in the operative management of varicocele. The Urologic and Cutaneous Review 1950;54(5):257–62.
7. Takebayashi S, Ueki T, Ikeda N, Fujikawa A. Diagnosis of the nutcracker syndrome with Doppler sonography: Correlation with flow patterns in retrograde leftelefrenal venography. AJR Am J Roentgenol 1999;172(1):39–43.
8. Rogers A, Beech A, Braithwaite B. Transperitoneal laparoscopic left gonadal vein ligation can be the right treatment option for pelvic congestion symptoms.
secondary to nutcracker syndrome. vascular 2007;15(4):238–40.

9. Wendel RG, Crawford ED, Hehman KN. The “nutcracker” phenomenon: An unusual cause for renal varicosities with hematuria. J urol 1980;123(5):761–3.

10. Coolsaet BL. Ureteric pathology in relation to right and left gonadal veins. Urology 1978;12(1):40–9.

11. Shintaku N, Takahashi Y, Akaishi K, Sano A, Kuroda Y. Entrapment of left renal vein in children with orthostatic proteinuria. Pediatr Nephrol 1990;4(4):324–7.

12. Dever DP, Ginsburg ME, Millet DJ, Feinstein MJ, Cockett AT. Nutcracker phenomenon. Urology 1986;27(6):540–2.

13. Buschi AJ, Harrison RB, Norman A, et al. Distended left renal vein: CT/sonographic normal variant. AJR Am J Roentgenol 1980;135(2):339–42.

14. Kurklnsky AK, Rookc TW. Nutcracker Phenomenon and Nutcracker Syndrome. Mayo Clin Proc 2010;85(6):552–9.

15. Zerhouni EA, Siegelman SS, Walsh PC, White RI. Elevated pressure in the left renal vein in patients with varicoceles: preliminary observations. J Urol 1980;123(4):512–3.

16. Campbell-Walsh Urology, vol. 4 of Section XVII, 10th edition 2012.

17. Takemura T, Iwasa H, Yamamoto S, et al. Clinical and radiological features in four adolescents with nutcracker syndrome. Pediatr Nephrol 2000;14(10-11):1002–5.

18. Shin JI, Park JM, Lee JS, Kim MJ. Effect of renal Doppler ultrasound on the detection of nutcracker syndrome in children with hematuria. Eur J Pediatr 2007;166(5):399–404.

19. Martinez-Salamanca Garcia JI, Herranz Amo F, Gordillo Gutiérrez I, et al. Nutcracker’s syndrome: Demonstration with helicoidal TC with volumetric reconstruction “3D”. Actas Urol Esp 2004;28(7):549–52. [Article in Spanish].

20. Ahmed K, Sampath R, Khan MS. Current trends in the diagnosis and management of renal nutcracker syndrome: A review. Eur J Vasc Endovasc Surg 2006;31(4):410–6.

21. Zhang HK, Shen LG, Li M, et al. Diagnosis and treatment of left renal vein entrapment Syndrome. Chinese Journal of General Surgery 2001;16:511.

22. Neste MG, Narasimham DL, Belcher KK. Endovascular stent placement as a treatment for renal venous hypertension. J Vasc Interv Radiol 1996;7(6):859–61.

23. Marone EM, Psacharopulo D, Kahlberg A, Coppi G, Chiesa R. Surgical treatment of posterior nutcracker syndrome. J Vasc Surg 2011;54(3):844–7.

ABOUT THE AUTHORS

Article citation: Barka M, Mallat F, Hmida W, Chavey SO, Ahmed KB, Abdallah AB, Tlili K. Posterior nutcracker syndrome with left renal vein duplication as a cause of gross hematuria and recurrent left varicocele in an eight-year-old boy. Int J Case Rep Images 2014;5(8):572–575.

Malek Barka is Resident at Department of Visceral Surgery, University of Sousse School of Medicine. His area of interest include oncology and transplantation.
About Edorium Journals
Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission
We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?
In less than 10 words - we give you what no one does.

Vision of being the best
We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services
We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review
All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review
All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version
Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status
From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks
You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.

Four weeks
After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.

Mentored Review Articles (MRA)
Our academic program “Mentored Review Article” (MRA) gives you a unique opportunity to publish papers under mentorship of international faculty. These articles are published free of charges.

Most Favored Author program
Join this program and publish any number of articles free of charge for one to five years.

Favored Author program
One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program
Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence
We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...
We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

CONNECT WITH US

Edorium Journals: On Web
Browse Journals