**Nutcracker syndrome: a case report**

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**ABSTRACT**

**Background:** The Nutcracker Syndrome is a compression of the left renal vein (LRV) by the abdominal aorta (AA) and the superior mesenteric artery (SMA). It is manifested by pain, hematuria, varicocele or lower or pelvic urinary symptoms. There are less than 200 cases published in the literature.

**Materials and methods:** The patient is a 15-year-old boy who presented with a 2-year progression of pain in the lumbar fossa and left flank. Ultrasonogram revealed dilated LRV and Doppler sonogram and multislice CAT detected obstruction, ectasia and increased blood flow at the LRV level.

**Results:** Given symptom intermittence and scant interference with patient’s daily activities, patient was left under surveillance with periodic control.

**Conclusions:** Incidence of this disease is low. It should be suspected in young men or women with symptoms of long duration and who present with no other suspicious pathology. Open surgery is presently the most indicated treatment and extensive follow-up is required to confirm long-term endovascular treatment results.

**Key words:** nutcracker syndrome, left renal vein, aortomesenteric compass, venous obstruction, endovascular treatment.

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**RESUMEN**

**Antecedentes:** El síndrome de cascanueces se produce por obstrucción de la vena renal izquierda (VRI) debida a atrapamiento entre la aorta y la arteria mesentérica superior (AMS). Se manifiesta por dolor, hematuria, varicocele o síntomas urinarios bajos o pelvianos. Existen menos de 200 casos publicados en la literatura.

**Material y métodos (informe del caso):** Se presenta el caso de un varón de 15 años con dolor en fosa lumbar y flanco izquierdo de 2 años de evolución. La ecografía encuentra VRI dilatada, y por ecografía-Doppler y TAC “multislice” se detecta obstrucción, ectasia y aumento de flujo de la VRI.

**Resultados:** Ante la intermitencia de los síntomas y la escasa interferencia en la actividad diaria del paciente, se decide asumir una conducta expectante, con controles periódicos.

**Conclusiones:** Enfermedad de baja incidencia, debe sospecharse en pacientes jóvenes, varones o mujeres, con síntomas de larga evolución sin otras afecciones que lo justifiquen. En la actualidad, la cirugía abierta es el tratamiento más indicado y se requiere mayor seguimiento para confirmar el resultado a largo plazo del tratamiento endovascular.

**Palabras clave:** síndrome de cascanueces, vena renal izquierda, compás aortomesentérico, obstrucción venosa, tratamiento endovascular, Argentina.
INTRODUCTION
Initially described by De Schepper in 1972 as the “left renal vein entrapment syndrome”, it occurs when the left renal vein (LRV) is compressed by the abdominal aorta (AA) and the superior mesenteric artery (SMA) due to the closing of the angle between those two arterial trunks. Other less frequent causes are venous vascular abnormalities such as retroaortic or circumaortic renal vein or arteries with variations in origin and tract. Extrinsic compressions due to retroperitoneal lesions such as splenic tumors are uncommon. It presents with hematuria, left lumbar or abdominal pain and less frequently with varicocele, chronic pelvic pain or dysuria.

MATERIALS AND METHODS
The patient is a 15-year-old boy with no relevant clinical or urological history, with a 2-year progression of left lumbalgia and dull intermittent pain in the left flank and hemiabdomen that increased with exercise. Patient never presented with macro or microscopic hematuria. Doppler ultrasonogram led to suspicion of the pathology (Image 1) and it was confirmed by multislice CAT upon detecting through both studies proximal dilatation and compression of the LRV with hyperflow at the level of the aortomesenteric compass from vein stenosis (Image 2). Neither varicocele nor collateral circulation was detected (Images 3 to 4).

RESULTS
With the consent of the patient and his parents, conservative treatment was decided upon. This involves periodic clinical, laboratory and imaging controls along with analgesics as requested. Future endovascular or surgical approach has not been discarded, and will depend on pathology progression.

DISCUSSION
The nutcracker syndrome appears when there is a reduction of the angle between the AA and the SMA. This abnormality can be congenital or associated with left renal ptosis, reduction of perirenal fat, accentuated lumbar lordosis and also with pregnancy. The presence of intrarenal arterial venous fistulas with flow and pressure increase in the renal vein is called the “pseudo-nutcracker effect”.

Physiopathology is very similar to the aortomesenteric compass syndrome in which the third portion of the duodenum is obstructed by these same vascular structures, resulting in high digestive obstruction. The authors have seen no reports of coexistence of the two pathologies in the literature.

This pathology is more frequent in young women. Clinically it can be silent or there can be macro or microscopic hematuria (66% of cases), pain in the left back flank or abdomen or congestive pelvic symptoms (from obstruction of the gonadal venous flow) such as chronic pain, dyspareunia, dysmenorrhea or dysuria. Pain and hematuria are exacerbated with orthostatism or exercise.

In the differential diagnosis, pathologies that cause pain and hematuria such as lithiasis, congenital vascular malformations, tumors, infections, parenchymal or urinary tract abnormalities, painful pelvic syndromes and organ or neighboring structure alterations must be excluded. In order to do so, routine studies should be resorted to (sonography, simple or contrasted radiology),
endoscopic studies (cystoscopy or ureteropyeloscopy) or more complex studies (tomography, magnetic resonance studies or angiography). Some of these diagnostic modalities can provide orienting data for nutcracker syndrome. Diagnosis is made through dilated LRV (sonogram), indirect signs of extrinsic compression of the urinary tract (excretory urogram), increase of LRV caliber, hyperflow and retrograde varicosities (Doppler sonogram), observation of renal vein compression from the aortomesenteric compass and resulting collateral circulation (computed multislice tomography has the best definition and reconstruction, as well as selectively showing the vascular system by subtraction or nuclear magnetic resonance), evidence of varicose veins and slow contrast wash in venous phase (arteriography) or by the determination of renocaval pressure gradient and direct LRV observation and of its varicose collaterals (retrograde phlebography is considered to be the gold standard). Renocaval gradient above 3 mm Hg confirms diagnosis, 1-3 mm Hg is borderline and under 1 mm Hg is normal. However, advanced cases with collateral circulation may not have hypertension but LRV will be dilated. Persistent or anemizing hematuria, lumbar or incapacitating pelvic pain, or the presence of severe congestive pelvic symptoms are treatment indications, with the exclusion of other differential diagnoses.

Surgical treatment can be endovascular (balloon dilatation or stent placement) and has immediate good results but lacks long-term follow-up and presents risk of thrombosis, tearing or prosthesis migration. Vein dissection can be carried out with open surgery (only in cases in which symptoms can be resolved with this), gonadocaval bypass (in patients with predominant pelvic symptoms), venous transposition in front of the SMA or behind the AA or arterial (high risk for ischemia in the entire area irrigated by the SMA), autotransplant (to the iliac fossa) or nephrectomy when there is severe organ damage. As conservative treatment, when the predominant symptom is hematuria, hypertension in the urinary tract with a perfusion pump has been proposed and is reported to be successful in select cases.

**CONCLUSIONS**

The nutcracker syndrome is a rare pathology that is manifested by pain and/or hematuria in patients of both sexes. Phlebography is the diagnostic gold standard and Doppler sonogram, tomography (preferably multislice) or magnetic resonance studies are also used as less invasive diagnostic tools. Open or endovascular surgery is carried out to both resolve obstruction and alleviate hypertension of the left renal vein.

**BIBLIOGRAPHY**