

Exploring vascular shadows: Case report of young adult nutcracker syndrome

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Abstract

Nutcracker syndrome is defined as an uncommon vascular condition where the left renal vein is compressed between the superior mesenteric artery and aorta, causing renal vascular congestion. This can lead to renal dysfunction and symptoms such as hematuria, proteinuria and orthostatic hypotension. Its prevalence is low, approximately 1 in 2,500 people, and it has no prevalence by sex, which makes its diagnosis challenging due to clinical variability and lack of specific criteria. Therefore, the importance of early diagnosis and treatment to prevent serious complications is emphasized. The main objective of this report was to describe the natural history and the diagnostic-therapeutic approach of a case of Nutcracker Syndrome in a 25-year-old young patient who had been consulted for persistent pelvic pain. Transvaginal ultrasound identified pelvic varices and abdominal angiotomography and venography studies confirmed left renal vein compression with venous dilation, findings typical of nutcracker syndrome. The Doppler confirmed extrinsic compression and decreased venous flow. Given the absence of severe symptoms and clinical stability, management was conservative, with half-yearly controls and follow-up by nephrology, without the need for pharmacological or surgical treatment. This syndrome may resolve spontaneously or develop into complications such as persistent hematuria, proteinuria and chronic pelvic pain. Conservative treatment is appropriate in mild cases without renal involvement, but endovascular or surgical interventions should be considered in patients with severe or persistent symptoms.

Keywords: Conservative Treatment; Hematuria; Nutcracker Syndrome; Pelvic Pain; Phlebography

1. Introduction

Nutcracker syndrome (NCS) is a rare condition characterized by extrinsic compression of the left renal vein (LVD) between the superior mesenteric artery (SMA) before and the aorta afterwards. This understanding results in renal vascular congestion, clinically manifested with symptoms such as hematuria, proteinuria, orthostatic hypotension, pain and even renal dysfunction. Prolonged venous compression can stimulate collateral drainage pathways through the gonadal and pelvic veins, which may explain the overlap of other syndromes such as pelvic congestion syndrome (1).

Although the exact prevalence is not well established, it is estimated to affect approximately 1 in 2,500 people. It is important to note that the prevalence of nutcracker syndrome is similar between men and women. Diagnosis can be challenging due to variability in clinical presentation and lack of specific diagnostic criteria (2).

The main objective of this case is to analyze the natural history of a rare disease and promote knowledge about it.

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2. Presentation of the case

A 25-year-old female patient born in Barranquilla/Atlántico with a possible diagnosis of nutcracker syndrome with history of hospital admission for approximately one year of evolution (June 2023), characterized by pelvic pain.

In June of last year, the patient is performed a transvaginal ultrasound control where they return results of pelvic varices, she is referred to perform an abdomen Angiotac on July 6, 2023, where it shows reduction of aorto-anglemesenteric that causes compression of the left renal vein which is slightly dilated. Which coincides with a probable nutcracker syndrome.

The patient reports no other discomfort, however, on January 19 of this year a venogram is sent to confirm the aforementioned diagnosis where findings in the left renal vein are suggestive of nutcracker syndrome.

In February of this year, a mesenteric abdominal vessel Doppler was performed and results were obtained from an abrupt decrease in the calibre of the left renal vein and extrinsic compression between the superior mesenteric artery and the aorta, confirming the patient’s diagnosis.

After this, on February 21, 2024, an ultrasound of the urinary tract was performed where dilation of both renal pelvis was shown and following this examination on March 8 of this year a Doppler of mesenteric abdominal vessels was again.

performed which showed a decrease in the flow rates of the left renal vein compatible with nutcracker syndrome, On March 11, a renal artery Doppler is performed, which confirms the decrease in flow rates of the left renal vein.

Following these procedures, in the same month she is referred to gynecology where they find varicocele pelvic and decide her referral to vascular surgery for diagnostic confirmation, control. Where she was then referred to nephrology for control and treatment.

You are denied pathological, allergic, gynecosurgical, hospital, surgical and traumatic history, for treatment they send you control with nephrology every six (6) months, urinary computed tomography of control and control tests as albumin in serum, creatinine, complete hemogram, uric acid, sodium, calcium, potassium, phosphorus, PTH and uroanalysis.

