Case Report

A Rare Cause of Recurrent Hematuria in Children: Nutcracker Syndrome

by Gurkan Genc,[1] Ozan Ozkaya,[1] Kenan Bek,[1] Yonca Acikgoz,[1] *and* Murat Danaci[2]
[1]*Pediatric Nephrology Department, Ondukuz Mayis University Faculty of Medicine, Samsun, Turkey*
[2]*Radiology Department, Ondukuz Mayis University Faculty of Medicine, Samsun, Turkey*

Correspondence: Gurkan Genc, Department of Pediatric Nephrology Ondokuz Mayis University Children Hospital, Kurupelit 55139, Samsun, Turkey. Tel.: +90 362 3121919/3754; E-mail: <gencdoc@yahoo.com>

Summary

Nutcracker syndrome (NCS), the left renal vein hypertension due to compression between aorta and superior mesenteric artery, may present with hematuria, flank pain and proteinuria. We report a 6-year-old girl with episodic macroscopic hematuria who was diagnosed as NCS with Doppler ultrasound and 3D computerized tomography angiography. She was managed conservatively with spontaneous resolution of macroscopic hematuria. With this case we would like to emphasize that in children with hematuria and orthostatic proteinuria without an apparent cause, a great index of suspicion and appropriate imaging is necessary for the diagnosis of NCS.

Key words: children, hematuria, Nutcracker syndrome.

Introduction

Macroscopic hematuria in children is a common cause of referral to hospital, with a long list of etiology including urinary tract infections, hypercalciuria, nephrolithiasis, glomerulonephritis, etc. In most cases, the diagnosis is easy with typical history, physical and laboratory examination. But extensive work-up may be required in rare cases. Nutcracker syndrome (NCS), which is the compression of left renal vein (LRV) between superior mesenteric artery (SMA) and aorta, is such a condition [1]. We present here a 6-year-old girl with episodic macroscopic hematuria who was diagnosed as NCS.

Case Report

A 6-year-old girl was admitted to the hospital with intermittent hematuria of 2 weeks. Physical examination, blood pressure (100/70 mmHg), biochemical tests including blood urea nitrogen, creatinine ($0.6\,mg\,dl^{-1}$), electrolytes, albumin, total protein and C-reactive protein and erythrocyte sedimentation rate were normal. Urinalysis revealed proteinuria ($30\,mg\,dl^{-1}$), hematuria (80% isomorphic) and leucocyturia. Prothrombin, partial thromboplastine and bleeding times and complement levels were normal. Antinuclear and antidouble-stranded DNA antibodies were negative. Daily protein excretion was $2.5\,mg\,m^{-2}\,h^{-1}$ and Ca excretion was $0.8\,mg\,kg^{-1}\,day^{-1}$ with a urine volume of $1360\,ml\,day^{-1}$. Urine culture was negative, and renal ultrasound was normal. Hematuria has resolved spontaneously in 1 week. Doppler ultrasound was planned but the patient did not come to her appointment.

Two years later, she was admitted to our hospital again with macroscopic hematuria lasting for a few days. It was learnt that she had macroscopic hematuria 1 year ago and applied to another hospital. Renal biopsy had been planned but the parents had not given consent. On her second admission to our hospital, her blood pressure was 100/70 mmHg with normal physical examination and biochemical tests (serum creatinine was $0.41\,mg\,dl^{-1}$). Urinalysis revealed a specific gravity of 1020, pH 6, negative nitrite and glucose $75\,mg\,dl^{-1}$ protein, 21 red blood cells/high power field (HPF), one to two white blood cells/HPF and a few hyaline casts (spot urinary protein/creatinine ratio being 1). Complement levels were normal. Antibodies to hepatitis B and C viruses were negative. Daily urine collection (total volume: 1220 ml) revealed $14\,mg\,m^{-2}\,h^{-1}$ proteinuria and $1.2\,mg\,kg^{-1}\,day^{-1}$ calciuria. Nocturnal and diurnal urinary protein excretions were $3.9\,mg\,m^{-2}\,h^{-1}$ and $34\,mg\,m^{-2}\,h^{-1}$, respectively. Renal ultrasound was normal and Doppler ultrasound revealed LRV dilatation up to the angle between SMA and aorta with a diameter of 7 mm on hilar region. This diameter was 1–2 mm between aorto-mesenteric vessels. Peak flow

© The Author [2009]. Published by Oxford University Press. All rights reserved. For Permissions, please email: journals.permissions@oxfordjournals.org

doi:10.1093/tropej/fmp121 Advance Access published on 10 December 2009

Downloaded from https://academic.oup.com/tropej/article-abstract/56/4/275/1651722 by Kocaeli University Library (KOU) user on 23 July 2020

