

Nutcracker Syndrome

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Received: November 08, 2021

Published: December 15, 2021

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Abstract

Nutcracker Syndrome is a rare condition that presents with variable non-specific symptoms and signs including loin pain, chronic pain syndrome, macroscopic and microscopic haematuria, orthostatic albuminuria, occasionally varicocele, infrequently chronic renal failure. As a result, these patients are often treated as pyelonephritis or diagnosed with benign orthostatic albuminuria. They may also be subject further investigation with cystoscopy by urology. It is not uncommon for these patients to be labelled with chronic pain syndrome, functional pain syndrome or fibromyalgia, with significant morbidity involving chronic pain poorly controlled with analgesia.

We present a 29-year-old medical student with chronic pain, who was given a diagnosis of fibromyalgia. Following recurrent presentations to emergency department with left loin pain and intermittent haematuria, the patient was admitted under urology for further investigation. Doppler ultrasound, CT, and MRI showed compression of the Left Renal Vein (LRV) between superior mesenteric artery and abdominal aorta confirming the diagnosis of nutcracker syndrome. The patient opted for endovascular therapy with left renal vein stenting that resulted in resolution of all symptoms.

Although nutcracker syndrome is a rare condition, a low threshold to discuss radiological findings with a senior radiologist is required to exclude nutcracker syndrome once common causes of haematuria and loin pain have been ruled out. Failure to diagnose Nutcracker in a timely manner can lead to serious complication and irreversible kidney injury.

Keywords: Renal Vein; Kidney; Nutcracker Syndrome

Case Presentation

A 29-year-old medical student with chronic pain was initially diagnosed with fibromyalgia, which impacted his medical education. She resigned from university for two years due to the impact of poor symptom control on his ability to study. Due to recurrent episodes of loin pain and intermittent haematuria, she presented to the emergency department multiple times. Repeat clinical examination was unremarkable, apart from tender left loin on palpation.

The following investigation were done with results either negative or normal; full blood count, kidney function, liver function tests, CRP, ESR, vasculitis and immune screen. Urine sent for red

cell cast and dysmorphic red cells, serum immunoglobulin including IGA, complements.

Ultrasound of the abdomen and CT KUB were reported no abnormality.

The patient was admitted for further investigation. She was referred to urology and underwent cystoscopy which was normal. Albumin creatinine ratio (ACR) was 500 supine and increased to 800mg/L after exercise. The patient was referred to the renal service to exclude IGA Nephropathy, thin membrane disease and recurrent renal calculi. Kidney biopsy did not show inflammation, scarring or deposits on electron microscopy and immunofluorescence.

The patient was referred to chronic pain clinic, and commenced on ketamine infusion via patient-controlled analgesia (PCA).

Due to increasing pain, dyspareunia, pelvic congestion, she was reviewed gynaecology to exclude pelvic inflammatory disease. However, with increasing symptoms including haematuria, patient was readmitted under renal for observation and reinvestigation.

Doppler ultrasound, CT, and MRI showed compression of the Left Renal Vein (LRV) between superior mesenteric artery and abdominal aorta known as beak sign. Doppler ultrasound showed reduction in the peak speed velocity (PSV) of the pre-stenotic portion and increased peak speed velocity of the post stenotic segment with a flow post-stenotic/pre-stenotic ratio of more than 5 and high resistive index (RI) (< 0.77). The superior mesenteric branching angle was < 35 degrees. Phlebography showed significant venous pressure gradient between the left renal vein and inferior vena cava at 4mm in addition to peri-renal venous collaterals.

These results confirmed a diagnosis of nutcracker syndrome. Diagnosis and options for treatment were discussed with the patient, who opted for endovascular therapy with left renal vein stenting that resulted in symptom resolution. The patient did not require further analgesia, returned to medical school, and successfully graduated as an intern during the time of writing this article.

Discussion

Nutcracker syndrome is a very rare condition with abnormal anatomy of the left renal vein, abdominal aorta, and superior mesenteric artery [2]. It was first described by Grant in 1973, 'the left renal vein, which lies between the aorta and superior mesenteric artery, resembles a nut between the jaws of a nutcracker' [1]. Anatomically, it is characterised by stenosis of the aorta- mesenteric segment of left renal vein and post stenotic dilation of the distal segment of the left renal vein [2]. Where the anatomical abnormality is present without symptoms, it is known as the Nutcracker phenomena.

Nutcracker syndrome causes a plethora of symptoms in the form of, left loin pain, pelvic congestion in females, dyspareunia, and varicocele in males, microscopic and macroscopic haematuria, and orthostatic albuminuria. Very rarely it can progress to renal vein thrombosis and chronic kidney injury without treatment [3].

As nutcracker syndrome has a very nonspecific clinical pre-

sentation, it is rarely diagnosed in a timely manner. Furthermore, anatomical abnormalities can often not cause any symptoms at all [4]. A patient is often diagnosed with functional pain syndrome, and not uncommonly patient subject to invasive investigations including cystoscopy and kidney biopsy that are unlikely to assist in diagnosis. Some patients will be incorrectly diagnosed with post infectious glomerulonephritis if they have an infection preceding the presentation.

The prevalence is higher in the young and middle aged [5]. Nutcracker syndrome is higher in thin females who have accelerated development in vertebral bodies with decrease in paravertebral fats, which result in a narrow angle between the aorta and superior mesenteric artery.

Two forms have been found to cause nutcracker syndrome, anterior and posterior nutcracker syndrome. More commonly, anterior nutcracker syndrome where the angle between superior mesenteric artery (SMA) and abdominal aorta is less than 35 degrees. This is a core feature of the anterior nutcracker syndrome found on the sagittal plane on CT, with a sensitivity and specificity of 90% [5]. Other criteria which almost have sensitivity and specificity higher than 90% include beak sign (severe narrowing of the left renal vein less than 32 degrees), left renal vein diameter ratio (hilar to aorto-mesenteric ratio more than 5), collateral venous circulation in the renal hilum or retroperitoneum, renocaval pressure gradient higher than 3 is almost diagnostic.

Figure 1

Evidence of compression effect of the left renal vein between the superior mesenteric artery (SMA) and aorta with reduced aortic-SMA angle about 35° (the normal angle is approximately 45°).

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Case courtesy of Dr Safwat Mohammad Almoghazy, Radiopaedia.org, rID: 51924.

Left renal vein transposition and auto-transplantation are invasive but curative treatments [6]. Laparoscopic surgery has started to replace open surgery which include spleno-renal venous bypass and left renal vein-inferior vena cava transposition. This procedure avoids splenic injury and is considered current standard of care. It is less invasive and large case series have shown that patients treated with stenting have resolution of symptoms and no restenosis after 5 years [6-8].

Conclusion

Although symptoms and signs of nutcracker syndrome are vague and nonspecific, failure to diagnose Nutcracker in timely manner can lead to serious complications with irreversible kidney injury. Medical practitioners should have a low threshold to discuss radiological findings with a senior radiologist to exclude Nutcracker after ruling out other causes of haematuria and loin pain. This is particularly so, given endovascular treatment has shown considerable success.

Acknowledgment

Associate Professor adel Ekladios thanks Sharna Shughandem YI I Li for editing the Manuscript

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Volume 6 Issue 1 January 2022

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