

Circumaortic Renal Vein Compression Causing Nutcracker Syndrome with Pelvic Congestion: A Case Report

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Radiology Case. 2026 July; 20(7):1-6 :: DOI: 10.3941/jrcr.6081

AUTHORS CONTRIBUTION

Bayramov Rasim: clinical data acquisition, supervision, editing, author

Bayramli Farid: clinical data review, manuscript drafting, writing, corresponding author.

DISCLOSURES

The authors declare no conflict of interest.

HUMAN AND ANIMAL RIGHTS

No human and animal experiments were conducted in this study

CONSENT

Yes

ABSTRACT

A 32-year-old female patient with fever, cold extremities, abdominal and flank pain, infrequent urination and hypertension. Laboratory tests, ultrasound and computed tomography examinations were carried out to investigate the case. During computed tomography angiography scan circumaortic left renal vein with entrapment of anterior and posterior branches was revealed. The patient opted for conservative management and is currently under continuous medical observation. This case report emphasizes the necessity of imaging modalities in assessment of this rare entity.

CASE REPORT

BACKGROUND

Renal vascular malformations are a rare group of anatomical abnormalities, associated with unexplained clinical symptoms. We report a rare case of combined nutcracker syndrome with left renal vein branches entrapment. This entity has only been described six times in medical literature previously. Review of medical literature indicates the link between circumaortic renal vein compression and a variety of non-specific clinical symptoms. Therefore, the use of modern imaging techniques is essential for the diagnosis and follow-up management of this disorder. Our study also aims to increase clinical awareness among physicians regarding the diagnosis and treatment of this condition and its manifestations.

CASE REPORT

A 32-year-old female presented with cramping abdominal and flank pain and infrequent urination (two times per day). On physical examination, she weighed 55 kg with a BMI of 23.5. The patient also reported tachycardia (>100 bpm) and episodes of elevated systolic and diastolic blood pressure, often accompanied by facial flushing. Her hypertension was resistant

to treatment and poorly controlled. Fever and cold extremities had been present for the previous two to three days.

Prior to CT, the patient underwent a series of clinical and laboratory examinations, which were inconclusive. Abdominal ultrasonography demonstrated normal kidney size and echogenicity without detectable anatomical abnormalities. A comprehensive metabolic panel showed normal serum creatinine and urea levels, with a mildly elevated serum albumin level (57.2 µmol/L).

During CT scan with computer tomographic angiography (CTA) circumaortic left renal vein (LRV) was revealed (Figure 1). The aortomesenteric angle and aortomesenteric distance were found to be 11° and 6 mm respectively, leading to the compression of anterior branch between aorta and SMA. The posterior branch was also entrapped between the aorta and the L2 vertebra, with a remaining aortovertebral space of 3 mm (Figure 2). Thereby pelvic congestion syndrome, consistent with venous drainage insufficiency resulting in varicose dilatation of left ovarian vein and varices in the left pelvic region

was a salient finding (Figure 3,4). CT intravenous urography demonstrated no further abnormalities of the urinary system, and the remaining abdominal structures were unremarkable. Consequently, a diagnosis of combined nutcracker syndrome was established.

Management and follow up

After multidisciplinary consultation and discussion of treatment options, the patient opted for conservative management.

Follow up

The patient is currently under continuous medical observation

DISCUSSION

Introduction

Nutcracker syndrome (NS) is an extrinsic venous entrapment disorder, typically involving compression of the left renal vein between the superior mesenteric artery (SMA) and the aorta; however, other anatomical variations have also been described in the literature [1,2]. The term *nutcracker* originates from Dr. Schepper due to the resemblance of the compressing vessels to the jaws of a nutcracker crushing a nut [3].

Two classic forms of nutcracker syndrome have been described in the literature [2]. Anterior nutcracker syndrome refers to the typical compression of the left renal vein between the SMA and the aorta, whereas posterior nutcracker syndrome involves compression of a retroaortic anatomical variant of the left renal vein between the aorta and the lumbar vertebrae (L1 or L2) [4].

Combined nutcracker syndrome is a rare condition caused by the compression of a circumaortic left renal vein, where an accessory retroaortic renal vein exists alongside the normally positioned anterior renal vein, resulting in venous compression both anteriorly (between the SMA and aorta) and posteriorly (between the aorta and vertebrae) [5].

Etiology & Demographics

Renal venous system abnormalities are caused during embryological development [6]. Fetal kidneys are drained by both ventral and dorsal renal veins, with the latter usually disappearing during development [5]. The overall incidence of a retroaortic renal vein is approximately 3% and circumaortic renal veins is about 3.5% of individuals, most often as incidental CT findings [7]. A circumaortic renal vein is characterized by two LRV passing anteriorly and posteriorly to the aorta and predisposing venous entrapment known as combined nutcracker syndrome. Combined Nutcracker Syndrome is a rare condition, described previously only six times in the medical literature.