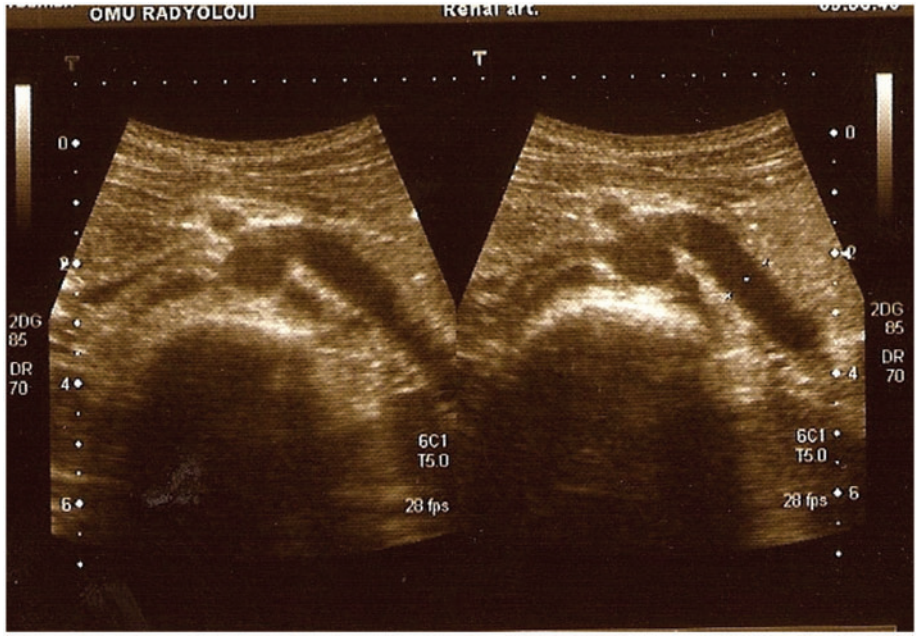


FIG. 1. Doppler ultrasound imaging showing narrowing LRV section.

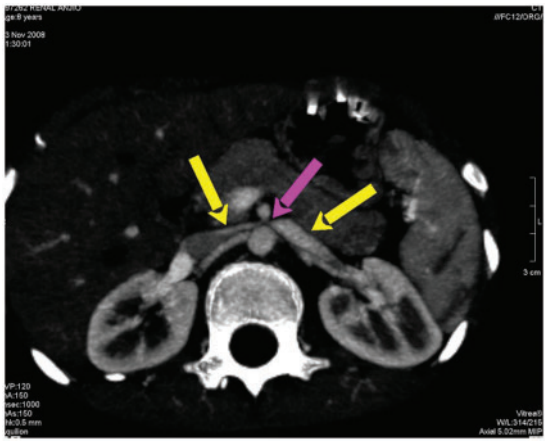


FIG. 2. CT angiography revealed narrowing LRV.

rate measured 11 cm s^{-1} in renal hilus, and 137 cm s^{-1} between aorta and SMA (Fig. 1). The diagnosis was confirmed by 3D computed tomography (CT) angiography by showing pre- and post-narrowing diameters of 6 mm and 1.4 mm, respectively (Figs 2 and 3).

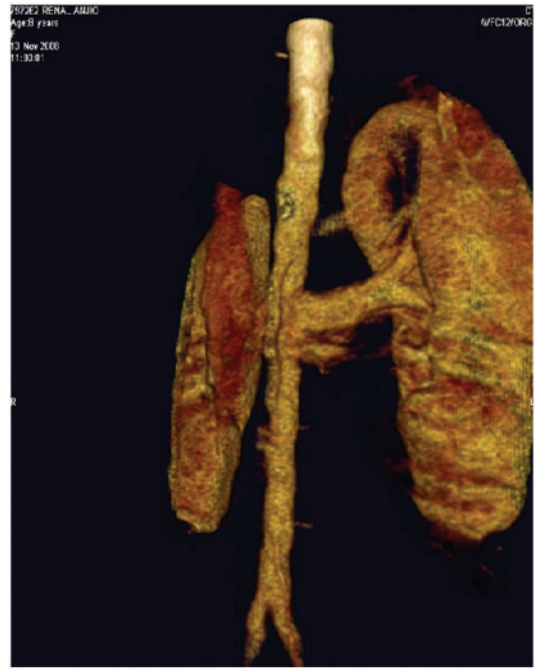


FIG. 3. 3D computerized tomography imaging of patient with narrowing LRV.

