

A Successful Endourological Treatment of Hematuria in Nutcracker Syndrome

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1. Abstract

1.1. Background: The Nutcracker Syndrome (NS) consists of the compression of the left renal vein causing a regime of high venous pressures that affected the retroperitoneum. Hematuria is the most common clinical symptom. The clinical investigation is carried out with Doppler Ultrasonography or Abdominal Angiotomography. The treatment is based on the measurement of symptoms, and invasive treatments are reserved for symptomatic patients who present severe symptoms or are refractory to conservative treatment. Age is an important factor in clinical management, and patients under 18 years-old have a tendency to conservative approach based on clinical observation and lifestyle changes.

The importance of this case report is to present a minimally invasive alternative treatment as an option (flexible ureteroscopy with cauterization of renal papillae) to open surgery and endovascular procedures.

1.2. Case summary: We present a case of an adolescent (17 years old) with NS and macroscopic hematuria. After 7 days with conservative treatment, he was underwent to Urethrocytoscopy and Ureterorenoscopy, which identified a hematuric jet in the left ureteral meatus, and has been treated with flexible ureterorenoscopy with fulguration of the hemorrhagic renal papillae. As a complementary treatment, a high calorie diet was recommended for 3 months and was advised to avoid physical activities.

No recurrences or symptoms after one-year follow-up.

1.3. Discussion: Nutcracker Syndrome treatment is based on patient characteristics and severity of symptoms, and includes conservative treatment, open surgery and endovascular surgery. This case report presents an alternative conduct to expand minimally invasive options for treatment hematuria in Nutcracker Syndrome.

2. Introduction

The Nutcracker Syndrome (NS) is the clinical expression of left renal vein (LRV) compression. Venous hypertension in the left renal vein could cause macroscopic hematuria, varicocele on the left side, flank pain and orthostatic proteinuria [2].

Hematuria is the most common clinical symptom, its spectrum ranges from microhematuria to severe macroscopic hematuria [2]. The anatomical description of the pathology was made by Grant in 1937, however the clinical description with the current name of the syndrome was elaborated almost 40 years later in 1971 by Chait [3].

The most frequent cause of NS is the compression of the LRV between Abdominal Aorta and Superior Mesenteric Artery (SMA), whilst the compression of the LRV between spine and Aorta is called the Posterior Nutcracker [3].

There are no specific clinical diagnostic criteria, so it is necessary to exclude other more common causes of hematuria and flank pain [4], such as nephrolithiasis, urinary tract infection and neoplasms [5]. In general, the diagnosis is made around the second or third decade of life, and the methods used are abdominal Doppler Ultrasound (DUS), Computed Tomography Angiography (Angio CT) of the abdomen and pelvis, Contrasted Magnetic Resonance imaging of the abdomen and pelvis and, lastly, Phlebography [6].

The mesenteric-aortic angle found in imaging exams varies classically from 38° to 56°, when there is an interposition of the Duodenum between aorta and Superior Mesenteric Artery (SMA), the pathological angle can be greater [3].

The difference in diameter between the LRV extended to the narrow portion is 4 to 1, and the pressure gradient between inferior vena cava and left renal vein found in the DUS is between 3 and

7 mmHg. Finally, the caliber of the gonadal vein is greater than 3mm [3].

Treatment may vary due to severity of symptoms. Conservative management is usually destined for young patients and those with mild symptoms. Otherwise, surgical and endovascular approaches are chosen for those with severe symptoms or who do not improve after conservative treatment [7].

This case report describes a patient with NutCracker Syndrome that has been treated successfully with use of minimally invasive technique.

3. Case Report

We present the case of a male patient, 17 years old, student, who was admitted to the emergency room due to episodes of urine bleeding with clots elimination 7 days ago with worsening complaints in the last 24 hours. He had difficulty urinating before the clots left the urine. He denied a history of trauma, fever, polyuria or dysuria. No known comorbidities.

On physical examination, he was 1.80 m tall and weighed 55 kg (BMI 17kg/m²), with no signs of palpable varicocele in the Valsalva maneuver. No other alterations in the propaedeutic examination. Primary laboratory tests revealed hemoglobin of 11 g/dL. No other changes. The initial approach was to hydrate and perform angiotomography of the abdomen and pelvis. Angiotomography is the result of union between the angiography and tomography and It's a diagnostic imaging test which uses high technology to obtain visualization of the body's vein and arteries, through administration of intravenous contrast. The aortomesenteric angle on the exam was remarkable, as it was 25° and the left renal vein was being compressed, causing a sharp tapering when passing between SMA and the aorta (Figure 1 and 2).

The patient was admitted and duly evaluated by a multidisciplinary team of vascular surgeons, nephrologists and urologists. Conservative clinical treatment was adopted with rest and bladder irrigation for 7 days.