Epidemiologically NS is diagnosed mostly in women in their 3rd and 4th decade of life with lower than 18,5 BMI [5]; in

our case the patient is a 32-year-old-female within normal BMI range. Low BMI is identified as a significant risk factor for NS [2].

Clinical and Imaging findings

The primary symptoms of combined nutcracker syndrome are micro- or macrohematuria, proteinuria, flank pain, pelvic pain, dysuria and gonadal varices [8]. Apart from these presented in our patient elevated systolic and diastolic pressure, indirectly associated with the renal venous congestion syndrome and is an uncommon, but well documented manifestation [9]. Clinically NS is a diagnosis of exclusion [2]. Clinical manifestations are caused primarily from increased left renal vein pressure and corresponding pelvic congestion [1].

Radiographic features are similar on different imaging modalities. Retrograde venography is considered as the gold standard but is an invasive procedure [10]. Doppler US provides valuable hemodynamic assessment of the LRV at the compression site [11]. In most cases CTA is performed and can demonstrate vein narrowing, pelvic and gonadal varicosities, and allow measurement of the aortomesenteric angle and distance (normal ranges: 38–65° and 10–34 mm, respectively), both of which were significantly reduced in this patient. MRI in the venous phase offers similar diagnostic value [5].

Treatment and prognosis

There is a wide range of treatment options for NS, ranging from conservative management to endovascular and reconstructive surgeries. The appropriate course of action depends on various factors, such as presence of hematuria, age groups and severity of congestion symptoms [4]. Interventional procedures include primarily external or internal stenting. Open surgical procedures like transposition of LRV are novel options for a pediatric age group [12]. Renal autotransplantation, gonadocaval bypass or even nephrectomy could be carried out during end-stage organ damage [13]. Given the mildness of symptoms, presence of arterial hypertension and patient's convenience a conservative treatment with constant observation was chosen as the most appropriate management.

TEACHING POINT

Combined Nutcracker Syndrome is a rare clinical entity causing a range of non-specific symptoms and is therefore difficult to diagnose without the use of imaging techniques. Doppler US and CTA are considered most suitable diagnostic modalities and should be performed in cases of unexplained hematuria, abdominal and flank pain, gonadal varices and as demonstrated in our case elevated pressure. The primary therapeutic approach is conservative management, invasive and surgical procedures being possible in more severe and end-stage cases. This case report emphasizes the necessity of imaging modalities in assessment of this pathology and preventing of false negatives, making it possible to undertake appropriate management.

QUESTIONS

Question 1: What anatomical structure involved in Nutcracker Syndrome?

1. Compression of left renal vein (applies)
2. Compression of 3rd part of duodenum
3. Compression of left renal artery
4. Compression of left ureter
5. Compression of right renal vein

Explanation: Nutcracker Syndrome is compression of left renal vein between aorta and SMA [Nutcracker syndrome (NS) is an extrinsic venous entrapment disorder, typically involving compression of the left renal vein between the superior mesenteric artery (SMA) and the aorta]

Question 2: What is the etiology of Nutcracker Syndrome?

1. Embryologic (applies)
2. Iatrogenic
3. Infectious
4. Toxins
5. Idiopathic

Explanation: Nutcracker Syndrome is embryological malformation [Renal venous system abnormalities are caused during embryological development]

Question 3: What is the gold standard imaging modality for Nutcracker Syndrome?

1. Retrograde venography (applies)
2. CTA
3. MRI
4. Doppler US
5. Native CT

Explanation: Retrograde venography is gold standard for NS [Retrograde venography is considered as the gold standard but is an invasive procedure]

Question 4: What is not a common clinical manifestation of Nutcracker Syndrome?

1. Right sided varicocele (applies)
2. Macrohematuria
3. Flank pain
4. Left sided pelvic congestion
5. Microhematuria

Explanation: Right sided varicocele is not common due to compression of left renal vein [Combined nutcracker syndrome is a rare condition caused by the compression of a circumaortic left renal vein]

Question 5: What is a risk factor for Nutcracker Syndrome?

