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Minimally invasive treatment of nutcracker syndrome in a young girl

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ABSTRACT

Nutcracker syndrome is a vascular anomaly consisting of external compression of the left renal vein by the superior mesenteric artery and the aorta artery. It may manifest with recurrent abdominal or pelvic pain, flank pain, macro- or microscopic hematuria, gonadal varices, or asymptomatic.

We present a 10-year-old female patient with chronic progressive pain of more than two years of evolution in the left flank and radiating to the pelvic area. A diagnosis of nutcracker syndrome was made. The surgical resolutuion consisted in the transposition of the left ovarian vein to the left iliac vein. The patient remains asymptomatic at one year of follow-up.

In selected patients, venous decongestion of the left renal vein to the inferior vena cava using the ovarian vein is a low-complexity therapeutic possibility with a low incidence of complications.

Keywords: *renal nutcracker syndrome; abdominal pain; hematuria.*

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INTRODUCTION

Nutcracker syndrome (NS) is the external compression of the left renal vein (LRV) by the superior mesenteric artery (SMA) and the aorta artery, which causes venous congestion in the left kidney and alternative venous drainage to the pelvic floor through the ovarian vein.[1] This syndrome is one of the differential diagnoses of chronic abdominal pain, especially in women. Treatment depends on the severity of symptoms. Numerous techniques have been described, such as renal vein reimplantation,[2] nephropexy, renal vein plasty, renal autotransplantation, meso-aortic transposition, endovascular stenting,[3] and extravascular stenting. Many of these surgical techniques have limitations and can lead to severe complications.[4]

In this case, we present the surgical resolution of NC in a pediatric patient, applying a minimally invasive technique to pediatric patients with a low incidence of complications and a short postoperative hospital stay.

CLINICAL CASE

We present a 10-year-old female patient with a history of duodenal membrane resection at the age of 2 days old and who remained hospitalized for 17 days for nutritional recovery. She presented with abdominal pain of more than two years of evolution, progressive, localized in the left flank, radiating to the pelvic area, often disabling, and that did not improve with standard analgesics. On physical examination, the abdomen was flat, soft, and not painful on palpation.

Abdominal radiography and abdominal ultrasound were performed without pathological images, and blood and urine laboratories were performed without particularities. Angiotomography showed a reduction of the aortomesenteric angle (17°), compressing the LRV with the presence of varicose veins of the utero-ovarian veins (*Figure 1*). Renal Doppler ultrasound showed increased blood flow velocity in the left renal vein, with decreased caliber at the level of the aortomesenteric compass (3.1 mm) for a hilar caliber of 6 mm. Renal scintigraphy was requested, showing both kidneys' preserved function (*Figure 2*).

The surgical resolution was decided with the Nephrology, General Surgery, and Pediatric Cardiovascular Surgery teams. Diagnostic catheterization was performed, which showed narrowing of the LRV with a pressure difference with the inferior vena cava of more than 3 mmHg and dilatation of the ovarian vein with valvular incompetence (*Figure 3*). The diameter of the ovarian vein was measured at the junction level with the external iliac vein of 4.5 mm. Due to the degree of narrowing of the LVR, endovascular stenting was ruled out.

Surgical technique

A combined approach was performed. The cavity was entered through the umbilical approach with an open technique, pneumoperitoneum was performed, and ports of 5 mm were placed in the right iliac fossa and left flank. An incision was made in the peritoneum, and a delicate

Figure 1. Angiotomography of the abdomen with reconstruction

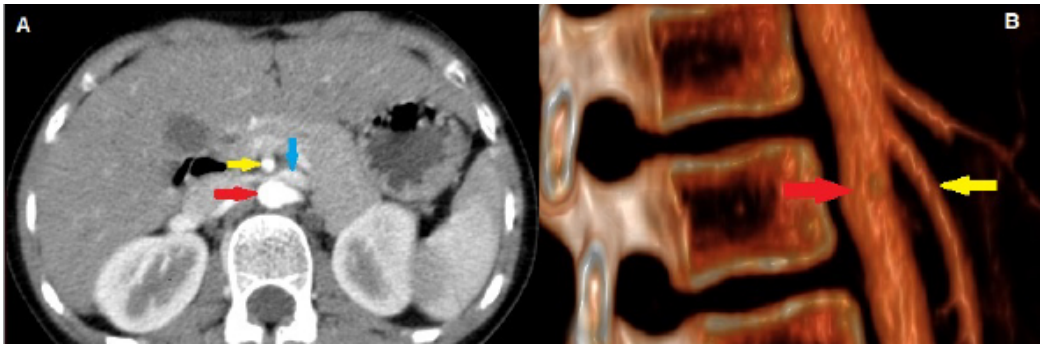


Figure A, coronal section showing external compression of the left renal vein (blue arrow) by the aorta (red arrow) and the superior mesenteric artery (yellow arrow).
Figure B, 3D reconstruction of the aortomesenteric angle, formed by the aorta artery (red arrow) and the superior mesenteric artery (yellow arrow).

