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Treatment of nutcracker syndrome: a mini review

Abstract

Nutcracker syndrome is a rare condition caused by the compression of the left renal vein by the superior mesenteric artery and aorta. Diagnosis is challenging due to symptom variability, with hematuria being the most common symptom. Imaging techniques such as ultrasound, tomography, resonance imaging, or venography are required for confirmation. Treatment remains controversial, with options ranging from non-surgical management to open surgery and endovascular interventions, each with its own indications, advantages, and disadvantages. This article presents a mini-review of the current therapeutic options for treating this rare syndrome.

Keywords: nutcracker syndrome, left renal vein, superior mesenteric artery, aorta, hematuria, pelvic pain, endovascular, open surgery

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Introduction

"Nutcracker syndrome" (NS), first mentioned by Grant in the 1930's,[1,2] is characterized by the compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the abdominal aorta ("anterior nutcracker syndrome").[1,3,4] This results in impaired blood outflow and congestion.[5] Variations such as "posterior nutcracker syndrome," involving compression by the spinal column and the aorta, have also been described.[3,4] Due to the wide range of symptoms and the lack of well-established diagnostic criteria, the prevalence of NS is unknown.[1,5–7] Incidence rates are higher in the second decade of life to middle age, affecting a higher proportion of females.[4,6,7]

Symptoms include hematuria, pelvic and flank pain, orthostatic proteinuria, and renovascular hypertension.[1,4–8] Less common symptoms include congestion syndrome, dysmenorrhea, dyspareunia, left varicocele, hypercalciuria, syncope, hypotension, tachycardia, Henoch-Schönlein purpura, membranous nephropathy, nephrolithiasis, and Berger's disease.[1,2,4-6] Diagnosis is often made after excluding other pathologies due to the absence of well-established clinical criteria.[4,6] Routine blood tests are non-diagnostic, though anemia secondary to bleeding from hematuria may be observed. It can also be observed micro- or macrohematuria and proteinuria in the urinalysis.[2] Imaging techniques such as duplex ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), intravascular ultrasound (IVUS), and venography are required for confirmation.[1,2,6] Ultrasound is recommended as a first-line study with sensitivity and specificity of 69%-90% and 89%-100%, respectively. CT can identify LRV compression and other signs, with a "beak sign" and an aortomesenteric angle of <45° being indicative.[2,7,9,10] Despite the above, several researchers have reported as a cutting point an SMA branch angle 3 mm Hg, can be taken as a diagnostic criteria of NS 2.

Treatment

Treatment is controversial and ranges from conservative management to open and endovascular surgery, depending on symptom duration and severity.[2,4,6] Asymptomatic NS does not require treatment.[2]

Non-operative /conservative treatment

Conservative treatment is indicated for patients with mild symptoms and those under 18 years of age.[2,4,6] In young patients is recommended because the increase in intra-abdominal and fibrous tissue at the SMA origin during growth releases the obstruction of the LRV.[1,4] Weight gain can help by increasing retroperitoneal adipose tissue, repositioning the left kidney, and reducing tension on the LRV, resolving symptoms in around 30% of patients.[1,2] Low-dose acetylsalicylic acid (ASA) can improve renal perfusion.[1,4,12] Angiotensin-converting enzyme (ACE) inhibitors can help with orthostatic proteinuria.[1,4,7]

Intervention

Indicated for adults with severe symptoms such as hematuria and anemia or severe pain affecting quality of life.[2,4]

Open surgical treatment

Open surgery remains the standard treatment. Several techniques are available:

- Left renal vein transposition: Considered the standard care for persistent symptoms, it involves re-implanting the LRV to the inferior vena cava.[7] This surgery consists of sectioning the left renal vein and distal re-implantation to the inferior vena cava via a transabdominal, transperitoneal midline approach.[4] This intervention corrects the compression at the aortomesenteric angle by repositioning the confluence of the left renal vein to the inferior vena cava caudally by 3 - 5 cm 1. Vein tributaries (gonadal, adrenal and lumbar veins) are ligated and this allow mobilize the left renal vein with a subsecuente tension-free repositioning or transposition.[2] The completion assessment can include perform intraoperative ultrasound and assessment of intravenous pressure, which should be < 1 mmHg.[2] This procedure can present complications such as deep venous thrombosis, intestinal obstruction, ileus, retroperitoneal hematoma and LVR restenosis.[2,4,7] This intervention has several advantages such the short period of renal ischemia and few anastomoses, with good success rates (80-100% of symptoms resolution). It is considered, by experts, the gold standard treatment for NS.[1,4]