Downloaded from https://academic.oup.com/tropej/article-abstract/56/4/275/1651722 by Kocaeli University Library (KOU) user on 23 July 2020

Discussion

NCS is defined as compression of LRV between the aorta and SMA resulting in elevation of LRV pressure and collateral vein development. LRV hypertension is the most commonly pronounced pathogenetic mechanism. This pressure may cause rupture of thin-walled septa between collecting system and small veins causing hematuria. Glomerular basement membrane changes and mild immune injury are also approved as possible mechanisms [1, 2–6].

Symptoms of NCS include abdominal and left flank pain, lower limb varices, left varicocele, pelvic congestion syndrome (lower abdominal pain, dysmenorrhea, post-coital ache, dysuria, dyspareunia, pelvic, vulvar, gluteal or thigh varices), hematuria and orthostatic proteinuria [2–4]. Our patient had only macroscopic hematuria with no apparent cause at first evaluation where Doppler ultrasound could not be carried out due to parental incompliance. Upon a second similar episode, she applied to another hospital where renal biopsy was offered. NCS is an uncommon cause of hematuria with unremarkable history. Thus it is difficult to diagnose unless being suspected. In our patient, after excluding other causes, Doppler ultrasound was performed with suspicion of NCS and the diagnosis was confirmed with 3D CT angiography with a 2-year delay. The diagnosis of NCS depends on demonstration of compressed LRV with methods such as magnetic resonance imaging, Doppler ultrasound, CT angiography and venography. Previous studies suggest that initial method should be Doppler ultrasound and confirmation should be done with other imaging techniques, if required [7,8]. Peak velocity ratios between aortomesenteric and LRV hilar portion were used to diagnose NCS [7]. Suggested cut-off value is 4.8 for peak velocity ratio and that was 12.4 in our patient. Before using more invasive methods, 3D CT angiography is the preferred technique to diagnose vascular abnormalities [8]. Retrograde phlebography, video-angiography, reno-caval pressure gradient determination are accepted as the gold standard for the diagnosis of NCS. Renal biopsy is always normal if other renal diseases are not associated [9, 10]. Left-sided hematuria can be shown with cystoscopy.

Conservative approach, open surgical procedures, intra or extra vascular stents, intra-pelvic chemical cauterization have been proposed for the treatment. Patients with mild symptoms may be followed conservatively especially in puberty [6]. But severe pain, persistent macroscopic hematuria and progressive renal function impairment are potential surgical indications [10]. Surgical procedures include anterior nephropexy with excision of renal varicosities, LRV transposition, SMA transposition, renal auto-transplantation and gonado-caval bypass. Because of mild urinary symptoms, our patient was managed conservatively.

In conclusion, NCS is a rare condition the diagnosis of which is dependent on a high index of suspicion in children with recurrent hematuria and orthostatic proteinuria, especially when there is no apparent cause or clinical finding. Doppler ultrasound and CT angiography should be performed in such patients in order to avoid unnecessary renal biopsy.

References

1. de Schepper A. Nutcracker phenomenon of the renal vein and venous pathology of the left kidney. J Belge Radiol 1972;55:507–11.
2. Little AF, Lavoipierre AM. Unusual clinical manifestations of the nutcracker syndrome. Australas Radiol 2002;46:197–200.
3. Takemura T, Iwasa H, Yamamoto S, *et al*. Clinical and radiological features in four adolescents with nutcracker syndrome. Pediatr Nephrol 2000;14:1002–5.
4. Scultetus AH, Villavicencio LL, Gillespie DL. The nutcracker syndrome: its role in the pelvic venous disorders. J Vasc Surg 2001;34:812–19.
5. 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;31:410–16.
6. Tanaka H, Waga S. Spontaneous remission of persistent severe haematuria in an adolescent with nutcracker syndrome: seven years' observation. Clin Exp Nephrol 2004;8:68–70.
7. Shin IS, park JM, Lee JS, Kim MJ. Doppler ultrasonographic indices in diagnosing nutcracker syndrome in children. Pediatr Nephrol 2007;22:409–13.
8. Kaneko K, Kiya K, Nishimura K, *et al*. Nutcracker phenomenon demonstrated by three-dimensional computed tomography. Pediatr Nephrol 2001;16:745–7.
9. Ozono Y, Harada T, Namie S, *et al*. The nutcracker phenomenon with IgA nephropathy. J Intern Med Res 1995;23:126–31.
10. Zhang H, Li M, Jin W, *et al*. The left renal entrapment syndrome: diagnosis and treatment. Ann Vasc Surg 2007;21:198–203.

Downloaded from https://academic.oup.com/tropej/article-abstract/56/4/275/1651722 by Kocaeli University Library (KOU) user on 23 July 2020