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Hematuria as a Common Symptom of the Nutcracker Syndrome: Two Case Reports and Mini-review

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Nutcracker syndrome results from compression of the left renal vein (LRV), usually in the range formed by the abdominal aorta and the superior mesenteric artery (SMA), leading to stenosis of the aorto-mesenteric part of the left renal vein and dilation of its distal part.
The symptomatology remains dominated by abdominal pain and hematuria. Its diagnosis is essentially based on modern imaging means (computed tomography, ultrasound-Doppler, phlebography) and its treatment is controversial. We report two observations of patients, the first admitted for intermittent macroscopic hematuria and the second for incidentally discovered microscopic hematuria, and whose radiological exploration revealed Nutcracker syndrome.

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1. INTRODUCTION

"Nutcracker syndrome encompasses all manifestations linked to venous stasis induced by stricture of the left renal vein: either between the aorta and the superior mesenteric artery, or between the aorta and the spine. It is revealed in half of the cases by atypical pain in the left flank, leading the diagnosis towards other pathologies" [1].

We report two observations of patients admitted to the nephrology and hemodialysis department of the Ibn Rochd University Hospital of Casablanca in Morocco for microscopic or macroscopic hematuria, in whom the diagnosis was made based on the scan signs.

1.1 Case Presentation 1

A 19-year-old patient, presented to our department with a history of five episodes of f macroscopic hematuria. The clinical examination was unremarkable. The complete biological workup was normal with minimal pyelocalic dilatation on the radiological workup on renal-vesical ultrasound. An abdominal CT scan was performed with and without injection of contrast medium and did not reveal any specific pathology. However, it showed compression of the left renal vein as it passed between the aorta and the superior mesenteric artery with dilatation of the left renal vein (ratio of the diameter of the hilar and aorto-mesenteric portions: 6.47 (>5.5)) with narrowing of the portion trapped in the fork formed by the abdominal aorta and the superior mesenteric artery, the angle of which measures 22° (Figs. 1 and 2).

The therapeutic abstention was indicated by the vascular surgeons, considering the intermittent character of the clinical symptomatology and its moderate intensity.

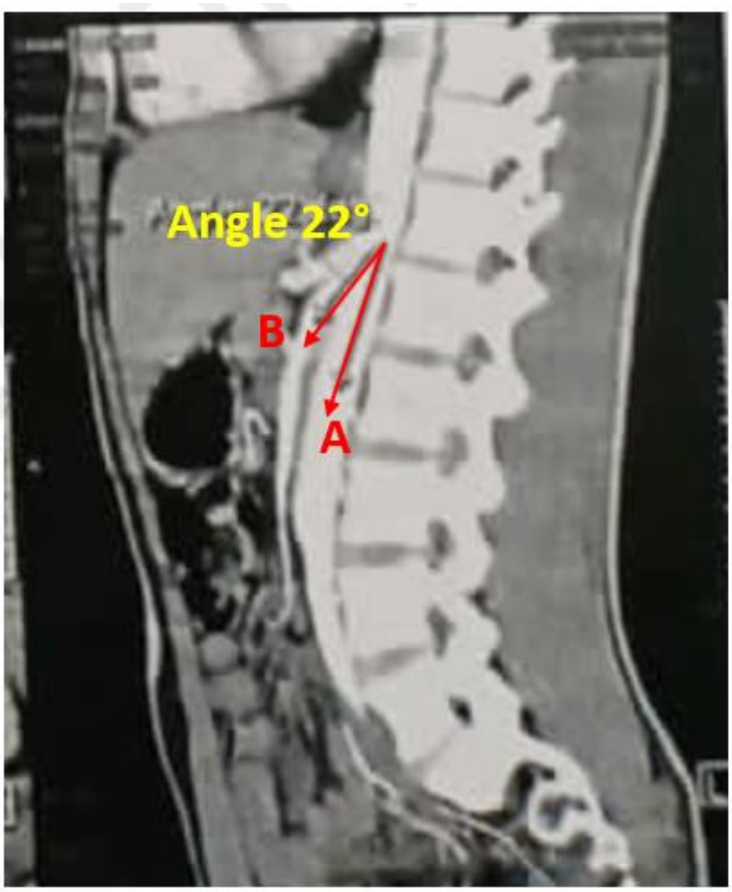

Fig. 1. Sagittal section of the abdominal-pelvic scanner before and after PDC injection with A) angulation between the abdominal aorta and the superior mesenteric artery; B) less than 41° (measured at 22°)

