

Nutcracker Syndrome with Hypertension as an Unusual Initial Presentation

Abstract

Hypertension has been rarely reported in patients with the nutcracker phenomenon (NCP)/syndrome. We describe a case of a young adolescent female patient where a computed tomography angiography (CTA) provided evidence of left renal vein dilatation, probably due to its compression through the angle between the aorta and the superior mesenteric artery, during the evaluation of secondary hypertension. Blood levels of renin and aldosterone were within normal limits. Ultrasonography of the kidney showed minimal scarring on the left side. As there were no other signs of secondary hypertension, we proceeded with a CTA, which revealed findings compatible with the so-called NCP/syndrome.

Keywords: Adolescent, hypertension, nutcracker syndrome

**N. A. Narkhede,
A. B. Deokar,
K. P. Mehta,
N. N. Kamat**

*Department of Pediatrics
and Radiology, Nanavati
Super Speciality Hospital,
Vileparle (W), Mumbai,
Maharashtra, India*

Introduction

Nutcracker phenomenon (NCP) refers to compression of the left renal vein (LRV), most commonly between the aorta and the superior mesenteric artery, with impaired blood outflow often accompanied by distention of the distal portion of the vein. The nutcracker syndrome (NCS) is the clinical equivalent of NCP characterized by a complex of symptoms with substantial variations. When symptomatic, this syndrome is manifested by left flank and abdominal pain, with or without macroscopic or microscopic hematuria. Loin pain and hematuria are common symptoms, presenting in patients of all ages. There are various underlying pathologic causes, including renal or ureteric calculi or tumors, intrinsic kidney disease (including glomerulonephritis), and loin pain hematuria syndrome. Patients therefore often undergo multiple investigations and procedures. However, in some cases, no cause is identified unless rarer pathologic states are considered. We describe a case of a young female with hypertension, loin pain, hematuria, and multiple seizure episodes, where an extensive clinico-laboratorial investigation revealed that she also presented elements of the NCP.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Case Report

A 17-year-old female presenting with complaints of repeated episodes of convulsions was admitted for further workup. The patient also had a history of headache, gross hematuria, and loin pain intermittently for the past 6 months. Loin pain was associated with left loin tenderness for the past 4 months. The loin pain used to be so severe that she had to lie down in the bed for 3–4 h. The patient had a history of hypertension intermittently for the past 4 years. Intermittently, she was on antihypertensive (tablet amlodipine and losartan), which was stopped for the past 1½ years. She was investigated for hypertension 3 years back. Pressure track ambulatory blood pressure (BP) record at that time showed poor BP control with normal diurnal variation (85% of all systolic BP and 36% of all diastolic BP exceeded threshold of 135/85 mmHg while awake and 120/70 mmHg while sleep). Blood levels of renin and aldosterone were within normal limits. Twenty-four-hour urine collections did not reveal any proteinuria. Urinary tests for pheochromocytoma were also normal. Immunological investigations including anti-nuclear antibodies, complement (C3/C4), and serum immunoglobulin levels were within normal limits. Electroencephalogram and magnetic resonance imaging (MRI) of the brain were normal.

How to cite this article: Narkhede NA, Deokar AB, Mehta KP, Kamat NN. Nutcracker syndrome with hypertension as an unusual initial presentation. *Indian J Nephrol* 2017;27:472-4.

Address for correspondence:

*Dr. N. A. Narkhede,
Room No. 25, Doctors
Quarters, Nanavati Super
Speciality Hospital,
S. V. Road, Vileparle (W),
Mumbai, Maharashtra, India.
E-mail: nilimanarkhede12@
gmail.com*

Access this article online

Website: www.indianj nephrol.org

DOI: 10.4103/ijn.IJN_184_16

Quick Response Code:



In the past from 8 to 12 years of age, she had a history of recurrent urinary tract infection, which once required hospitalization. Her father had a history of hypertension that was diagnosed at the age of 29 years. Paternal uncle and younger brother had a history of Type 1 diabetes mellitus. Physical examination was remarkable for left renal angle tenderness with BP recorded to be normal.

Before admission, the patient had undergone radiological examination including a renal ultrasound, which showed both kidneys normal in size and shape with minimal scarring in the left kidney. Subsequent repeat Dimercaptosuccinic acid scan was normal. Doppler renal arteries' scan was performed to rule out renal artery stenosis, which showed borderline renal artery resistive index values of 0.68 on the right side and 0.7 on the left side. Venous parameters were normal. Hence, computed tomography angiography (CTA) was planned. CTA had revealed signs of LRV stenosis and dilation [Figure 1].

