

## WHICH IS YOUR DIAGNOSIS?

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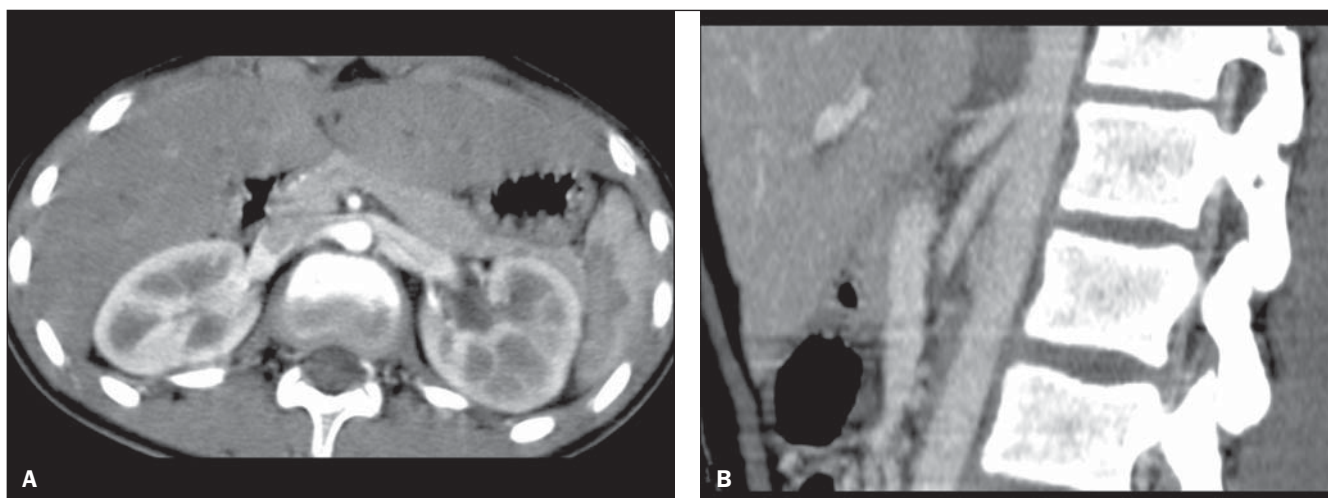
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Female, 11-year-old patient with lumbar pain and intermittent macroscopic hematuria for three months, sometimes with blood clots. The patient did not present any comorbidity and clinical examination was normal. All the laboratory blood tests results, including blood count, coagulation profile, protein electrophoresis, FAN, C3, PCR and biochemistry were normal. Simple urine test revealed reddish color urine and presence of hemoglobin and

red blood cells. The tests for alcohol-acid-resistant bacilli and deformed red blood cells were negative. The 24-hour urine test demonstrated subtle proteinuria.

Abdominal multidetector computed tomography angiography (MDCT-angiography) was performed (Figure 1). Uretrocystoscopy demonstrated intermittent bleeding and presence of blood clots throughout the left ureteral meatus.



**Figure 1.** Abdominal MDCT-angiography image in the axial plane (A) and MIP (maximum intensity projection) reformatted image in the sagittal plane (B).

## Images description

**Figure 1.** Abdominal MDCT-angiography image in the axial plane (A) and MIP (maximum intensity projection) reformatted image in the sagittal plane (B) show compression of left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA) due to narrowing of the space between those two arteries, characterizing the nutcracker phenomenon. Both the distance (A) and the mesoaortic angle (B) are reduced, measuring about 3 mm and 30°, respectively.