Figure 2. Renal scintigraphy with normal uptake

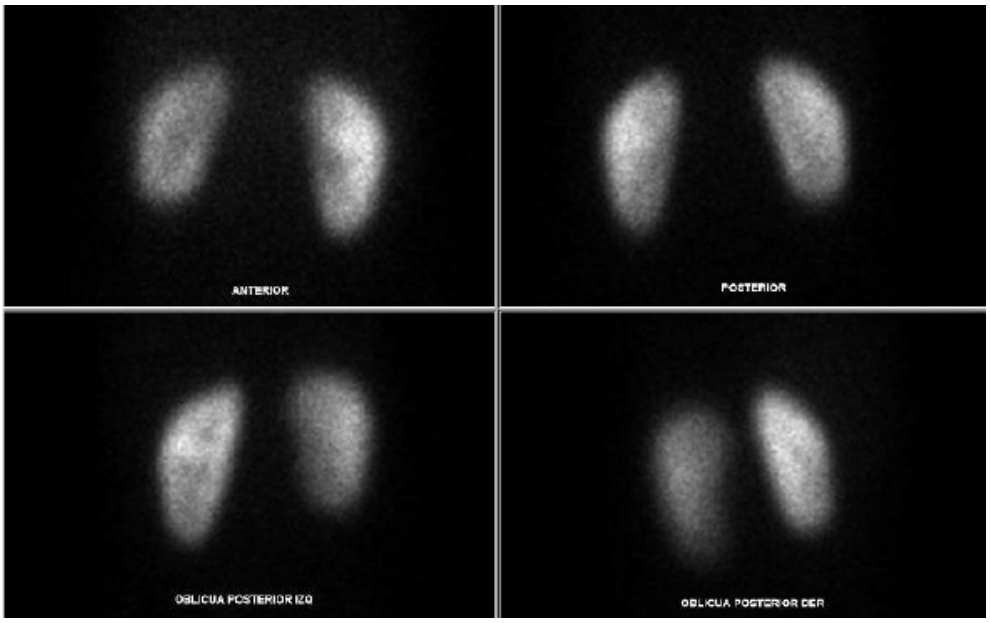
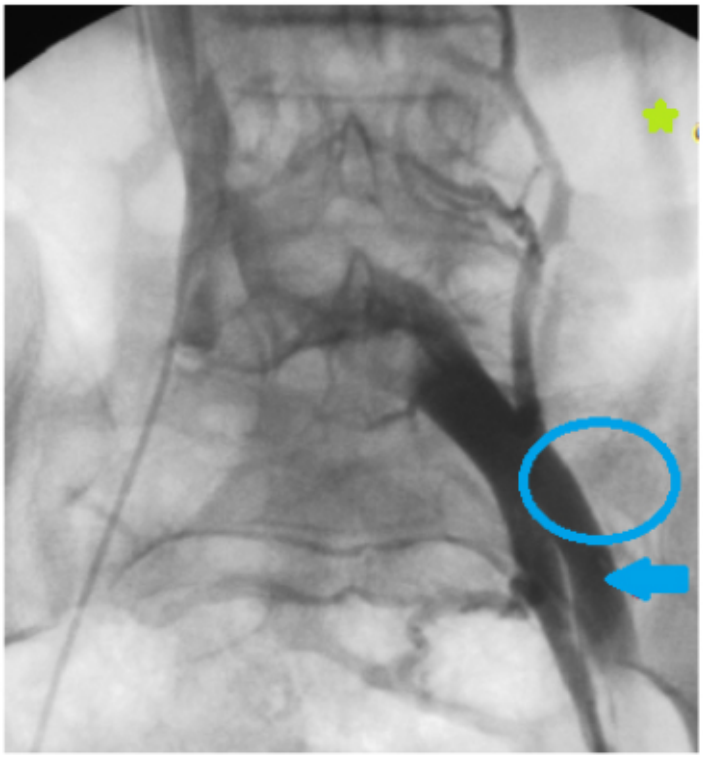