To the results of the above-mentioned paraclinics, the patient in June last year (2023) has a slight increase in urea. In February of this year (2024), a urine analysis was performed showing the presence of bacteria (+2), no hematuria or proteinuria and a complete blood count showed monocytes, eosinophils and elevated urea.

Table 1 Laboratories

Admission	
Tests	Results
Creatinine	0.8 mg/dL
Glycemia	85 mg/dL
Urea	26 mg/dL
Hemoglobin	13.20 g/dL

Table 2 Uranalysis

Uranalysis	
Tests	Results
Color	Yellow
Turbidity	Clear
pH	5.5
Specific gravity	1.021
Nitrites, glucose, ketone bodies, urobilinogen, bilirubin, mucus.	Negative
Cédulas epithelialize	3.0
Squamous epithelial cells	2.4
Transitional epithelial cells - Tran EC	0.0
Renal tubular epithelial cells (RTEC)	0.5
Leukocytes	3.3
Erythrocytes	0.7
Bacteria	+2
Casts, hyaline casts, pathological casts	Negative
Non-squamous epithelial cells	0.5
Proteinuria in 24-hour urine	0.1
Proteinuria	4.00
Microalbuminuria	7.35MG/G

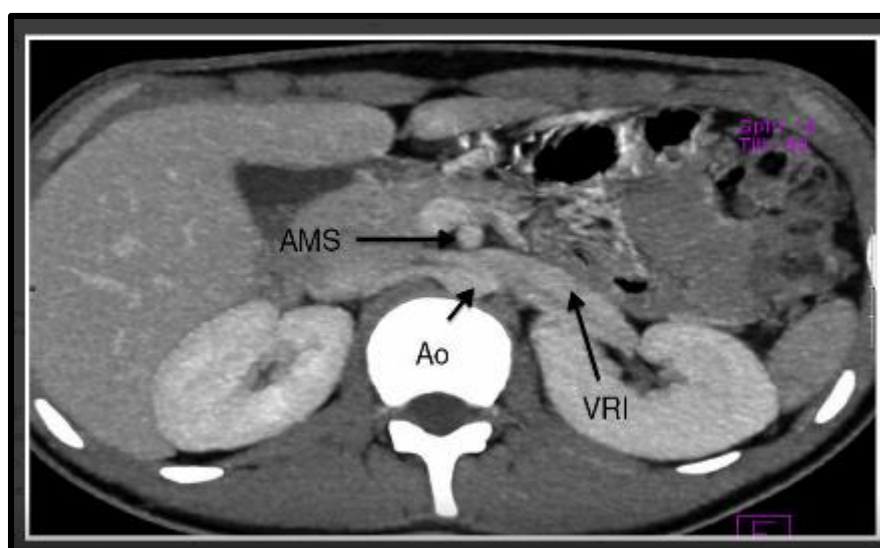
Table 3 Complete Blood Count (CBC)

Complete Blood Count (CBC)	
Laboratory parameters	Results
Hemoglobin	14 g/dL
Red blood cell count	4.34 mcl
RDW-wide distribution of erythrocytes	11.10%
Hematocrit	41.90%
Mean corpuscular volume	96.50 fl
Corpuscular hemoglobin	32.30 pg
Total leukocyte count	5.90
Neutrophils	2.74 ml (46.50%)
Lymphocytes	1,46 ml (24.70%)
Monocytes	0.75 ml (12.70%)
Eosinophils	0.88 ml (14.90%)
Basophils	0.06 ml (1.00%)
Platelet count	223.000 mcL

Platelet volume	9.70 fl
IG percentage	0.20
Ionized calcium	1.28 mg/dL
Creatinin	0.7 mg/dL
Phosphorus	3.76 mg/dL
BUN	17.40 mg/dL
potassium	4.31 mg/dL
Albumin	4.35 mg/dL
Proteins in urine	0.1g/24h
Sodium	137 mg/dL
Urea	37 mg/dL
Urinary volume	2460 L

3. Discussion and review of literature

Nutcracker syndrome (NCS) is a rare condition characterized by extrinsic compression of the left renal vein (VRI) between the anterior superior mesenteric artery (AMS) and the posterior aorta. This understanding results in renal vascular congestion (3).



Note. There is an increase in the diameter of the left renal vein.

