



Case Report

Utility of Point-of-Care Ultrasonography in the Diagnosis of the Nutcracker Syndrome

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Abstract

A 43-year-old man was evaluated because of left flank pain from three years ago. Laboratory tests were normal so a point-of-care ultrasound of the abdomen was performed, showing dilation of the left renal vein with an abrupt narrowing between the aorta and the superior mesenteric artery. These findings suggested a left renal vein entrapment syndrome or nutcracker syndrome. The nutcracker syndrome is an entrapment of the left renal vein at the aortomesenteric clamp, leading to hyperpressure at the left kidney and the left gonadal vein. It can cause hematuria, left flank pain, orthostatic proteinuria and gonadal symptoms (pelvic congestion syndrome in women and scrotal pain and varicocele in men). Although there are not clear diagnosis criteria, abdominal ultrasound is the diagnostic test that has proven most useful. However, a high clinical suspicion is important, since similar ultrasound findings can be encountered in the general population.

Introduction

We present a case of a man who complained of intermittent left flank pain from three years ago, and who had undergone several tests without establishing a diagnosis. We performed a point-of-care ultrasound at the emergency department which showed dilation of the left renal vein. These findings were compatible with a Nutcracker Syndrome. The Nutcracker syndrome is an entrapment of the left renal vein between the aorta and the superior mesenteric artery, causing increased pressure in the left renal vein and the left gonadal vein. It can cause left flank pain, hematuria and pelvic congestive symptoms. The nutcracker syndrome is not a prevalent health problem. However, rare entities must be considered once the most common conditions have been ruled out, since it can save the patients' years of testing. The rapid spread of Point-of-Care Ultrasound at the emergency departments and primary care facilities is helping to widen the differential diagnosis of various clinical syndromes.

Case

A 43-year-old man presented with intermittent left flank pain irradiated to the umbilical region that started three years earlier. The pain worsened after eating but did not improve with defecation. He did not associate fever, nausea, vomiting, hyporexia or any other symptoms. Prior imaging tests had all been normal. On physical examination, blood pressure, heart rate and temperature were normal. Body mass index was low at 17 kg/m². He presented tenderness of the left flank and periumbilical region, without any signs of peritoneal irritation.

Investigations

Blood analysis was normal. Urine sediment examination showed microhematuria with 1-5 red blood cells per high power field. We performed a point-of-care ultrasound, on which we found an enlargement of the left renal vein of 13.2 mm proximal to the aorto-mesenteric clamp with an abrupt narrowing of the LRV

between the abdominal aorta and the superior mesenteric artery. The normal diameter of the left renal vein is 4 to 5 mm [1]. The aortomesenteric distance and aortomesenteric angle were 3.6 mm and 19°, respectively. The normal aortomesenteric distance is 16 ± 6 mm, and the normal angle $51 \pm 25^\circ$. Both kidneys were normal with good corticomedullary differentiation. The left kidney measured 95.4 mm long and the right kidney, 98.6 mm. We did not identify pyelocaliceal dilation or kidney stones.

Differential Diagnosis

Given the clinical presentation, the normal blood work-up and the ultrasound imaging, we suspected a nutcracker syndrome. Other conditions such as nephrolithiasis, renal cell carcinoma and urinary tract infection were excluded.

Treatment

Since the patient had mild symptoms and did not present any warning signs, we agreed on conservative treatment and surveillance.

Outcome and Follow-Up

The patient was referred to his primary care physician for follow-up. He was instructed to come back to the emergency department if the symptoms worsened or if he presented with fever, incapacitating pain or incoercible hematuria.

Discussion

The nutcracker syndrome, also known as left renal vein entrapment syndrome, is caused by compression of the left renal vein (LRV) between the abdominal aorta and the superior mesenteric artery (SMA). The SMA has its origin at the abdominal aorta, and it usually forms an acute angle. The left renal vein, the pancreatic neck and the duodenum, in this order, pass between the abdominal aorta and the SMA. In some patients, this space is reduced, compressing the left renal vein (LRV) causing dysfunction of the venous system and venous congestion on the LRV territory, which can lead to renal and gonadal symptoms [1-3] (Figure 1-7).



Figure 1: Sagittal view showing the origin of the superior mesenteric artery at the abdominal aorta.

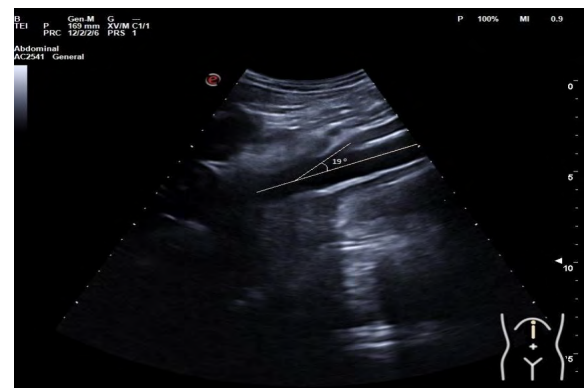


Figure 2: Superior mesenteric artery forming an acute angle at its origin at the abdominal aorta.



Figure 3: Axial view of the left renal vein passing between the SMA and the aorta, showing the “beak sign”, an abrupt narrowing of the LRV. AMS: superior mesenteric artery. VRI: left renal vein. Ao: aorta.



Figure 4: Axial view of the left renal vein at the aortomesenteric clamp. D1 is the diameter of the left renal vein (13.2 mm). D2 is the aortomesenteric distance, which is reduced to 3.6 mm, while the mean aortomesenteric distance in healthy individuals is 16 mm [1].



Figure 5: axial view of the LRV between the aorta and the superior mesenteric artery in a healthy individual (aortomesenteric distance 10.3 mm).