**Diagnosis:** Nutcracker syndrome.

## COMMENTS

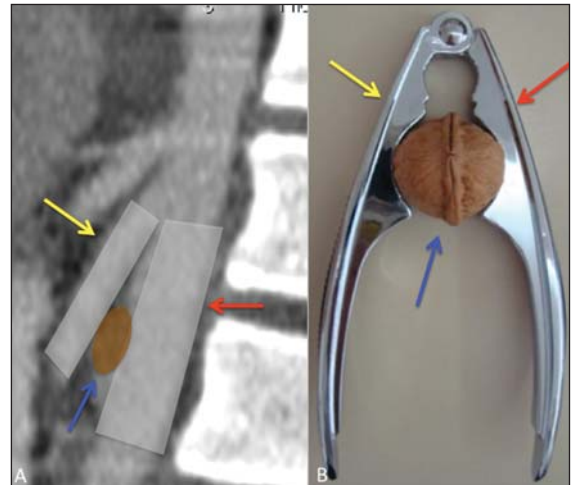
Nutcracker phenomenon (NCP) refers to compression of the LRV, generally because of a narrowing of the angle between the aorta and the SMA. In such a situation, it is described as anterior nutcracker phenomenon, corresponding to the anatomy observed in the present patient. Posterior NCP is also described, where the retro-aortic or circum-aortic renal vein is compressed between the aorta and the vertebral body. In cases where there is association with clinical manifestations, nutcracker syndrome (NCS) occurs. The anatomy of NCP without clinical manifestations may represent a variant of normality<sup>(1,2)</sup>.

The use of the term nutcracker is generally credited to de Schepper who, in 1972, reported the cases of two patients with hematuria and LRV compression. However, the term was first utilized by Chait et al. in 1971<sup>(3,4)</sup>. Figure 2 illustrates the analogy with a nutcracker.

The nutcracker syndrome is most frequently found in young women; and, because of symptoms variability and lack of consensus about diagnostic criteria, its prevalence remains unknown, but probably it is an underdiagnosed entity. Most common clinical manifestations include hematuria, lumbar or abdominal pain, pelvic varices, varicocele and proteinuria in varied degrees. Hematuria occurs because of rupture of the thin walls of the venous collaterals within the adjacent calyceal system due to venous hypertension secondary to the LRV compression<sup>(4,5)</sup>.

In the present case, the diagnosis of NCS was achieved after ruling out other

**Figure 2.** Analogy with a nutcracker (A,B). Nut = LRV (blue arrows); right leg of the nutcracker = aorta (red arrows); left leg of the nutcracker = AMS (yellow arrows).



possible causes of hematuria, upon visualization of bleeding through the left ureteral meatus at ureterocystoscopy and characterization of the anatomical findings of NCP by MDCT angiography.

Several imaging studies are utilized in the diagnostic workup of NCS, such as Doppler ultrasonography (US), MDCT angiography, magnetic resonance angiography (MRA) and venography. Although venography with measurement of the difference of pressure between LRV and superior vena cava is considered as the most informative method in diagnostic terms, it is invasive and is not performed in patients presenting mild symptoms as well as in children. Additionally, it is not an entirely reliable method, since overlap of pressoric levels may occur between asymptomatic patients and those with NCS<sup>(1,5)</sup>.

MDCT angiography with multiplanar reformatting allows the evaluation of the relationship between aorta, SMA and LRV, so it is easy to measure both the mesoaortic angle and the distance between the SMA and the aorta. This method has shown to be superior to venography and US in the identification of LRV compression<sup>(6)</sup>.

There is a great variability in the measurement of the mesoaortic angle, and small angles are associated with NCS patients as compared with asymptomatic patients. Typically, the mesoaortic angle and the distance between the aorta and the SMA at the level of the LRV in symptomatic patients are  $39.3^\circ \pm 4.3^\circ$  and  $3.1 \text{ mm} \pm 0.2 \text{ mm}$ , respectively. On the other hand, in asymptomatic patients, they are  $90^\circ \pm 10^\circ$

and  $12 \text{ mm} \pm 1.8 \text{ mm}$ <sup>(5)</sup>. The present patient presented an angle of about 30° and distance of 3 mm, in compatibility with the NCP anatomy.

The treatment for NCS is dependent on the symptoms severity. In patients under the age of 18 years, with mild to moderate hematuria, the best option is a conservative approach with clinical follow-up, considering that complete resolution is observed in 75% of cases over a two-year period<sup>(7)</sup>. Such approach was adopted in the present case.

One can conclude that MDCT-angiography can be utilized as a quite reliable, noninvasive method to characterize the NCP anatomy, allowing the diagnosis of NCS in patients with a compatible clinical presentation.

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