With no improvement in hematuria on the seventh day, but maintaining hemodynamic stability throughout hospitalization, the patient had a drop in hemoglobin from 11.3g/dL to 9.2g/dL. Urethroscopy and ureterorenoscopy were chosen to rule out other causes of hematuria before the vascular procedure. During the urethroscopy, a hematuric jet was identified in the left meatus (Figure 3). In ureterorenoscopy, the presence of bleeding in the renal papillae was detected (Figure 4). We opted for the fulguration of the hemorrhagic renal papillae with Holmium Laser with 4 mJ and 15 Hertz, in addition to the placement of a double J catheter for 2 weeks.

Hematuria has ceased on the first postoperative and has hospital discharged on the second day. A high calorie diet for 3 months was recommended because, together with collateral veins, the growth of adipose tissue close to the origin of Superior Mesenteric Artery and between it and Left Renal Vein may reduce compression in LRV, collaborating in remission from symptoms [1, 9]. Besides, the patient was advised to avoid physical activities because this could increase hematuria in Nutcracker Syndrome and it would retard weight gain.

After one year, he did not present new episodes of hematuria, maintaining a hemoglobin of 15 g/dL in laboratory tests, with no signs of hematuria in the urine test and the patient's current weight is 59 kg.



Figure 1: Axial abdominal CT angiography - compression of Left Renal Vein (LRV) between the Aorta and the Superior Mesenteric Artery (SMA). "Beak sign": an abrupt narrowing of the LRV between the Aorta and SMA, with proximal dilatation of the LRV.



Figure 2: Sagittal abdominal CT angiography - compression of Left Renal Vein (LRV) between the Aorta and the Superior Mesenteric Artery (SMA).



Figure 3: Left ureteral meatus flagrant showing macroscopic hematuria.



Figure 4: Macroscopic hematuria originating from left kidney's renal papilla.

4. Discussion

The Nutcracker Syndrome has as main anatomical alterations the compression of LRV in its path between the abdominal aorta and SMA, caused by decreasing the angle and distance between these two vessels. In the clinical case, the patient had the most common clinical manifestation of the syndrome, macroscopic hematuria [2].

There are no specific clinical diagnostic criteria, so diagnosis is made after exclusion of other more common cases of hematuria [4] and flank pain, like nephrolithiasis [5].

DUS is the most widely used method in the assessment [8]. Angio CT can be used as a very reliable non-invasive method to characterize the anatomy of the Nutcracker phenomenon, making it possible to infer the diagnosis of Nutcracker Syndrome [3]. In this case reported, US and doppler were not performed initially for this patient because Angio TC and urotomography are more available in São Paulo and are less dependent operator.

Currently, the medical literature proposes several treatment options. Conservative management is usually destined for young patients (under 18 years old) and those with wild symptoms, for 24 months. It includes weight gain because, with the development of venous collaterals, the growth of adipose tissue close to the ori-

gin of Superior Mesenteric Artery and between it and Left Renal Vein release the entrapped LRV, collaborating in remission from symptoms [1, 9]. Orthostatic proteinuria can be reduced with angiotensin-converting enzyme inhibitors [9]. Younger patients will have a higher rate of resolution with conservative measures than adult patients.

Surgical and endovascular approaches are chosen for those with severe symptoms or who do not improve after conservative conduct [7]. Severe symptoms are considered intense hematuria (with or without anemia), intense pelvic or abdominal flank pain, symptoms persisting for more than 6 months in adults and 24 months in those less than 18 years old [10].

Open surgery for anterior Nutcracker Syndrome includes: Transposition of the LRV, Left kidney autotransplantation, Transposition of the SMA, Nephropexy, Nephrectomy, Renocaval bypass and Transposition of the LGV [1]. Laparoscopic techniques have been increasing in popularity as minimally invasive surgery techniques improve [11].

Open surgery is indicated to treatment posterior Nutcracker Syndrome: Excision of the LRV, leaving a small margin from the Inferior Vena Cava (IVC) wall, translocation of LRV to a position anterior of the aorta, and reimplantation to the IVC in a superior position than the previous location, in order to relieve the pressure [12, 13].

Endovascular surgery (Stents and embolization of Left Gonadal Vein) has been increasingly used and involves less risk of morbidity and of complications than open surgeries [14].

Currently, endoscopic techniques have allowed more accuracy and less invasive diagnosis in the treatment of chronic unilateral hematuria (CUH) [15]. The first choice to endoscopic evaluation is the cystoscopy followed by rigid ureteroscopy, flexible ureteroscopy, combination of flexible ureteroscopy, semi-rigid ureteroscopy and the digital flexible ureteroscopy [15]. Actually, the association between flexible ureteroscope and semi-rigid ureteroscope with diathermy fulguration or laser ablation is the diagnostic and therapeutic technique of choice, with a high overall success rate (93%) and a low recurrence rate (10%) [15]. Nonetheless, there is not a consensus for the follow-up [15].

