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The Nutcracker Syndrome: Rare Cause of Pediatric Abdominal Pain

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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 Study

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ABSTRACT

Nutcracker syndrome results from compression of the left renal vein (LRV), usually in the fork 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. We report the case of a 10-year-old child, with no family history of renal disease, diabetes or hematuria. He was admitted to our clinic for paroxysmal abdominal pain in the epigastric area that was not improved by the usual analgesics, associated with vomiting without transit disorders or hematuria, and had been evolving for 10 days prior to his admission. The clinical examination was normal, the normal biological check-up, the abdominal CT scan was in favor of the Nutcracker syndrome. The treatment was symptomatic with a good evolution.
Nutcracker syndrome is a rare entity, to be evoked in the diagnostic range of rare etiologies of unexplained abdominal pain.

Keywords: Nutcracker syndrome; hematuria; abdominal CT scan; left renal vein.

1. INTRODUCTION

Nutcracker syndrome or Nutcraker 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 left aorto-mesenteric part of the left renal vein and dilation of its distal part. Its diagnosis is essentially based on modern imaging means

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(computed tomography, echography-Doppler, phlebography).

Nutcracker syndrome is a rare disease with only scare published literature, especially in children. Up until now only case reports and small case series are published. Because of this issue, clinical diagnostic criteria could not be established and treatment options remain controversial.

2. CASE PRESENTATION

We report the case of a 10-year-old child, with no family history of renal disease, diabetes or hematuria. He was admitted to our clinic for paroxysmal abdominal pain in the epigastric area that was not improved by the usual analgesics, associated with vomiting without transit disorders or hematuria, and had been evolving for 10 days prior to his admission. The clinical examination was normal, the BMI normal, no abdominal mass or organomegaly. The standard radiological workup (abdominal and pelvic ultrasound) was normal and the urinalysis was unremarkable, with no hematuria, proteinuria or signs suggestive of urinary tract infection. Amylase and lipase to assess pancreatic pathology were within normal limits. Abdominal CT scan was in favor of Nutcracker syndrome.

The treatment was symptomatic with a good evolution.

3. DISCUSSION

Nutcracker syndrome is a rare disease with only few published literature, especially in children.

“The presence of the clinical features forms a basis for the diagnosis. The presence of hematuria and proteinuria in a patient must be explored. Urine analysis, urine culture, and imaging of kidneys should be performed. Doppler US (DUS), CT angiography (CTA), magnetic resonance angiography (MRA) and retrograde venography are imaging methods used to diagnose NCS. DUS with sensitivity and specificity as high as 78% and 100%, respectively, is an appropriate initial diagnostic test in patients with suspected NCS” [1].

“The Nutcracker syndrome is difficult to diagnose in pediatrics in the face of the various causes of abdominal pain and other possible causes compatible with the clinical presentation of the patient” [2-6].

“The reference methods to identify this syndrome are always phlebography of the LRV in parallel with the measurement of the pressure gradient between the LRV and the inferior vena cava, or with intravascular USG” [2,6,7]. “However, it has been reported that measurement of LRV diameter and peak flow velocity with DUS can be used with significant success for diagnosis” [2,3,8-12]. Parc et al. [1] proposed that thresholds> 4.2 for AP diameter ratio and 4.0 for PV ratio should be used as sonographic criteria in the diagnosis of NCS in children.

The management of this syndrome is controversial and depends on the severity of clinical symptoms. Management modalities range from therapeutic abstention and monitoring to nephrectomy. In pediatrics, conservative treatment is recommended because of the possibility of spontaneous resolution after the development of adipose tissue.

“Analgesics may be offered to relieve pain, Aspirin therapy has been reported to improve the left-right renal perfusion ratio in children with NCS” [10].

4. CONCLUSION

NCS is a rare and underdiagnosed syndrome that has significant morbidity. Given its relatively recent individualization, the management of this syndrome is not yet well codified (surgery, endovascular, or abstention).

CONSENT

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

ETHICAL APPROVAL

As per international standard or university standard 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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