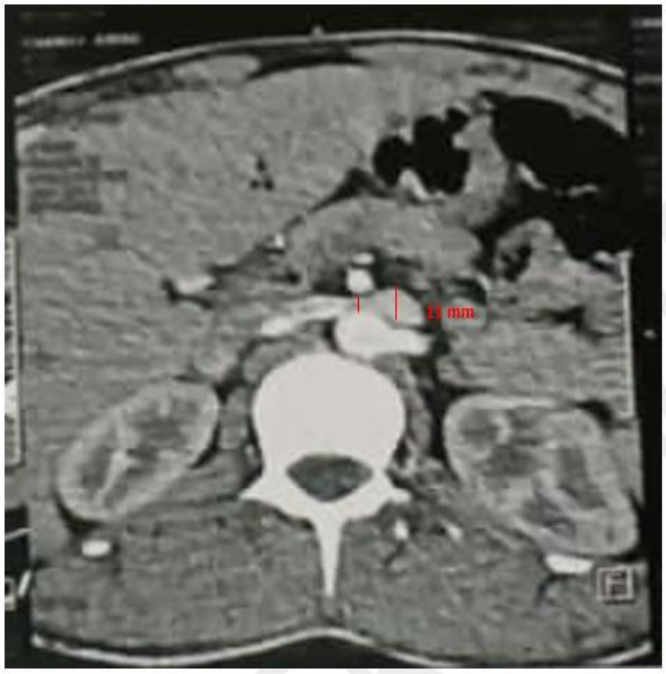

Fig. 2. Axial section of the abdominopelvic CT: dilatation of the hilar segment of the left renal vein with a hilar and aortomesenteric portion diameter ratio >5.5 (11:2)

1.2 Case Presentation 2

A 46 year old patient, presented to the emergency department with abdominal pain and vomiting associated with acute renal failure at 49 mg/l of plasma creatinine. The clinical examination revealed epigastric tenderness with signs of extracellular dehydration.

Urine analysis confirmed a microscopic hematuria without any other urine sediment anomaly and a negative 24-hour proteinuria.

As part of the etiological assessment of his symptoms, an abdominal ultrasound completed by an abdominal CT scan showed pancreatitis stage A of the Balthazar classification with evidence during the same examination of a pinched left renal vein between the abdominal aorta and the superior mesenteric artery in favour of a Nutcracker syndrome. (Fig. 3).

The evolution was marked after intravenous rehydration with isotonic saline by the improvement of the renal function to 28 mg/l of CP with the normalization of the creatinine figs at the end of 6 days. The patient was referred to the department of general surgery and vascular surgery for further follow-up.

In both cases, the diagnosis of this syndrome was confirmed after elimination of other causes of hematuria by means of an exhaustive workup.

2. DISCUSSION

The first anatomic description of LRV entrapment was reported in 1937 by Grant [2], while the first clinical impact of this anatomic condition was made by El-Sadr and Mina in 1950 [3]; as far as we know, the term nutcracker was probably first used in Schepper's papers in 1972 and Chait's in the following year [4].

This term nutcracker syndrome should be reserved for patients with clinical symptomatology associated with these anatomical features because, as mentioned by Shin and Lee [5], there are similar anatomical variants that do not have clinical repercussions and in these cases one should rather speak of "nutcracker phenomenon".

"The common anterior NCS is created by the entrapment of LRV between AA and SMA. Posterior NCS is rare, resulting from the compression of retro-aortic LRV by abdominal aorta and vertebral column" [6]. Mallat et al. [7] reported other variations of NCS; anteroposterior NCS and Wilkie syndrome (SMA syndrome) in association with anterior NCS which results from compression of the third portion of the duodenum between the aorta and the SMA [8].