In this case report, the patient was submitted to urethrocystoscopy and ureterorenoscopy before the vascular procedure, and after conservative clinical treatment with rest and bladder irrigation for 7 days. During the urethrocystoscopy, a hematuric jet was identified in the left meatus (Figure 3). In ureterorenoscopy, the presence of bleeding in the renal papillae was detected (Figure 4). The fulguration of the hemorrhagic renal papillae with Holmium Laser was performed and inserted a double J catheter for 2 weeks. Apart from that he was instructed to ingest a high calorie diet for 3 months in order to increase retroperitoneal adipose tissue and had gained 4 kg during one year.

No new episodes or symptoms after one year of follow-up.

In fact, the weight gain and endourological treatment with laser were effective to solve the hematuria in this patient, and could be a minimally invasive alternative to open surgery or endovascular treatment in young patients with mild hematuria.

References

1. Macedo G L, Santos M A, Sarris A B, Gomes R Z. Diagnosis And Treatment Of The Nutcracker Syndrome: A Review Of The Last 10 Years. *J. Vasc. Bras.* 2018; 17(3).
2. Hohenfellner M, Steinbach F, Schultz-Lampel D, et al. The Nutcracker Syndrome: New Aspects Of Pathophysiology, Diagnosis And Treatment. *J Urol.* 1991; 146(3): 685-8.
3. Kurklinsky Ak, Rooke Tw. Nutcracker Phenomenon And Nutcracker Syndrome. *Mayo Clin Proc.* 2010; 85(6): 552-9.
4. Venkatachalam S, Bumpus K, Kapadia Sr, Gray B, Lyden S, Shish-hbor Mh. The Nutcracker Syndrome. *Ann Vasc Surg.* 2011; 25(8): 1154-64.
5. Novaes Lf, Silva Saguia Ln, Di Migueli Ca, et al. Young Woman With Nutcracker Syndrome Without Main Clinic Manifestation: Hematuria-Case Report. *Int J Surg Case Rep.* 2017; 31: 225-8.
6. Miró I, Serrano A, Pérez-Ardavín J, et al. Eighteen Years Of Experience With Pediatric Nutcracker Syndrome: The Importance Of The Conservative Approach. *J Pediatr Urol.* 2020; 16(2): 218.E1-6.
7. Daily R, Matteo J, Loper T, Northup M. Nutcracker Syndrome: Symptoms Of Syncope And Hypotension Improved Following Endovascular Stenting. *Vascular.* 2012; 20(6): 337-41.
8. Zucker EJ, Ganguli S, Ghoshhajra B, Gupta R, Prabhakar Am. Imaging Of Venous Compression Syndromes. *Cardiovasc Diagn Ther.* 2016; 6(6): 519-32.
9. Ananthan K, Onida S, Davies AH. Nutcracker Syndrome: An Update On Current Diagnostic Criteria And Management Guidelines. *Eur J Vasc Surg.* 2017; 3(6): 886-94.
10. Avgerinos ED, Mccanney R, Chaer R. Surgical And Endovascular Interventions For Nutcracker Syndrome. *Semin Vasc Surg.* 2013; 26(4): 170-7.
11. He Y, Wu Z, Chen S, et al. Nutcracker Syndrome—How Well Do We Know It? *Urology.* 2014; 83(1): 12-7.
12. Skeik N, Gloviczki P, Macedo TA. Posterior Nutcracker Syndrome. *Vasc Endovascular Surg.* 2011; 45(8): 749-55.
13. Said Sm, Gloviczki P, Kalra M, et al. Renal Nutcracker Syndrome: Surgical Options. *Semin Vasc Surg.* 2013; 26(1): 35-42.
14. Hulsberg PC, McInerney E, Partovi S, Davidson JC, Patel IJ. Minimally Invasive Treatments For Venous Compression Syndromes. *Cardiovasc Diagn Ther.* 2010; 6(6): 582-92.
15. Tanimoto R, Kumon H, Bagley DH. Development Of Endoscopic Diagnosis And Treatment For Chronic Unilateral Hematuria: 35 Years Experience. *J Endourol.* 2017; 31(S1): S76-S80.
16. Wang SZ, Zhang WX, Meng QJ, Zhang XP, Wei JX, Qiao BP. Laparoscopic Extravascular Stent Placement For Nutcracker Syndrome: A Report Of 13 Cases. *J Endourol.* 2015; 29(9): 1025-9.