Figure 1 Renal Angio tomography

There is an increase in the diameter of the left renal vein. AMS: superior mesenteric artery; Ao: aorta; VRI: left renal vein (4)

What makes this clinical case striking is that it manifests in approximately 1 out of 2,500 people and this is where its medical importance lies. On the other hand, what has a greater impact on it is that this is a young patient, with no history of importance, no underlying pathologies, good healthy habits and above all the fact that it is an extremely unusual pathology which, if not addressed in an appropriate and timely manner, can trigger a real injury.

A clinical case of a 25-year-old female patient from the city of Barranquilla who consults a medical center for pelvic pain, which is why she is referred to gynecology where there are findings of pelvic varices. Subsequently, she is referred to the vascular surgeon where an AngioTAC and a venography are performed where she is diagnosed with Nutcracker syndrome.

Venography findings highlight the presence of a permeable left renal vein, with radiolucent defect in its secondary middle third due to extrinsic compressive effect on the superior mesenteric artery, associated with the presence of small intra-anal and perirenal varicose dilatations.

Taking into account the definition of NCS and the patient's findings obtained from venography, we can show that the diagnosis was made correctly, as stated by Dieleman F. and his collaborators in a study carried out in Amsterdam, Netherlands in 2023.

In this study, thirty-eight patients were identified through a personalized consultation of the local electronic medical record system. Of this group, 21 patients experienced symptoms, including pain in the flank, abdominal pain, hematuria and fatigue, along with duplex ultrasound findings and/or venography findings positive for NCS. Of the individuals diagnosed with NCS, 11 were treated with LRV transposition, based on a shared decision considering imaging results, duplex ultrasound, venography and the severity and impact of symptoms in daily life.

Due to patient preferences, shared decision-making or self-referral to a foreign hospital, 10 were treated conservatively. A nutcracker phenomenon was observed in another 17 patients, that is, those who had a dx per image but were clinically asymptomatic (5) (6).

As for the patient's approach, it was decided to keep her under observation and to carry out controls every 6 months without accompanying pharmacological treatment and without surgical intervention, based on a conservative treatment as proposed by Dieleman F. and collaborators in his study, in which half of the patients diagnosed with NSC received conservative treatment, the result of this study shows that conservative treatment for elderly patients with a wide variety of symptoms may also be the best option. However, this study presented a number of limitations, such as that some patients had been referred from another institution and therefore the complete evolution of symptoms was not available (5) (7). Likewise, the majority of patients in whom success has been used are in the pediatric population (8)

Regarding the implementation of new therapies for the treatment of NSC, it has been proposed the implementation of an endovascular treatment, which is minimally invasive and has presented multiple benefits, such as a rapid recovery time and resolution of symptoms, This was established by Kolber M. and collaborators in the review of a study of 75 patients who were placed a stent in the left renal vein (LRV), with an average follow-up of 55 months. Similarly, it is a technique that is being studied and does not offer safety in response to treatment and no evolution to complications, because a stent migration rate of 6.7% has been identified, either to ICV, right heart or retrograde to renal ilium, as opposed to the surgical treatment, which has shown greater safety (1)

4. Conclusion

From this case, it can be concluded that nutcracker syndrome is a rare medical condition characterized by extrinsic compression of the left renal vein between the superior mesenteric artery and aorta, resulting in renal vascular congestion. In the clinical case presented, it is particularly striking due to its manifestation in a young patient of 25 years who has no medical history in conservative treatment and this has significant underlying pathologies, and also with healthy habits. In particular, the medical importance of NCS is highlighted, since its manifestation is not frequent, the identification, proper and timely management of this pathology are crucial to prevent potentially serious complications. The diagnosis was made by angioma and venography, which showed a characteristic narrowing of the VRI. The patient in question was treated with caution, observation and protective care in some cases, as recommended in recent studies. In a study by Dieleman et al., a large proportion of patients diagnosed with SCN were treated without surgery, with good results in most cases. Despite advances in minimally invasive procedures, such as stent placement, they still carry the risk of complications such as stent migration. Therefore, the choice of treatment should be carefully considered, especially in young and asymptomatic patients such as this. Finally, SCN treatment should be individualized according to patient characteristics and clinical and radiological findings, follow-up and conservative treatment may be appropriate for some patients, but it is important to continue research and develop safer and more effective treatment.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interests in this section.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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