

Nutcracker Syndrome as Unusual Cause of Hematuria in a young woman: A Case Report

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Abstract

Case Report

Nutcracker syndrome is an uncommon vascular compression disorder that is characterized by the compression of left renal vein between the abdominal aorta and superior mesenteric artery. It tends to occur in female patients with a predilection of third and fourth decades. Symptoms are non-specific and diagnosis is possible with exclusion of more common causes and, with imaging studies. The diagnosis can be made with Doppler ultrasound, computed tomography angiography, magnetic resonance angiography and retrograd venography. We present a case with this rare syndrome diagnosed with computed tomography angiography. This entity can be easily overlooked unless it comes to mind and dedicated imaging is performed.

Key words: Nutcracker syndrome, computed tomography angiography, hematuria.

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INTRODUCTION

Hematuria may be a hint of an underlying disease, some of which are easily diagnosed and rest of them are required extensive workup. The causes vary according to age and gender, with the most common ones are inflammation/infection of the bladder or prostate, urinary stones, and, in elderly patients, urinary system malignancy or benign prostatic hyperplasia [1].

Nutcracker syndrome (NS) also known as left renal vein entrapment syndrome, is characterized by compression of the left renal vein between the abdominal aorta and superior mesenteric artery (SMA). This entity mostly is a rare cause of hematuria and presents with hematuria and pain although it may be asymptomatic [2]. Diagnosis is frequently delayed or missed, due to different manifestations of the syndrome and lack of consensus on diagnostic criteria [3].

CASE REPORT

A 28-year-old woman was referred to our clinic from another university hospital, with a history of intermittent gross hematuria and left flank pain of several months. She had no history of trauma or sexually transmitted diseases and there was no associated dysuria or fever. Physical examination was normal. She had low body mass index (17.6 kg/m²). Urinalysis confirmed 3+ blood cells without crystals or

microorganisms. Ultrasound and cystoscopy could not detect any renal, ureteral, or bladder anomaly. Family and past medical histories were unremarkable. Complete blood count, routine biochemical tests, complement serology, anti-GBM antibodies, and bleeding parameters were within normal limits. Computed tomography angiography (CTA) revealed the external compression of left renal vein by the abdominal aorta and SMA (Figure 1, 2). Considering patient's symptoms and radiological findings, NS was primarily assessed as the final diagnosis. The patient is given a diet to increase the abdominal fat volume.

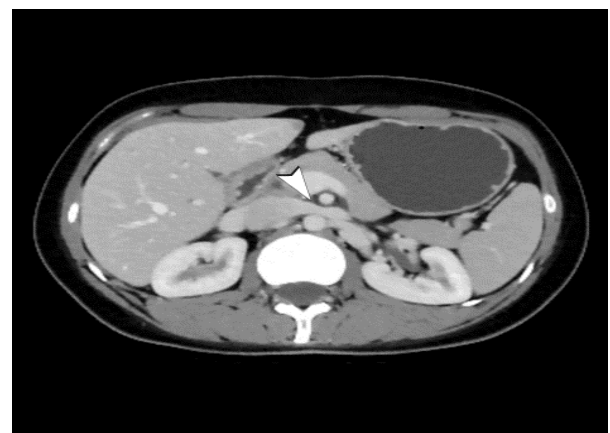


Fig-1: On contrast enhanced axial CT image, narrowed left renal vein between abdominal aorta and superior mesenteric artery was seen (white arrowhead)

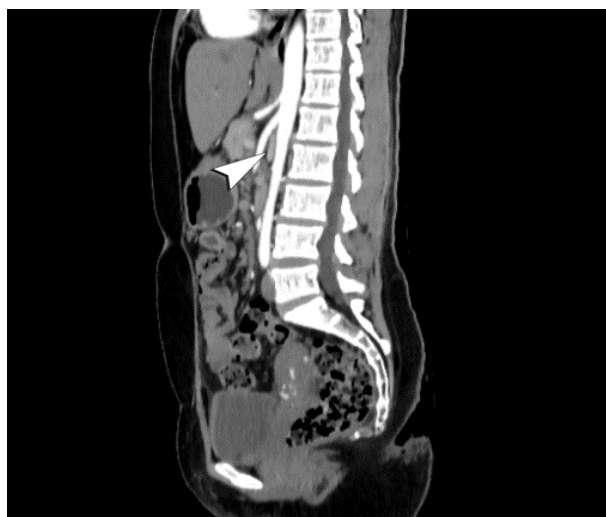


Fig-2: On contrast enhanced reconstructed sagittal CT image, decreased angle between abdominal aorta and superior mesenteric artery and compressed left renal vein between them (white arrowhead) were seen