The patient underwent LRV transposition and she is currently under close follow-up. No hypertension was reported on follow-up visits. Her clinical signs and symptoms have improved drastically since the surgery.

Discussion

The terms “NCS” and “NCP” are frequently used interchangeably in the literature referring to the known abnormal anatomy of the compression of the LRV, as it passes through the angle between the aorta and the superior mesenteric artery. Although the first clinical report was made by El-Sadr and Mina in 1950, the term “nutcracker” is credited to de Schepper in 1972,^[1] but was first used by Chait *et al.* in 1971.^[2] It usually affects women more than men and is mostly present in the third or fourth decades of life. Hypertension has not been included in the traditional clinical manifestations of the syndrome that is usually

accompanied with abdominal pain, hematuria, orthostatic proteinuria, chronic fatigue, pelvic congestion in females, and left-sided varicocele formation in males.^[3,4]

Doppler ultrasonography is a helpful, noninvasive modality and should be the first assessment after NCS is suspected clinically or when a large LRV diameter ratio is noted between its distended and narrowed portions ($D_{D/N}$) on computed tomography (CT) or MRI. Other imaging modalities that are used include renal angiography, CTA, digital subtraction angiography, standard MRI, and magnetic resonance angiography. Renal angiography shows delayed venous washout from the left kidney. Venography (retrograde phlebography) is not commonly performed in patients who do not have severe symptoms, especially in children. Although knowledge of renocaval pressure gradient is extremely helpful, there is some overlap between pressures seen in healthy individuals and patients with NCS.^[5]

The renal NCS management has evolved in the past four decades^[6-8] and there are several available options, from close expectant surveillance, endoscopic proceedings (e.g., external or internal stenting and hemostatic agents) to more complex open surgical procedures (e.g., LRV transposition, renal autotransplantation of the left kidney). Patients in prepubertal age should be offered less aggressive forms of treatment since the likelihood of spontaneous remission is due to normal physical development.^[3,9]

Acknowledgment

We acknowledge the support of Dr. Paras Kothari, Pediatric Surgeon at Nanavati Super Speciality Hospital, Mumbai, for the left renal vein transposition surgery.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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. Chait A, Matasar KW, Fabian CE, Mellins HZ. Vascular impressions on the ureters. *Am J Roentgenol Radium Ther Nucl Med.* 1971 Apr;111(4):729-49.
3. Reed NR, Kalra M, Bower TC, Vrtiska TJ, Ricotta JJ 2nd, Gloviczki P. Left renal vein transposition for nutcracker syndrome. *J Vasc Surg* 2009;49:386-93.
4. Mazarakis A, Almpanis G, Tragotsalou N, Karnabatidis D, Fourtounas C. Is hypertension a manifestation of the nutcracker phenomenon/syndrome? Case report and brief review of the literature. *Hippokratia* 2012;16:187-9.
5. Hohenfellner M, D’Elia G, Hampel C, Dahms S, Thüroff JW. Transposition of the left renal vein for treatment of the nutcracker

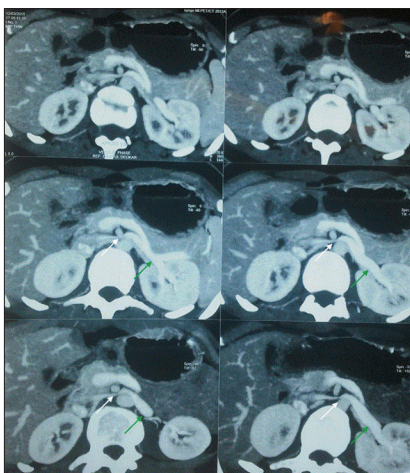


Figure 1: Axial Computed tomographic images with intravenous contrast in portal venous phase showing nutcracker phenomenon. Left renal vein is compressed between the aorta and the superior mesenteric artery (white arrow). Left renal vein with distended hilar portion (green arrow)

- phenomenon: Long-term follow-up. *Urology* 2002;59:354-7.
6. Kurklinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. *Mayo Clin Proc* 2010;85:552-9.
 7. Gulleroglu K, Gulleroglu B, Baskin E. Nutcracker syndrome. *World J Nephrol* 2014;3:277-81.
 8. Ullery BW, Itoga NK, Mell MW. Transposition of the left renal vein for the treatment of nutcracker syndrome in children: A short-term experience. *Ann Vasc Surg* 2014;28:1938.e5-8.
 9. Preza Fernandes J, Amorim R, Gomes MJ, Oliveira V, Reis A, Ribeiro-Castro J. Posterior nutcracker syndrome with left renal vein duplication: A rare cause of haematuria in a 12-year-old boy. *Case Rep Urol* 2012;2012:849681.