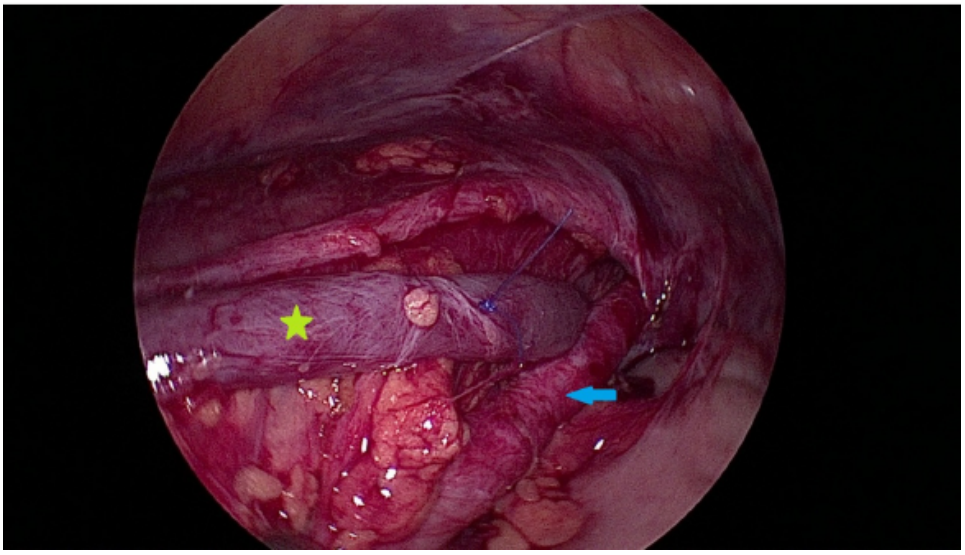
Figure 3. Diagnostic catheterization



The ovarian vein (green star) is shown crossed over the internal iliac vein (blue arrow). The blue circle shows the area where the terminolateral anastomosis is performed.

dissection of this vein was performed until it was individualized. It was left repaired with vessel loops. Subsequently, a transverse infra umbilical incision in the left iliac fossa of 4 cm was performed for surgical exposure of the distal gonadal vein and iliac vein. With the ovarian vein

Figure 4. Laparoscopic surgical imaging



Terminolateral anastomosis of the gonadal vein (green star) with the external iliac vein (blue arrow)..

completely freed, the proximal end of the gonadal vein was ligated and sectioned, and a clamp was placed on the distal end; the external iliac vein was clamped at the intersection with the gonadal vein, and terminolateral anastomosis of the distal end of the ovarian vein with the iliac vein was performed. Clamps were released, hemostasis was performed and closed as usual (*Figure 4*). Exploratory laparoscopy was performed to verify correct hemostasis.

The patient left the operating room extubated and in good hemodynamic condition. She recovered without complications and remained hospitalized for two days. Antiplatelet therapy with acetylsalicylic acid was indicated for six months.

At one year of follow-up, the patient remains asymptomatic. Doppler ultrasound shows adequate retrograde flow through the gonadal vein, with no signs of thrombosis.

DISCUSSION

Compression of the LRV between the aorta and the MSA is increasingly recognized as a differential diagnosis in women with pelvic and left flank pain. Initially, venous stasis in the LRV generates left flank pain and micro- or macrohematuria. As time goes by, dilatation of the left ovarian vein causes valvular insufficiency and reflux into the pelvis, producing pelvic pain.

Surgical indications for NS are as follows: 1) lack of improvement of symptoms two years after medical treatment; 2) complications, such as back pain, dizziness, weakness, and obvious varicoceles; and 3) signs of kidney damage, among others.[5]

The most common procedure performed in NS is the reimplantation of the LRV in a location below the inferior vena cava,[1] which can be performed by a transperitoneal or a laparosopic approach. These approaches have multiple disadvantages, particularly the limitation generated by the length of the LRV to reimplant it (with stenosis of this vessel). In addition, since the approach must be made through the abdominal midline, there are aesthetic complications inherent to the incision, which leaves a significant scar, considering the higher incidence of this disease in young women.

The endovascular approach has been described for treating NS, with excellent results, but in older patients.[3] The major limitation of this type of stenting procedure within the LRV is the size of the stent used and the extrinsic pressure it must tolerate. The follow-up time of this approach is limited, so the long-term consequences are unknown, considering the patients' 70- to 80-year survival.

Our procedure differs in that it is a relatively simple, aesthetic technique, presents no risk of eventrations or future obstetric complications, and does not exhaust other possibilities for resolution.[6] Venous blood flow in the NS is directed retrograde to the gonadal vein; it is dilated and with valvular insufficiency, so there is no change in the flow direction after the procedure. The proximity

of the gonadal vein to the external iliac vein suggests that this procedure could be performed by minimally invasive robotic surgery.

This technique is simple when there is already ingurgitation of the gonadal vein with reflux, so it is essential to perform venography to evaluate the size of this and the presence of valvular insufficiency.[7,8]

The laparoscopic approach allows complete dissection of the gonadal vein and avoids angulations after anastomosis with the iliac vein. It also allowed us to limit the size of the incision.

In selected patients, venous decongestion of the LRV into the inferior vena cava using the ovarian vein is reproducible and has a low incidence of complications. This technique avoids extensive abdominal surgeries and placement of intravascular materials. ■

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