Vein patch angioplasty: Involves venoplasty at the LRV and inferior vena cava (IVC) confluence using autologous vein or prosthetic material.[2]

Vein cuff: Reduces tension on the LRV-IVC anastomosis.[2]

Left kidney autotransplantion: Nephrectomy and retransplantation to the iliac fossa, normalizing LRV pressure.[1,4,7] This surgery normalizes LRV pressure levels and offers excellent results with low morbidity.[4] The risks are a large duration of renal ischemia; stenosis

of anastomoses of the renal artery, renal vein and ureter; an important disadvantage can be the requirement considerable surgical exposure.[4,7]

Transposition of the superior mesenteric artery (SMA): Less common, involves reimplantation below the LRV.[4] This carries a significant risk of arterial thrombosis and of mesenteric ischemia.[4,7]

Nephropexy: Simple nephropexy with excision of varicosities for young patients.[4]

Nephrectomy: radical surgical procedure. Recommended for persistent hematuria.[4]

Gonadal vein transposition: some patients with NS diagnosis have an incompetent and enlarged gonadal vein, with associated pelvic congestion. Transposition of the left gonadal vein, via transverse mesocolon, on to the inferior vena cava allows decompression of the kidney, and help to eliminate the gonadal reflux.[2,4]

Renocaval bypass: Uses the great saphenous vein to construct a bypass, avoiding LRV transposition. In this intervention it is not necessary ligate the lumbar, gonadal or left renal veins, if they are not refluxing. There are limited experience with this intervention.[4]

Endovascular treatment

Minimally invasive procedures have gained relevance,[1] due to rapid recovery and symptom improvement.[5,7] The use of the stent into the left renal vein was described in 1996 for the first time.[13] The ideal characteristics of stent are: enough radial strength to eliminate stenosis, conformability to fit the epithelium of the vessel and little length shrinkage to enable adequate positioning. The 6 or 8 cm long self expanding stents (Wallstents) are the best option 2-4. The principal complications of this intervention are: stent migration, incorrect stent placement, partial displacement of the stent, or erosion, into the inferior vena cava.[1,2,4] To avoid migration it is recommended that the stent should be 20% larger than the venous diameter at the renal hilum 4. In these patients anticoagulant and antiplatelet drugs should be indicated for up to 3 months, it is the necessary time for endothelization of the stent.[4,6,7] The duration indicated is 3 days on low molecular weight heparin, 30 days on clopidogrel, and 3 months on acetylsalicylic acid.[4,5] Left gonadal vein or ovarian vein embolization can help to treat pelvic congestion and pelvic varicoceles with significant rate success of symptoms relief (56% - 98%).[2,4]

Post-intervention management

After open or endovascular approach to decompress the left renal vein, anticoagulation therapy with a direct-acting oral anticoagulant is indicated for <3 months.[2] Asymptomatic patients postoperatively can be assessed with ultrasound at 3 months, annually for 3 years, and subsequently every 3-5 years.[2]

Conclusion

Despite being a rare entity, the nutcracker syndrome has many therapeutic options that include non-operative treatment, open surgery and endovascular interventions. It is important to mention that as technology advances, endovascular management takes on great relevance, being a minimally invasive treatment with less morbidity and good results demonstrating similar effectiveness to open treatment as mentioned in Sarikaya et al trial.[14] Knowing all the management alternatives allows us to choose the most appropriate for our patients, depending on their clinical condition as well as their own characteristics. In conclusion, we must adequately approach our patients and assess them comprehensively to define what treatment will be ideal for each of them.

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Conflicts of interest

The author declared that there are no conflicts of interest.

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