Hematuria revealing a nutcracker syndrome in the nephrology department of the CHU Ibn Rochd in Casablanca: about 2 cases and review of the literature

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.

Key words:

Nutcracker syndrome, hematuria, left renal vein, vascular compression .

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.

Observations:

Observation 1:

A 19-year-old patient with no previous pathological history was admitted to the department for intermittent macroscopic nonclotting 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° (Figure 1 and 2).

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

Observation 2:

A 46 year old patient with no particular pathological history 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.

The renal assessment showed 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. (Figure 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 figures 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.

Discussion:

Nutcracker syndrome has been rarely reported in the literature[2,3] ; the first description was made by pathologist Grant in 1937[4] and the first clinical case was reported by El-Sadr and Mina in 1950[5] .

This term nutcracker syndrome should be reserved for patients with clinical symptomatology associated with these anatomical features because, as mentioned by Shin and Lee [6], there are similar anatomical variants that do not have clinical repercussions and in these cases one should rather speak of "nutcracker phenomenon".

There are two types of NS: the most classic is anterior NS related to compression of the RVG between the MSA and the aorta; the other, rarer, is posterior NS related to compression of the RVG between the aorta and the vertebral body when the RVG is retro-aortic. The two types can be exceptionally associated in case of duplication of the RVG [7].

Its pathophysiology remains unknown but several hypotheses have been described: anatomical variants [6,7]; 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, the most frequent sign explained by connections between the renal varices and the collecting tubules [8]. According to LOPATKIN [9], hematuria is caused by a rupture of the thin-walled septum that separates the venous system from the collecting system at the level of the calcific fornix. In case of an obstruction on the left renal vein, the collateral venous system (safety valve) includes the gonadal, ureteral, adrenal, phrenal, retro-aortic lumbar, lumbo- azygos, vertebral and capsular veins [10].

However, most of the left renal vein compressions would remain asymptomatic, as suggested by a study by BUSCHI, who found in a series of phlebographies made in asymptomatic patients, a distended renal vein in 72% of cases [11].

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 [7].

The first treatment for nutcracker syndrome was described by Pastershank in 1974 [12]. As the correlations between clinical and paraclinical signs are loose, intervention should only be considered when symptoms are marked (severe pain, significant hematuria, even renal failure) and persist for at least 2 years [7]. There are many treatment options, from abstention to endovascular treatment to nephrectomy, depending on the severity of the symptoms [7].

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.

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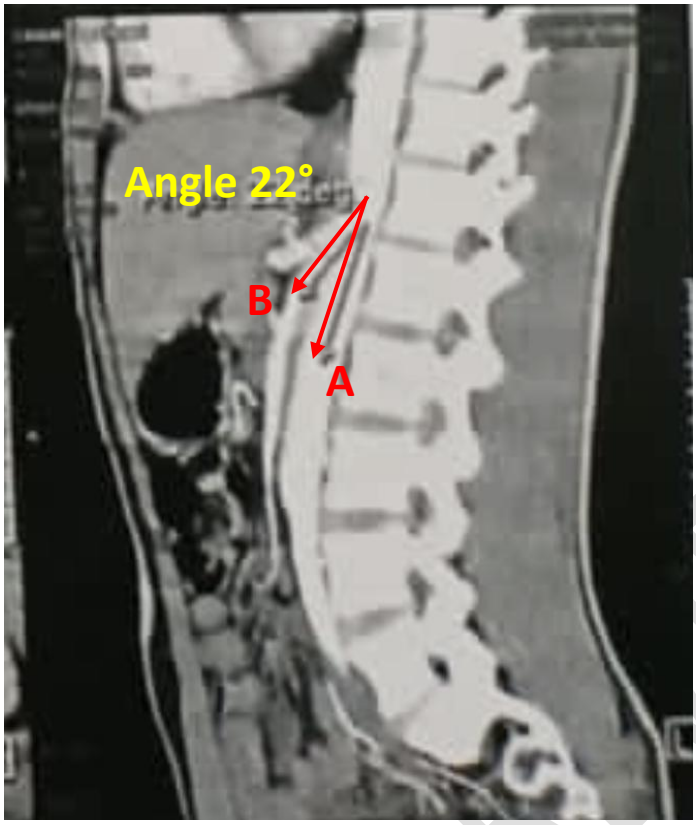

Figure 1 :Sagittal section of the abdominal-pelvic scanner before and after PDC injection with A) angulation between the abdominal aorta and the superior mesenteric artery; B) less than 41° (measured at 22°).

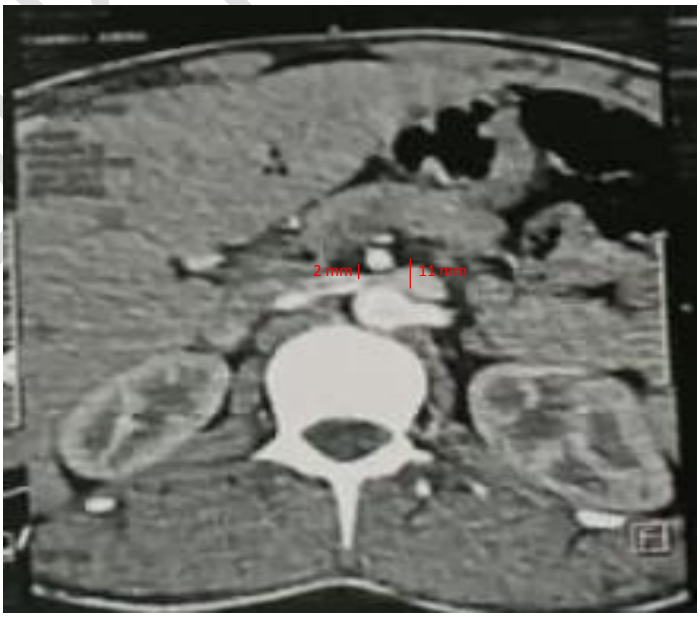

Figure 2 : Axial section of the abdominopelvic CT: dilatation of the hilar segment of the left renal vein with a hilar and aortomesenteric portion diameter ratio >5.5 (11:2)