1. Low BMI (<18.5) (applies)
2. Senior males
3. Diabetes mellitus
4. Children under age of 5
5. None of these

Explanation: Low BMI is associated with NS [Epidemiologically NS is diagnosed mostly in women in their 3rd and 4th decade of life with lower than 18,5 BMI]

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FIGURES

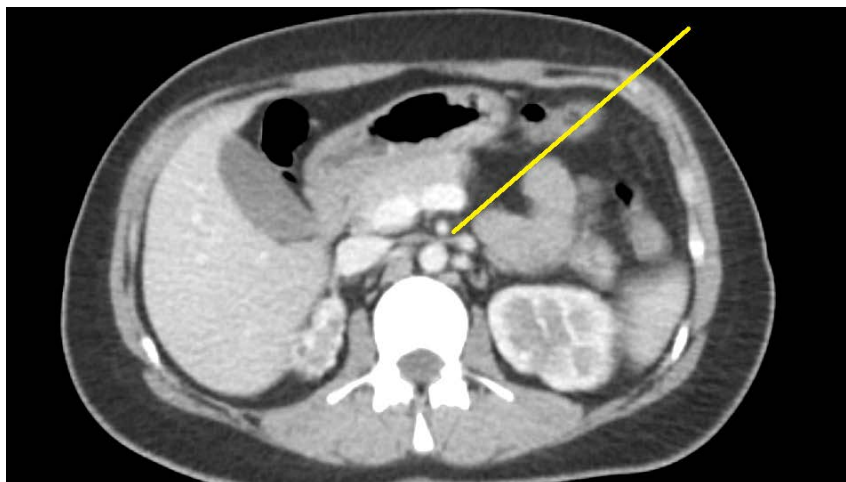


Figure 1: A 32-year-old female with Combined Nutcracker Syndrome: Coronal contrast enhanced CT images in venous phase showing entrapment of anterior branch of renal vein between SMA and aorta.

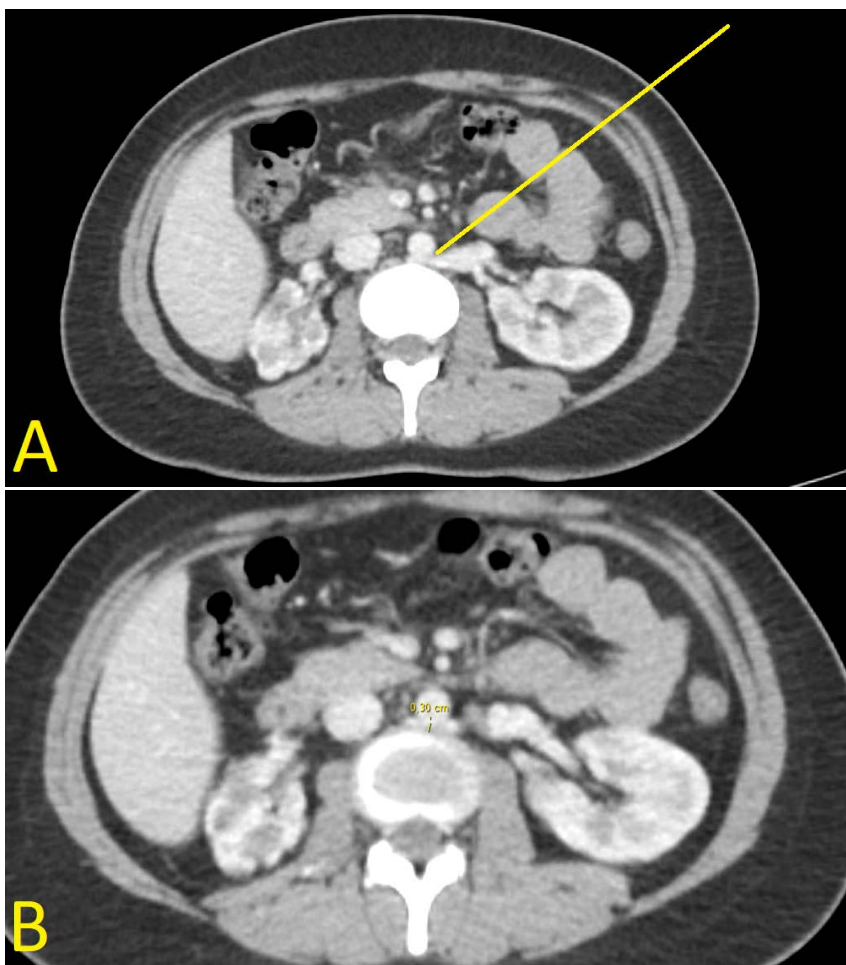


Figure 2: A 32-year-old female with Combined Nutcracker Syndrome:(A) and (B) Coronal contrast enhanced CT images in venous phase showing entrapment and distance of posterior branch of renal vein between aorta and L2.

