Young woman with nutcracker syndrome without main clinic manifestation: Hematuria—Case report

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ABSTRACT

INTRODUCTION: The nutcracker syndrome occurs due to the compression of the left renal vein between the aorta and the superior mesenteric artery. The diagnosis of the syndrome is often difficult and under diagnosed. The main clinical manifestations are hematuria and pelvic or back pain.

PRESENTATION OF CASE: A 25 years old female patient with severe abdominal pain in the lower abdomen without hematuria. Diagnosis with nutcracker syndrome after performing computed tomography. Presented the first episode of pain with 17 years old and was diagnosed at age 25.

DISCUSSION: In the nutcracker syndrome anatomical changes do not generate specific symptoms, causing the disease to be underdiagnosed. In this syndrome, there is increased pressure on the left renal vein, generating several signs and symptoms, and hematuria is one of present. Our patient did not have hematuria. Because of common symptoms, it makes nutcracker syndrome difficult diagnosis, confusing with other diseases, especially with nephrolithiasis. It is usually diagnosed after exclusion of other diseases.

CONCLUSION: The diagnosis of nutcracker syndrome is done after exclusion of other causes of chronic pelvic pain. In most cases, macroscopic and microscopic hematuria are present but was unobserved in this case. Making it more difficult diagnosis.

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1. Introduction

The nutcracker syndrome occurs due to the compression of the left renal vein between the aorta and the superior mesenteric artery, thereby reducing the outflow of blood from the renal vein. The diagnosis of the syndrome is often both difficult and under diagnosed. The criteria for diagnosis and treatment are not well established, and the natural history is not well understood.

The main clinical manifestations are hematuria and pelvic or back pain and it affects mostly females between ages 20 and 40 years old. The advancement of imaging exams has helped better identify nutcracker syndrome, and also treatment with major developments in endovascular surgery, making it less invasive. In the recent past treatment was exclusively dependent on open surgical therapy. Conventional surgery involves procedures associated with increased morbidity, as transposition of the renal vein, renal autotransplant and even nephrectomia [1].

The purpose of this case report is to demonstrate the delay and difficulty of diagnosis of nutcracker syndrome in young patients without hematuria.

2. Presentation of case

D.A.O patient 25 years, female, black. Walked into emergency department (ED) with severe abdominal pain in the lower abdomen which started on the same day with progressive worsening, with nausea and vomiting, more pronounced diffuse abdominal pain in hypogastric and left lower abdomen. Denies hematuria, denies gynecologic complaints. Denies nephrology disease, diabetes or hematuria, in the family.

Menarche at 17, and since then start presenting abdominal pain of high intensity, always associated with the premenstrual period. Often requiring hospital care to perform analgesia. Because these episodes tried consult a gynecologist, which began using continuous combined oral contraceptive. Which made use up to 25 years, remaining without pain conditions in that period was periodic monitoring with gynecologists, performing laboratory tests and ultrasound scans, showing no change. Refers stopped the use contraceptive pills by herself one month before the visit to the

This work has been reported in line with SCARE criteria [21].

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ED. She never had hematuria. Previous total abdominal ultrasound examination carried out eight months ago without changes.

Examination shows vital signs within normal limits, pain in deep palpation in the lower abdomen without signs of peritoneal irritation.

Laboratory tests performed on the day were unchanged. Renal function, blood count and urinalysis unchanged. Urinalysis: leucocytes <10mil, erythrocytes <10mil, negative protein, negative nitrate.

Performed CT scan, abdominal ultrasound and transvaginal ultrasound in ED. Abdomen ultrasound showed discreet paraietal thickening of the bowel in the right lower abdomen, this finding is an unspecific examination finding, and is not evidenced in any article used as reference in this study. Transvaginal ultrasound viewed the presence of ectasia of the uterine vessels. In tomography with intravenous contrast injection was observed changes nutcracker syndrome characteristics (Fig. 1) and dilation of the gonadal vein (Fig. 3) and pelvic vessels (Fig. 2).

Patient remained hospitalized for two days for pain control and evaluation of vascular surgery, which continued its follow-up ambulatory to programming surgical procedure.

3. Discussion

In the nutcracker syndrome anatomical changes generate no specific symptomatology, making the disease underdiagnosed [2]. D’Archembeau et al. [3] reported 83% incidence of the syndrome in patients with pelvic congestion.

