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Case Report

Nutcracker Syndrome: A Rare Cause of Hematuria, a Case Report

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Abstract

Introduction: Nutcracker syndrome is a rare and often unrecognized cause of hematuria. It results from compression of the left renal vein, typically between the abdominal aorta and the superior mesenteric artery, leading to stenosis of its aortomesenteric portion and dilation of its distal portion.

Case Report: We report the case of a 39-year-old woman who presented chronic abdominal pain and microscopic hematuria, in whom clinical and radiological findings were consistent with this syndrome.

Discussion: The variety of clinical manifestations makes the diagnosis difficult and often delayed. Therefore, imaging plays a key role in the diagnosis by confirming the stenosis of the left renal vein and ruling out any differential diagnosis. Its treatment remains controversial.

Conclusion: Nutcracker syndrome is a rare entity. Lack of awareness of this syndrome leads to diagnostic delays. Imaging plays a crucial role in confirming the diagnosis.

Keywords: *Nutcracker Syndrome; Left Renal Vein; Hematuria*

Introduction

Nutcracker syndrome is defined by the compression of the left renal vein between the abdominal aorta and the superior mesenteric artery [1]. Symptoms can be misleading, dominated by hematuria, which may or may not be associated to left lumbar pain, pelvic congestion syndrome for women and varicocele for men. The diagnosis is evoked by clinical and biological arguments, then confirmed by imaging. Management is multifactorial and multidisciplinary. Our case study highlights the key role of imaging in the recognition of this syndrome, which unfortunately remains unknown to many clinicians.

Case Report

A 39-year-old young women, mother of two, with no previous pathological history, presented with chronic abdominal pain predominating in the left side, associated with hematuria, progressing for two months. Clinical examination and biological tests were normal. Abdominal ultrasound was normal. An abdomino-pelvic CT scan was performed before and after injection of contrast, and revealed dilatation of the left renal vein (LRV) (Figure 1) with compression in the angle formed between abdominal aorta and the superior mesenteric artery (SMA), which measures 16.4° (Figure 2).

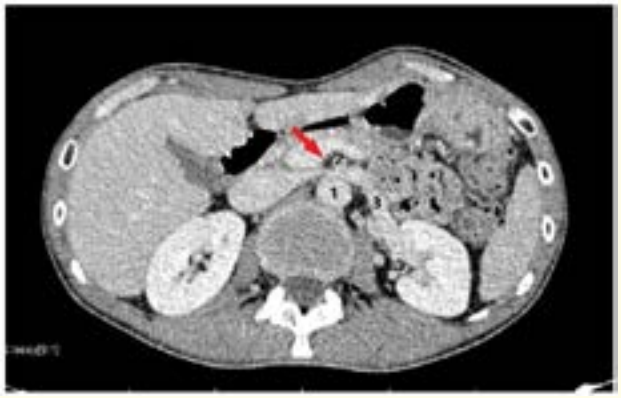

Figure 1: *Axial CT section after contrast injection showing the beak sign (arrow) at the aorto-mesenteric space; 1) abdominal aorta; 2) superior mesenteric artery; 3) left renal vein.*

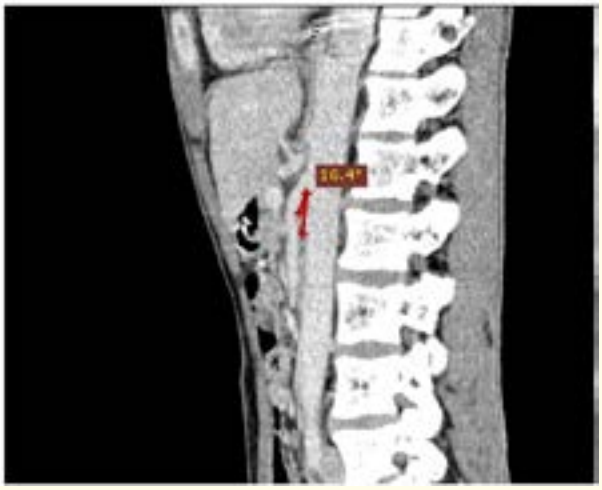

Figure 2: *CT sagittal section after contrast injection showing angulation between the abdominal aorta and superior mesenteric artery measured at 16.4°, confirming the diagnostic criteria for Nutcraker syndrome.*