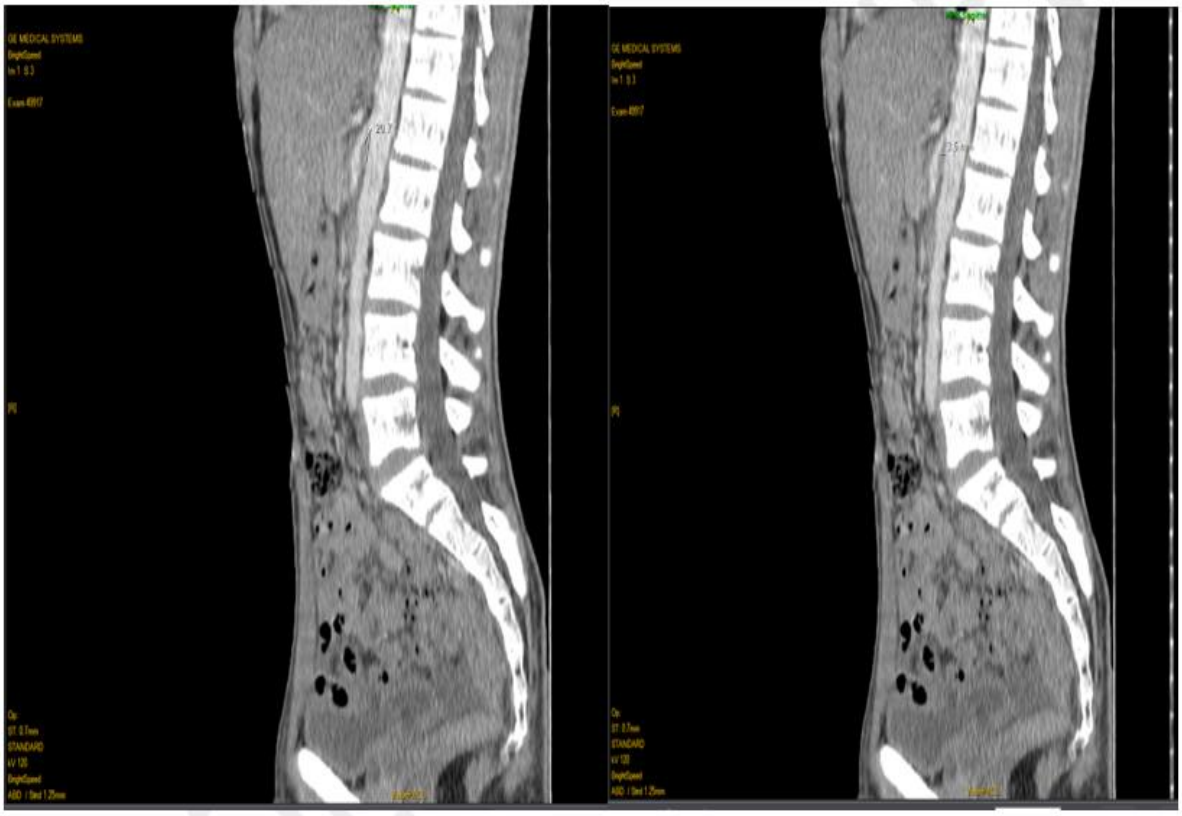

Fig. 3. Axial sections of the abdominopelvic scanner: A) Compression ratio >2.25 and B) aorto-mesenteric angle measured at 20.7°

Its pathophysiology remains unknown but several hypotheses have been described: anatomical variants [5,8]; duplicity of the left renal vein, in which case patients may suffer from both anterior and posterior components; ectopic or horseshoe kidneys; ectopic birth of the spermatic and ovarian arteries may also constrict the renal vein; cofactors: hyper-pressure of the venous network (vena cava more than portal) may contribute to the increase or appearance of the signs.

The prevalence of the syndrome seems to be higher in young people, with an average age at diagnosis between 30 and 40 years. Women are more often affected than men.

The clinical manifestations are twofold:

- Abdominal pain, caused by pelvic congestion syndrome, due to venous stasis upstream of the MAP;
- "Micro or macroscopic Hematuria is the most frequent sign in NS patients. The pathogenetic mechanism proposed to explain hematuria is that increased venous pressure into the LRV and left gonadal vein can lead to rupture of the septa between the venules and the collecting system in the renal parenchyma. On the other hand, no specific glomerular damage is reported in the literature" [9].

However, most of the left renal vein compressions would remain asymptomatic, as suggested by BUSCHI et al, who found in a series of phlebographies made in asymptomatic patients, a distended renal vein in 72% of cases [10].

"The basis of the diagnosis is an accurate history and a careful clinical examination. The diagnosis of SNA is based primarily on modern imaging techniques. The multi-bar scanner, with its multi-planar acquisitions, offers a definite advantage in establishing the diagnosis. Doppler ultrasound is a technique that can be very useful to confirm the diagnosis if it can be shown that the ratio between the maximum velocities of the RVG at the level of the stenosis and the maximum upstream distension is greater than or equal to five. Additional diagnostic confirmation can be based on phlebography, which allows measurement of the pressure gradient between

the inferior vena cava and the RVG: this is on average 1 mm Hg and is at least 3 mm Hg in the case of SCN" [8].

The first treatment for nutcracker syndrome was described by Pastershank in 1974 [11].

"The treatment is variable, and conservative management, interventional endovascular approaches, or more invasive surgical treatments are possible; the choice of whether to proceed with invasive treatment or not depends on the severity of the symptoms and clinical signs" [12].

"Conservative treatment is recommended for patients with modest hematuria and who are under 18 years of age, and any procedure should follow at least six months of conservative follow up; it has been reported that in most patients with mild symptoms, there may be complete spontaneous resolution of symptoms" [6].

"When clinical symptoms of NS are not tolerable and result in significant clinical manifestations (chronic pain, recurrent hematuria, pelvic congestion), conservative treatment may not be enough. Many surgical approaches, including medial nephropexy, renal vein bypass, transposition of the left renal vein, transposition of the superior mesenteric artery, gonad-caval bypass, and auto-transplantation of the left kidney, were reported" [8].

"Many patients refuse an invasive surgical treatment, opting for endovascular procedures. However, these procedures are not free from complications. The most important (for incidence and potentially damage) is stent migration, while in-stent restenosis and venous occlusion resulting from fibromuscular hyperplasia or thrombosis rarely occur. Anticoagulant and antiplatelet treatment is necessary to reduce the risk of thrombosis" [13].

4. CONCLUSION

The nutcracker syndrome, although rare and often under-diagnosed, could explain a set of algic and urinary symptoms, in particular hematuria, which can sometimes be the only revealing symptom. The management is nowadays better codified.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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