Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Report

Nut-cracker Phenomenon Masquerading as Loin Pain Micro-hematuria Syndrome: Anatomical and Urological Perspectives

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Abstract

Nutcracker syndrome (NCS) is a rare congenital anomaly, where the left renal vein (LRV) is sandwiched between two structures. It is defined as anterior or posterior NCS (PNCS), depending on whether the course of the LRV is anterior or posterior to descending aorta. We report a rare case of a 40-year-old lady, who presented to us with left lumbar pain, nausea, vomiting, and dyspepsia. On initial clinical evaluation, based on her diabetic status, biochemical findings of an elevated absolute leukocyte count and microscopic hematuria with an ultrasonography finding of cystitis with bilateral increased renal cortical echogenicity, a provisional diagnosis of the left acute pyelonephritis was made. On further evaluation with axial imaging, she was diagnosed to have a retro-aortic LRV, where the LRV is compressed between the aorta and vertebral body. Correlating her clinical and radiological findings, a diagnosis of PNCS was made. She was symptomatically better with antibiotics and anti-inflammatory drugs and was advised a regular follow-up. This manuscript emphasizes the need for a higher index of clinical suspicion to make a prompt diagnosis of such rare congenital venous anomalies that present with vague gastrointestinal symptoms or mimic pyelonephritis. The majority of such patients are conservatively managed. Those with intractable symptoms might need surgical intervention.

Key words: Kidney, Left renal vein, Microhematuria, Nutcracker, Pyelonephritis

INTRODUCTION

Nutcracker syndrome (NCS) is an uncommon clinical condition caused by mechanical compression of the left renal vein (LRV).[1] This condition is caused by compression of the LRV, either between the Superior Mesenteric artery and abdominal aorta Anterior NCS (ANCS) or between the vertebral column and the abdominal aorta Posterior NCS (PNCS).[2] The NCS must be distinguished from the nutcracker phenomenon. This phenomenon is a common anatomical abnormality, which is asymptomatic and diagnosed in routinely performed abdominal imaging. It usually affects women more than men and, in most cases, present in the 3rd or 4th decades of life.[3]

Access this article online

www.ijss-sn.com

Month of Submission : 11-2022
Month of Peer Review : 12-2022
Month of Acceptance : 12-2022
Month of Publishing : 01-2023

Anatomical Perspectives

The ANCS occurs when the LRV is sandwiched between the abdominal aorta and the origin of the superior mesenteric artery. Figure 1 illustrates the anatomical course of the LRV in ANCS. When the LRV gets compressed between these two arteries, there is a mechanical impediment to the return of blood from the renal vein, resulting in congestion of the left kidney.

On the other hand, PNCS occurs when the LRV gets compressed between the aorta and vertebral body.[4] Usually, the LRV runs anterior to the aorta, but in PNCS, the LRV runs behind the aorta, getting compressed from behind by the vertebral body. The course of this vein is called a retro-aortic LRV (RLRV). Figure 2 illustrates the

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PNCS e, with the yellow arrow pointing at the course of the LRV.

Due to mechanical compression on LRV, a renocaval pressure gradient is created which is the fundamental reason for all symptoms associated with PNCS. Venostasis or distension of the left ovarian or spermatic vein caused by LRV entrapment often results in loin pain, which may lead to confusion in making a proper diagnosis.[6] As PNCS is often under-reported, its clinical manifestations closely mimic and are often mistaken as presentations of other more common illnesses.[7]

Urological Perspectives

Loin pain and hematuria have always been perplexing problems for urologists since time immemorial. Most patients with PNCS present with vague loin pain or upper abdominal discomfort. In the absence of overt hematuria, most of these patients seek gastroenterologists' consultation. The presence of either micro or macroscopic hematuria adds to the confusion for urologists. Various authors have described renal cysts, angiomas, micro-hemangiomas, vascular anomalies, and renal veno-calyceal fistulae in the renal fornices as potential causes for this condition.[8] The lack of specific clinical findings necessitate a higher suspicion of PNCS. Non-invasive computed tomography (CT) angiogram imaging may be needed for the confirmation of PNCS.[9] This manuscript discusses an interesting case of PNCS associated with dyspepsia, vomiting, left loin pain, and microhematuria. The purpose of this manuscript is to highlight the myriad of symptoms that the PNCS may present with and also to give a clinico-anatomical overview of this underlying problem.