DISCUSSION

Left renal vein entrapment syndrome, described as the compression of the left renal vein between the superior mesenteric artery and abdominal aorta was firstly mentioned in 1950 by El Sadr and Mina [4]. Chait et al. defined the abdominal aorta and SMA as the two components of a ‘nutcracker’ that may compress and narrow the left renal vein [5]. This term is credited by De Schepper denominating this phenomenon as NS [6].

The term “nutcracker syndrome” should not be used as an alternative to the term “nutcracker phenomenon” which is elementally the compression of the aortomesenteric portion of the left renal vein. This phenomenon is not link with clinical symptoms all the time and such anatomic findings that are reminder of nutcracker may exemplify a normal variant or be associated with other conditions. Hence, the term “nutcracker syndrome” should only be used for patients showing particular clinical symptoms connected to morphologic findings belong to nutcracker phenomenon [2]. A type of nutcracker syndrome occurred when retro-aortic left renal vein may be compressed between the abdominal aorta and vertebral column [7, 8].

Nutcracker phenomenon is often found incidentally in patients undergoing medical imaging for other purposes. NS is more frequently seen in young women with a predilection in third and fourth decades and our patient was 28 years old. This syndrome is typically seen in slim patients due to the narrowed angle between SMA and abdominal aorta (normal values ranging 38°–65°) and a decrease in the distance between the abdominal aorta and SMA (normal values ranging 10–14 mm) probably due to decrease in the volume of retroperitoneal and mesenteric fat tissue [9]. In our case, the patient was a slender woman and the

angle and the distance between abdominal aorta and SMA were measured on CTA as 25° and 9 mm, respectively.

This syndrome is associated with varying spectrum of symptoms and findings from microscopic hematuria to progressive pelvic congestion, and, may be accompanied by left flank pain, gonadal vein syndrome and varicocele. Proteinuria may occur to some extent. Symptoms are frequently exacerbated with physical activity [2,10]. Hematuria is the most common symptom and mainly caused by rupture of intrarenal vessels owing to increased venous pressure, into the collecting system [11]. Varicoceles may develop secondary to elevated pressure in left renal vein which is regarded by Zerhouni et al. as the main reason for occurrence of varicoceles in NS [13]. However, this findings was not found in our case.

It is obvious that the definitive diagnosis can not be reached without dedicated imaging studies. Making the diagnosis is also difficult due to ambiguous complaints attributable to a number of more common etiologies, like urinary stones [14]. To rule out more common renal conditions presents with hematuria and flank pain, various diagnostic studies can be performed such as routine blood tests, urinalysis, urine culture, cytology, urethrocytostcopy, intravenous pyelography, and renal biopsy [2].

Appropriate imaging modalities include CTA, Doppler ultrasound, magnetic resonance angiography and renal angiography. Intravenous pyelography may be used to show notching of the renal pelvis and ureters caused by varicosities. Cystoscopy may be show the bleeding from the left ureter [12]. Doppler ultrasound can provide additional information for the diagnosis, such as defining the dilated left renal vein and possible flow abnormalities [10]. In addition, magnetic resonance angiography may be used as a noninvasive option to demonstrate the narrow and dilated portions of left renal vein.

In our case, CTA revealed the distended portion proximal to the narrowed left renal vein between abdominal aorta and SMA. In the light of the symptoms of the patient and absence of more common etiologies, the diagnosis is established as NS. Conservative management is preferred and a diet plan is prepared to help patient to gain weight. Interventional radiology techniques using endovascular stents or surgery are the other treatment options for the patients who were not get benefit from the conservative treatment or with more severe and frequent symptoms [15].

CONCLUSION

It is important to be familiar with the diagnostic approach for NS, considering the high possibility of missing the diagnosis even in

symptomatic patients. Renal vein thrombosis and kidney injury might occur, if left untreated. The physicians should remind this syndrome, especially in the patients with hematuria of unknown origin and other suggestive symptoms when other routine diagnostic studies are inconclusive. Additionally, the radiologists should be familiar with the findings of this rare syndrome.

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