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Treatment of Nutcracker Syndrome: A Mini Review

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Abstract

The nutcracker syndrome is a rare entity caused by compression of the left renal vein by the superior mesenteric artery and aorta. The diagnosis is a challenge because the variability of symptoms (hematuria is the most common) and required imaging such an ultrasound, tomography, resonance or venography to confirm. The treatment is still controversial but there are multiples options from non-surgical treatment to open surgery and endovascular interventions, each with its indications, advantages and disadvantages. In this article we present a mini review of the therapeutic options we currently have to treat this rare syndrome.

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

Introduction

The "Nutcracker syndrome" (NS) was mentioned by Grant in 1930's [1,2], it is a rare disease characterized by compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the abdominal aorta ("anterior nutcracker syndrome") [1,3,4] resulting in impaired blood outflow and congestion [5]. Certain variations have been described such a "posterior nutcracker syndrome", which consists in compression of the left renal vein by the spinal column and the abdominal aorta [3,4].

Due to the wide range of symptoms at presentation and the absence of well- established diagnostic criteria, the prevalence of NS is unknown [1,5-7]. Incidence rates are higher in second decade of life to middle-age (childhood to seventh decade) [6,7]. This syndrome apparently affects a higher proportion of female patients [4].

More cases are asymptomatic. There are a wide variety of symptoms such as hematuria (most common), pelvic and flank pain, proteinuria (orthostatic), renovascular hypertension [1-4,5-8]. Lest common symptoms are congestion syndrome, dysmenorrhea, dyspareunia, left varicocele, hypercalciuria, syncope, hypotension, tachycardia, Henoch-Schönle in pur pura, membranous nephropathy, nephrolithiasis and Berger's disease [1,2,4-6].

Diagnosis is complicated and is, usually, made after exclude other pathologies, because there are no well-established clinical diagnostic criteria [4,6]. Routine blood test results are nondiagnostic, although anemia secondary to bleeding due to hematuria may be observed. It can also be observed micro- or macrohematuria and proteinuria in the urinalysis [2]. Diagnosis tests such duplex ultrasound, computed tomography venography (CTV), computed tomography (CT), magnetic resonance (MRI), intravascular ultrasound (IVUS) and venography are required to confirm this syndrome [1,2,6]. Ultrasound is recommended as a first-line study, with sensitivity and specificity of 69%-90% and 89%-100% [1,2,11]. Computed tomography can identify the left renal vein compression by the aorta and SMA, it can be seen a "beak sign" that consist in triangular shape at the narrowing of the left renal vein at the aortomesenteric space, with sensitivity and specificity of 92% and 89%; other findings are the presence of venous collateral vessels and angle between the abdominal aorta and the SMA of <45° [2,7,9,10]. Despite the above, several researchers have reported as a cutting point an SMA branch angle <35° for the diagnosis of NS [1,7]. Magnetic Resonance Imaging (MRI) is preferred in young patients due to the absence of radiation [7]. Venography with measurement of the renal vein pressure gradient is the gold-standard diagnosis method [4,7]. The gradient of venous pressure from the LRV to the inferior vena cava (IVC) normal is <1 mmHg, but a gradient >3 mm Hg, can be taken as a diagnostic criterion of NS [2].

Treatment

Treatment is still controversial and includes conservative treatment, open or endovascular surgery procedures, depending on the duration and severity

of symptoms [2,4,6]. Treatment of asymptomatic NS is not indicated [2]. Nonoperative/Conservative treatment Indicated in patients with mild symptoms and young patients <18 years [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]. A treatment option that consists in weight gain increases the retroperitoneal adipose tissue, causing a repositioning of the left kidney with subsequent reduction of tension on the LRV; this therapeutic approach has been shown to resolve symptoms of NS in around 30% of patients [1,2]. Low dose acetylsalicylic acid (ASA) can be part of the treatment in patients with NS, because it helps to improve renal perfusion [1,4,12]. Angiotensin-converting enzyme (ACE) inhibitors can be used to help against orthostatic proteinuria [1,4,7].

Intervention

Indicated for adults with severe symptoms such hematuria, especially for those with anemia and requiring transfusion, and for patients with severe pain that decreases the quality of life and, sometimes, requires hospitalization [2,4].

Open Surgical Treatment

Open surgery has been, and continues to be, the standard of treatment. Several surgical techniques are available to manage nutcracker syndrome, I will list below the most common:

Left Renal Vein Transposition: Is considered the standard of care for patients with persistent symptoms and is the most common intervention for anterior and posterior NS, with excellent results in symptom control [7]. This surgery consists of sectioning the left renal vein and distal reimplantation 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 subsequent 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: Venoplasty of the LRV at the LRV e IVC confluence with segment of autologous vein, prosthetic material, or bovine pericardium, is another alternative. This can be performed in conjunction with LRV transposition to reduce the tension on the left renal vein e inferior vena cava anastomosis [2].

Vein Cuff: (Of the saphenous vein, typically) Can help with tension reduction on the LRV e IVC transposition anastomosis [2].

Left Kidney Auto Transplantation: It is an invasive intervention, and consists in nephrectomy and retransplantion of the kidney to the iliac fossa [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. Consists in transposition of the superior mesenteric artery from its origin at the aorta and reimplantation at a point 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. This intervention represents a low risk of injury to intraperitoneal organs, and can be considered such a management alternative for young patients [4].

Nephrectomy: Radical surgical procedure. Recommended in patients with persistent hematuria after different therapeutic approaches [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: This intervention uses the great saphenous vein to construct a bypass proximally to IVC and distally to LRV, and not require transposition of the LRV. In this intervention it is not necessary ligate the lumbar, gonadal or left renal

veins, if they are not refluxing. There is limited experience with this intervention [4].

Endovascular Treatment

In recent years these minimally invasive procedures have taken relevance [1]. Benefits of this interventions include rapid recovery time and symptoms 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].

Postintervention Management

After open or endovascular approach to de compress 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].

Conclusions

Despite being a rare entity, the nutcracker syndrome has many therapeutic options that include nonoperative 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.

Conflicts of Interest

The author declares that there is no conflict of interest.

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