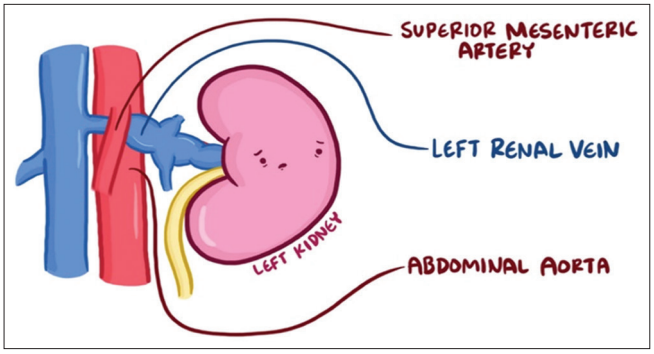

Figure 1: Diagrammatic illustration of the anterior nutcracker syndrome

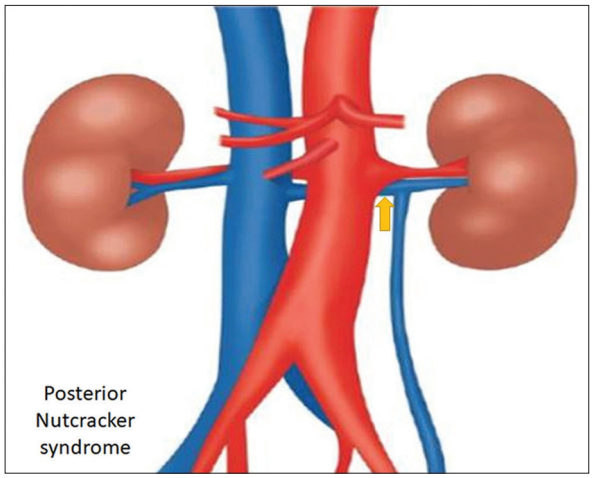

Figure 2: Diagrammatic illustration of Posterior NCS (reproduced with permission from Jang *et al.*[5])

CASE REPORT

A 40-year-old female presented with abdominal pain for the past 5 months. The pain was mainly in the left upper back. The left lumbar pain was non-radiating, dull aching, and relieved on analgesic intake. She also had epigastric pain that was burning in nature, associated with vomiting and not related to food intake. There was no abdominal distension and her bowel movements were normal. She had no macro-hematuria, dysuria or decreased urine output. Her appetite was normal. She was a known diabetic and on regular oral hypoglycemic agents. Physical examination was unremarkable.

On evaluation, she was anemic. Her renal function tests and Serum electrolyte levels were normal. Urine microscopy showed 25–30 RBCs per high power field with plenty of pus cells and albuminuria. Urine culture grew *Escherichia coli*. Ultrasound of the whole abdomen revealed a thick-walled urinary bladder and a bilateral mild increase in renal cortical echoes. All other abdominal viscera were normal. Based on the clinical symptoms, her diabetic status, and laboratory and ultrasound abdomen findings, a provisional diagnosis of acute infective pyelonephritis was made. Contrast-enhanced CT of the abdomen was suggestive of left pyelonephritis and cystitis. Contrast-enhanced CT abdomen revealed a compressed RLRV between the aorta and vertebral column [Figure 3].

Figure 3 illustrates that the left renal vein was single and identified to course behind the abdominal aorta. There

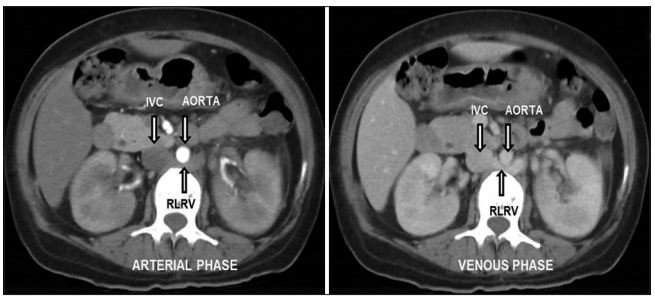

Figure 3: Computed tomography scan of the abdomen showing the course of RLRV

were no collateral veins. Correlating the clinical symptoms and the radiological findings, a diagnosis of PNCS was made. She was started on antibiotics and anti-inflammatory drugs, following which her symptoms improved. She was discharged with advice to follow-up regularly for the recurrence of symptoms.