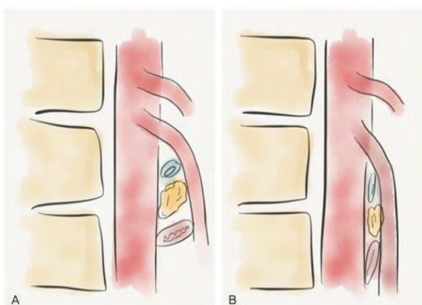


Figure 6: A. sagittal view representing a healthy subject. From cranial to caudal, the left renal vein, the neck of the pancreas and the third portion of the duodenum pass between the SMA and the abdominal aorta. B. Sagittal view. Representation of the nutcracker phenomenon with a narrowing of the aortomesenteric space.

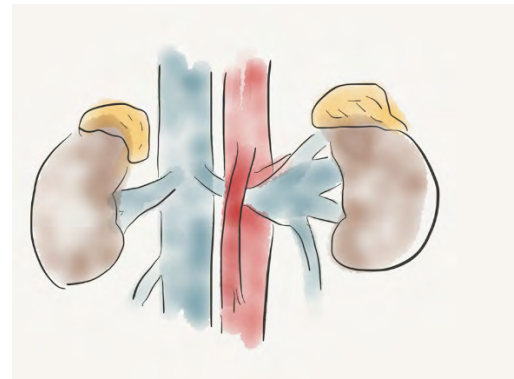


Figure 7: coronal view representing the origin of the SMA at the abdominal aorta, with the LRV passing between them before draining into the vena cava. In this image we can see the dilation of the proximal LRV due to its compression at the aortomesenteric clamp.

It is important to establish the difference between the nutcracker phenomenon and the nutcracker syndrome. The nutcracker phenomenon refers to the dilation of the LRV proximal to the aortomesenteric clamp, which is a relatively common finding encountered in 10 to 14% of the general population [4]. The nutcracker syndrome is the presence of clinical symptoms associated with a compatible anatomy. Nutcracker syndrome is an uncommon entity, although it is thought to be underdiagnosed due to its nonspecific clinical presentation and to the lack of established diagnostic criteria [2,5]. It is more common in the third and fourth decades of life and, even though it was thought to be more prevalent in women, the latest studies suggest that there is a similar prevalence in both men and women [3]. It is well documented that men are diagnosed earlier than women [6,7]. The normal aortomesenteric distance in healthy individuals is 16 ± 6 mm with an angle of $51 \pm 25^\circ$ between the aorta and the superior mesenteric artery at its origin [1]. The posterior nutcracker syndrome is a variant produced by a retroaortic or circumaortic left renal vein as an anatomic variant [8]. The compression of the left renal vein at the aortomesenteric clamp causes increased pressure on the ipsilateral kidney that is transmitted to the renal calyces, which can break into the collector system causing hematuria [9,10]. The nutcracker syndrome can also cause orthostatic proteinuria, although the physiopathology of this symptom is not well understood [1].

The increased pressure on the left venous system can also cause pelvic symptoms, since the left gonadal vein drains into the LRV while the right gonadal vein drains directly into the vena cava. The most common finding is scrotal pain and varicocele in males and pelvic congestion syndrome in women [11]. Hematuria is the most common symptom, presenting in 80% of patients, followed by left flank pain (40%) and varicocele (35%) [6].

Diagnosis must start with a thorough clinical history and physical examination and must be completed with diagnostic imaging [1]. A high clinical suspicion is important for imaging interpretation and to avoid overdiagnosis, given the high prevalence of the nutcracker phenomenon as an anatomic variant among the general population [12]. Given its accessibility, non-invasiveness and elevated specificity and sensitivity, Doppler ultrasound should be the first diagnostic test [13]. CT and MRI can be used to confirm ultrasound findings. Cystoscopy can be useful as well to demonstrate hematuria with origin at the left ureter [14].

Standardized diagnostic criteria are lacking, which in addition to the increased availability of imaging testing, can lead to overdiagnosis [4]. Treatment varies according to the degree of clinical severity. In patients presenting with mild-to-moderate symptoms, surveillance is indicated. Some authors recommend AAS and angiotensin-converting enzyme inhibitors to improve renal perfusion and to reduce proteinuria. In patients presenting with severe or incapacitating pain, hematuria causing anaemia, proteinuria not responding to pharmacological treatment or renal failure, surgical treatment must be considered [15,16]. Surgical techniques range from venous stent implantation to open bypass and even nephrectomy [16]. In patients under 18 years old, conservative treatment is recommended for at least two years since spontaneous resolution during growth is not uncommon [15]. The increased availability of diagnostic tests and less-invasive therapeutic options like venous stents are leading to an increase in surgically treated patients. However, it is well established that the disease is often self-limiting even without treatment. In addition, there is little clinical experience with the new treatments, with long-term side effects remaining unknown [4].

Learning Points/Take Home Messages 3-5 Bullet Points

- The nutcracker syndrome is an uncommon entity that can cause hematuria and left flank pain, and must be considered once more common alternative diagnosis have been ruled out.
- Doppler ultrasound is the most useful imaging test for the diagnosis of the nutcracker syndrome, so Point-of-Care ultrasound can be a useful diagnostic approach on the differential diagnosis of abdominal pain and hematuria.
- A high clinical suspicion is necessary for the diagnosis of the nutcracker syndrome, given the lack of standardized diagnostic criteria and the high prevalence of the nutcracker phenomenon on the general population.
- Treatment should always be individualized according to the patient's symptoms and beliefs. For most patients, the most adequate treatment is surveillance.

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