In view of this highly suggestive scannographic picture, the CT scan was sufficient to make the diagnosis of Nutcracker syndrome. After multidisciplinary discussion, the patient underwent transposition of the superior mesenteric artery (SMA) below the LRV (Figure 3 and 4). The surgical consequences was good.

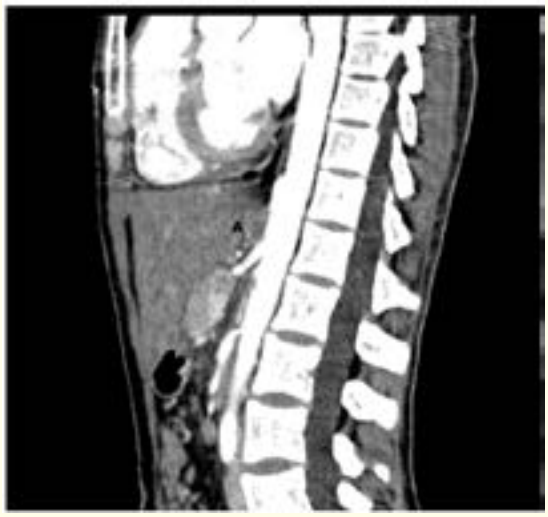

Figure 3: *CT sagittal section after postoperative contrast injection showing transposition of the superior mesenteric artery below the left renal vein.*

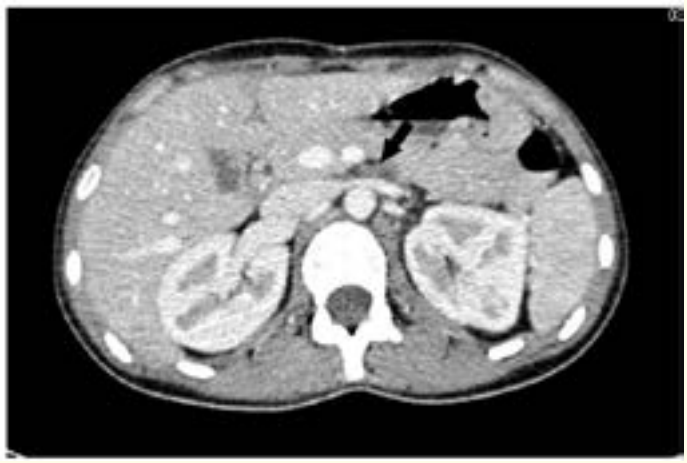

Figure 4: *Axial CT section after postoperative contrast injection showing free passage of the left renal vein (arrow).*

Discussion

Nutcracker syndrome is a rare entity defined by compression of the LRV as it passes through the aorto-mesenteric clamp (Figure 5). Its prevalence is highest in young subjects aged 30 to 40, with a clear female predominance [2], but remains underestimated as it is often misdiagnosed and remains asymptomatic for a long time. The syndrome was first described in 1937 by the pathologist Grant [3].

Nutcracker syndrome can be divided into three types: anterior, posterior and mixed [4]. In the anterior type, the SMA forms an acute angle, compressing the LRV and creating hyperpressure in the vein. In the posterior type, the LRV passes behind the abdominal aorta