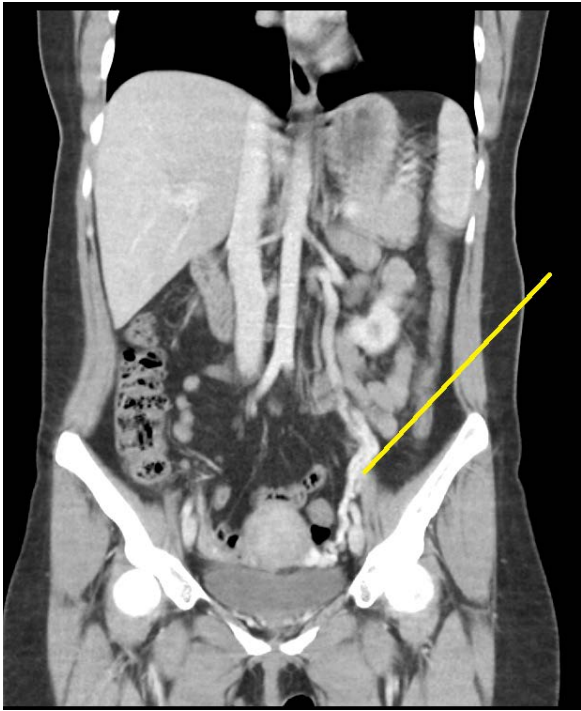


Figure 3: A 32-year-old female with Combined Nutcracker Syndrome: Coronal contrast enhanced CT images in venous phase demonstrating left ovarian vein varices and pelvic congestion.

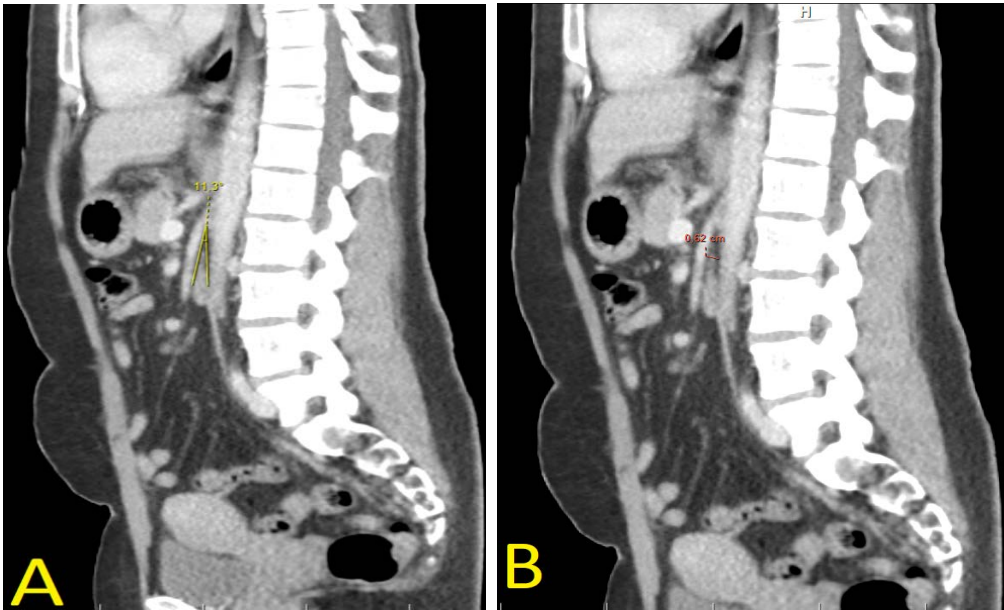


Figure 4: A 32-year-old female with Combined Nutcracker Syndrome: (A) and (B) Saggital contrast enhanced CT images in venous phase demonstrating measurements of aortomesenteric angle and distance.

SUMMARY TABLE

Etiology	Development malformation
Incidence	3.5% of individuals, mostly incidental finding
Gender Ratio	Most common if females
Age predilection	Most common in 3 rd and 4 th decade
Risk factors	Low MBI
Treatment	Symptomatic management and reconstructive surgery
Prognosis	Controversial; depends on risk factors and treatment options
Imaging Findings	Entrapment of anterior and posterior branches of LRV between SMA, aorta and L2 in venous phase CT scan

KEYWORDS

Combined nutcracker syndrome; renal development abnormalities; venous phase contrast CT; pelvic congestion; urological pathologies

ABBREVIATIONS

US = Ultra Sound
CT = Computed Tomography
CTA = Computed Tomography Angiography
MRI = Magnetic Resonance Imaging
NS = Nutcracker Syndrome
BMI = Body Mass Index
SMA = Superior Mesenteric Artery
LRV = Left Renal Vein

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