Initially the disease was described by El-Sadr, [4] and later named by Schepper [2] as nutcracker syndrome (nutcracker), which is the left renal vein compression by the aorta and superior mesenteric arteries. Having a higher prevalence in women [1,5,6]. It is assumed that it is associated with nephroptosis above or decreased retroperitoneal adipose tissue, leading to elongation of the renal vein and reduce the angle between the superior mesenteric artery and the aorta. The etiopathogenesis of pain is multifactorial, and suggests a hormonal etiology because pains are worse in the premenstrual period, progesterone induces vasodilation and increases blood flow at this stage of the menstrual cycle. [22,23]

In the syndrome, there is increased pressure on the left renal vein [7] and may result in several signs and symptoms such as pain in the flanks, hematuria, left varicocele, fatigue, proteinuria, pelvic congestion. The hematuria can be associated with rupture of the small septum separating veins of the urinary collecting system [8].

Because of common symptoms, it makes it difficult to diagnosis, and confused with other diseases, especially with nephrolithiasis. It is usually diagnosed after exclusion of other diseases [9]. Upon entering the hospital, it was suspected that our patient had kidney stones due to the strong acute pain and cramps in the abdominal area. Therefore, her diagnosis took longer because it was treated as a mere pre-menstrual cramps for years with combined hormonal contraceptives, masked the symptoms of the syndrome. It was assertively diagnosed after performing a CT scan, where images showed compression of the renal vein, ectasia of pelvic vessels associated with dilation of the gonadal vein. Our patient did not have hematuria, which is a typical symptom of this syndrome making diagnosis difficult. According Orczyk K et al., 2016 which 112 patients evaluated with nutcracker syndrome about 80% were above hematuria [10].

The diagnosis of nutcracker syndrome is made when it shows a reduction of left renal vein diameter is greater than 50% [11].

The ultrasound vascular Doppler can also be used for diagnosis, but it was not performed in this case.

Treatment of the syndrome remains controversial. Whether it should be treated surgically or not, should depend on the individual case and status of each patient and severity. In patients presenting symptoms acceptable as little abdominal pain, mild hematuria, may be tempted conservative treatment. In cases of severe anemia due to severe hematuria, significant pain and no improvement with common analgesics or functional renal impairment is indicated surgical treatment [12–15].

In clinical drug treatments, estrogens and anti-inflammatory drugs have been used, but have not achieved much success [16].

Petershank et al., 1970, was the first to propose surgical treatment [17] has since been appearing several new techniques for disease correction.

Surgery to left renal vein transposition has been demonstrated as an effective and safe procedure [9,18]. Nephropey excluding varicose veins is no longer recommended, because do not treat the
initial pathophysiology [9, 19]. The most invasive technique is renal auto transplantation, which is associated with better results and low morbidity and mortality rates, and some authors report that this treatment is the most effective [19]. Another form of surgical correction is made by ligation of the ovarian vein and varicose veins pelvic, via transabdominal or retroperitoneal even by laparoscopy [3].

Koyama A. et al. Published in 2004, had done conservative treatment and there was improvement in hematuria and proteinuria [24]. In the study by Ali-E.I-Dein et al. three patients were treated with stent an six with renal autotransplantation, all were free of pain [25]. Feng Hua et al. One patient treated with transposition of the left renal vein, three with conservative treatment and two with transposition of the superior mesenteric artery, all remained without symptoms. [26]

For our patient a stent through the endovascular treatment was proposed, which many studies show efficacy and safety, being less aggressive choice and more appropriate in this case [20].

4. Conclusion

The diagnosis of nutcracker syndrome gets done after exclusion of other causes of chronic pelvic pain, such as pelvic inflammatory disease, endometriosis, interstitial cystitis, pelvic tumors or inflammatory bowel disease.

As the clinical presentation present in most cases, macroscopic or microscopic hematuria, was not observed in this case, making it more difficult diagnosis.

Therefore there is a greater need for dissemination and understanding of the syndrome and its clinical presentation, between the main specialties involved with chronic pelvic pain, especially gynecology, urology. Knowing the clinical variants can also present like in this case, without hematuria.

Conflicts of interest

The authors have no conflicts of interest to declare.

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Luis Felipe Couto Novaes - study concept/ data analysis.
Leonardo Neves Saguia - writing the paper.
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