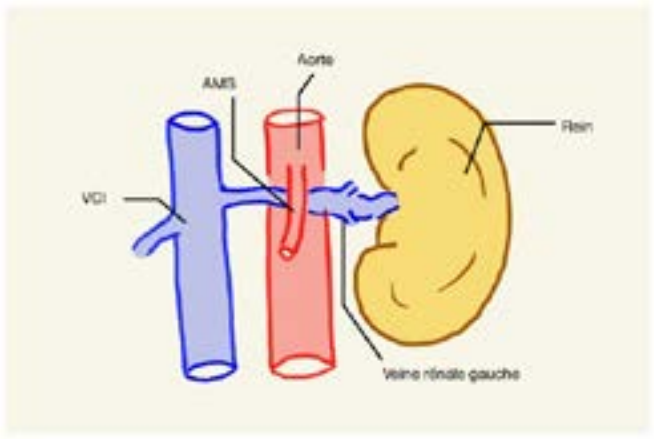

Figure 5: *Anatomical drawing illustrating the passage of the left renal vein through the aorto-mesenteric clamp.*

and is compressed between the aorta and the vertebral spine. In the mixed type, the anterior branch of the duplicated LRV is compressed between the SMA and the aorta, and the posterior branch between the aorta and the spine.

Several etiopathogenic hypotheses has been reported in the literature: dorsolateral ptosis of the left kidney with stretching of the LRV, too low or lateral origin of the SMA implanted at a pronounced angulation when the normal angle of insertion is around 90°, duplicity of the left renal vein, ectopic or horseshoe-shaped kidneys. However, all causes of extrinsic renal vein compression can induce secondary Nutcracker syndromes, including pancreatic cancers, retroperitoneal tumors and para-aortic adenopathies.

The clinical manifestations of Nutcracker syndrome are highly variable: many patients are asymptomatic and the abnormality is discovered by chance during an imaging examination performed for another indication. Symptomatic cases mainly present in the 2nd or 3rd decade with urological or gynaecological symptoms. Urological signs include abdominal or lumbar pain, micro or macroscopic hematuria (from the left urinary tract), varicocele or varicose veins of the lower limbs. Gynaecological signs are suggestive of pelvic congestion syndrome, and include dysmenorrhoea, dyspareunia and post-coital pain, lower abdominal pain, dysuria, pelvic, vulvar, gonadal, thigh varicose veins, and emotional troubles.

Diagnosis is based primarily on imaging. Echo-Doppler is usually the first imaging test, revealing collateral veins around the LRV, which reflect venous hyperpressure and are a radiological criterion for the syndrome. However, abdominal CT-scan with contrast injection is the modality of choice for visualizing compression of the LRV and its hemodynamic consequences. The characteristic sign is the visualization of a beak corresponding to compression of the LRV by the aorto-mesenteric clamp. On sagittal reconstructions, the normal angle formed by the SMA and the aorta should be between 35° and 56°. Narrowing is confirmed when this angle is between 7° and 22°. The distance between the SMA and the aorta is reduced, measuring between 2 and 8 mm, with disappearance of normal surrounding retroperitoneal fat, while the normal distance is 10 to 28 mm [5].

Phlebography used to be the gold standard for confirming the diagnosis, but it remains an invasive examination, now supplanted by non invasive imaging. It is currently used for treatment rather than diagnosis. It measures the pressure gradient between the inferior vena cava and the left renal vein (LRV), which is greater than 3 mmHg in nutcracker syndrome, compared with a normal value of 0 to 1 mmHg.

Treatment remains highly controversial. In addition to surgery (transposition of the LRV and/or SMA) indicated in cases of severe pain and massive hematuria, some teams use endovascular treatment. Finally, a wait-and-see attitude is currently recommended in moderate cases.

Conclusion

Nutcracker syndrome is a rare entity. Lack of awareness of this syndrome leads to delays in diagnosis. It should be suspected in the presence of chronic pelvic pain syndrome, atypical lumbar pain and micro or macroscopic hematuria. Imaging plays a major role in confirming the diagnosis. CT-Scan is the gold standard.

Patient Consent

Written informed consent has been obtained from the patients for the publication of this case report and any accompanying photographs. This case report is an incidental finding in the course of clinical work and has no ethical implications.

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