DISCUSSION

The development of the inferior vena cava by itself is such a complex process that efforts to understand the development of the renal vein make it even more complicated. LRV entrapment, also known as nutcracker syndrome, was first described by El-Sadr and Mina in 1950.[10] Belgian physician De Schepper was first credited with the terminology, NCS, in 1972. [11] The overall incidence of RLRV was reported as 3% and a vast majority of them are asymptomatic.

Traditionally, RLRV is classified into two distinct types. Type 1 is associated with an obliterated ventral preaortic limb of the LRV, but persistent dorsal retro aortic limb which subsequently joins the IVC in the orthotopic position. Type II anomaly results from the obliteration of the ventral limb of the LRV. The dorsal limb turns into the RLRV and lies at the level of L4 L5 and joins the gonadal and ascending lumbar veins before joining IVC. Jang *et al.* subsequently reported the congenital anomalies of the RLRV as four types. Type III anomaly is the circum-aortic LRV. One vein passes posterior and the other vein passes anterior to the aorta to join the IVC. In type IV anomaly, the ventral preaortic limb of the LRV is obliterated, and the remaining dorsal limb becomes the RLRV and joins the left common iliac vein.[5]

Most of the patients remain asymptomatic throughout their lifetime. While most remain healthy, some of the symptomatic ones develop clinical manifestations during their second or third decade of life. Few patients show only microscopic hematuria, which may be picked up during routine evaluation. Others may present with left loin pain, and unilateral left varicocele due to venous stasis at the gonadal vessel, also known as pelvic congestion syndrome. Gastrointestinal symptoms and arterial hypertension may occasionally be the presenting symptoms.

Microhematuria in PCNS, though rare, is not so uncommon. The mechanical compression of the LRV between the aorta and the vertebral body results in a rise in the pressure gradient between the LRV and the vena cava. This rise in pressure gradient, in turn, leads to the rupture of the membranous barrier between the smaller veins and the collecting system in the renal fornix, resulting in microhematuria.[12] Daily *et al.* reported a direct correlation between the LRV pressure (LRVP) and the degree of hematuria.[13] Patients with PNCS are best managed based on their symptomatology, clinical manifestations, the severity of LRVP, and hypertension.

In patients with mild hematuria and in young individuals, who are willing to come for a regular follow-up, conservative and supportive treatment is offered. Young adults, aged <18 years can be followed up for a minimum period of 2 years as there is a 75% chance that there may be a complete resolution of microhematuria. Endovascular stenting is a viable option for patients who have bothersome pain and hypertension due to elevated LRVP.[14] However, its future role in the management of this condition remains to be established and the potential complications such as fibromuscular dysplasia, stent migration, thrombosis, restenosis, and embolization have to be borne in mind.

The various treatment options include conservative treatment, intra-vascular stents, chemical cauterization, open surgical intervention, auto-transplantation, and finally if everything fails, nephrectomy.[15]

CONCLUSION

The authors report a case of a 40-year-old female who presented with left loin pain and microscopic hematuria, diagnosed as PNCS due to the compression noted in the retro aortic renal vein. Our case report discusses a rare presentation of PNCS. A thorough anatomical knowledge of this rare anomaly is imperative as this rare entity may present with a variety of signs and symptoms. This manuscript also reinforces the need for a high index of clinical suspicion to achieve a prompt diagnosis. Conservative treatment is recommended for patients with mild symptoms. Patients with serious impairment or severe symptoms may benefit from surgical or endovascular intervention.

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How to cite this article: Bose JD, Krishnamoorthy S, Ramachandran K. Nut-cracker Phenomenon Masquerading as Loin Pain Micro-hematuria Syndrome: Anatomical and Urological Perspectives. Int J Sci Stud 2023;10(10):7-10.

Source of Support: Nil, **Conflicts of Interest:** None declared.