CASE REPORT

NUTCRACKER SYNDROME: A RARE ETIOLOGY OF VARICOCELE

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ABSTRACT

Nutcracker syndrome (NS) describes left renal vein compression between the superior mesenteric artery and the aorta. NS is a rare entity. We report a case of nutcracker syndrome diagnosed with varicocele. A 15-year-old man with good past health presented with a left varicocele and intermittent left flank pain. He had no urinary symptoms and no haematuria. Doppler ultrasound and MRI showed a grossly distended left renal vein, and left varicocele secondary to the nutcracker syndrome. The patient underwent laparoscopic varicocelectomy and conservative treatment was adopted for the left renal vein compression. The evolution is favorable. NS is a rare entity. The treatment options are ranged from surveillance to nephrectomy. Treatment decision should be based on the severity of symptoms and their expected reversibility with regard to patient’s age and the stage of the syndrome.

INTRODUCTION

Nutcracker syndrome (NS) also known as left renal vein (LRV) entrapment syndrome, was first defined anatomically by Grant in 1937 as follows: “. the left renal vein, as it lies between the aorta and superior mesenteric artery, resembles an nut between the jaws of a nutcracker”. The nutcracker phenomenon was first described by de Schepper in 1972. Compression of the left renal vein between the superior mesenteric artery (SMA) and the aorta causes left renal vein hypertension.

The presence of symptoms and signs related to LRV outflow obstruction, including abdominal and left flank pain, macroscopic and microscopic hematuria, proteinuria and orthostatic proteinuria, and varicocele. The diagnosis of NS requires a high index of suspicion. It is extremely rare, and it represents a diagnosis of exclusion. Doppler ultra sonography is recommended as a first-line study. CT Scan and MRI can also be obtained, and they can be used to demonstrate compression of the LRV, gonadal vein distention, and pelvic congestion. Patients presenting with symptoms or signs and anatomic compression of the left renal vein (LRV) can be considered for intervention. Open, laparoscopic, and endovascular techniques have been developed to decrease the venous out flow of the LRV. We report a case of nutcracker syndrome diagnosed with varicocele.

Figure 1: A- Doppler ultrasound showed left varicocele.
DISCUSSION

The exact prevalence of NCS is unknown. Patients can present at any age from childhood to the seventh decade, with prevalence peaking in young (second or third decade) and middle-aged adults.6

Left renal ptosis, lordosis, and decreased retroperitoneal and mesenteric fat tissue may cause to NS.6,7 Right-sided NS is a more rare condition. Pregnancy is defining as a factor contributing to right-sided NS by compression of large veins.6 Left-sided IVC, hemiazygous continuation and persistent left superior vena cava combination is another rare cause of right NCS.6 All of the anatomic mechanisms involved in renal vein compression are resulting without flow obstruction leads to LRV hypertension with a measurable renocaval pressure gradient Clinical features of patients with NS are various. The symptoms vary from asymptomatic hematuria to severe pelvic congestion. Some patients have severe and persistent symptoms. Symptoms are aggravated by physical activity.9 Symptoms include hematuria, orthostatic proteinuria, flank pain, abdominal pain, varicocele, dyspareunia, dysmenorrhea, fatigue and orthostatic intolerance.10 The symptoms of autonomic dysfunction such as hypotension, syncope, and tachycardia could be seen but they are rare,11 while some patients suffer from severe, persistent symptoms, it is possible for others, especially children, to remain asymptomatic.12

Doppler ultrasonography (DUS) is recommended as a first-line study. It allows real-time assessment of the flow and peak velocities within the lumen of the LRV. A ratio of peak systolic velocity of the aortomesenteric segment to the hilar portion of <4.2 to 5.0 is considered one of the diagnostic criteria of NS.13 CT and MRI can also be obtained, and they can be used to demonstrate compression of the LRV, gonadal vein distention, and pelvic congestion. Furthermore, findings such as an LRV lumen diameter to aortomesenteric diameter ratio of >4.9, the “beak sign,” and an SMA branching angle of <35 degrees from the aortic origin can be also useful for the diagnosis of NS.13 Retrograde venography is the most informative method although it is an invasive test. It is not commonly performed in patients who have not severe symptoms.14

Spontaneous resolution by physical development during childhood is possible.15 Conservative approach with observation during minimum 2 years without medication is the best option for patients younger than 18 years old. As in the case of our patient, a laparoscopic varicocelectomy and conservative treatment was adopted for the left renal vein compression.

Surgical procedures are used for treatment in patients with severe symptoms. Nephrectomy, intraocular and extravascular stent implantation, transposition of the LRV or SMA, gonadal cava by pass, renal auto transplantation and nephrectomy are surgical procedures.

Open surgical techniques for anterior NS include LRV transposition, LRV transposition with patch venoplasty, patch venoplasty without LRV transposition, LRV transposition with saphenous vein cuff, gonadal vein transposition and saphenous vein bypass.16

findings suggestive of nutcracker are present without clinical symptoms. Term of NS is used for patients with clinical symptoms associated with nutcracker anatomy.

A 15-year-old man with good past health presented with a left varicocele and intermittent left flank pain. He had no urinary symptoms and no haematuria. The clinical examination showed a grade II varicocele. Doppler ultra sonography showed a grossly distended left renal vein, and left varicocele secondary to the nutcracker syndrome (Fig. 1). The cross sectional view of magnetic resonance imaging (MRI) showed compression of the left renal vein between the superior mesenteric artery and the aorta (Fig. 2). This compression caused the marked dilatation of the distal left renal vein with associated venous hypertension (nutcracker phenomenon). The patient subsequently, under went laparoscopic varicocelectomy and conservative treatment was adopted for the left renal vein compression, due to the intermittent nature of the clinical symptoms and its moderate intensity. The evolution is favorable.

Case Summary

A 15-year-old man with good past health presented with a left varicocele and intermittent left flank pain. He had no urinary symptoms and no haematuria. The clinical examination showed a grade II varicocele. Doppler ultra sonography showed a grossly distended left renal vein, and left varicocele secondary to the nutcracker syndrome (Fig. 1). The cross sectional view of magnetic resonance imaging (MRI) showed compression of the left renal vein between the superior mesenteric artery and the aorta (Fig. 2). This compression caused the marked dilatation of the distal left renal vein with associated venous hypertension (nutcracker phenomenon). The patient subsequently, under went laparoscopic varicocelectomy and conservative treatment was adopted for the left renal vein compression, due to the intermittent nature of the clinical symptoms and its moderate intensity. The evolution is favorable.

DISCUSSION

Nutcracker syndrome (NS), first described in 1937 by Grant,1 refers to patients presenting with symptoms and signs associated with the anatomic compression of the left renal vein (LRV). Most commonly known is the anterior NS, which refers to the compression of the LRV by the superior mesenteric artery (SMA) and the aorta.3,4 A second variant is the posterior NS, in which the LRV is compressed between the aorta and the vertebral body.3,5 The terms nutcracker phenomenon and nutcracker syndrome (NS) are sometimes used as synonym in the literature. Nutcracker phenomenon descript anatomic
Surgical placement of an externals tent to the LRV is another surgical approach to NS. Endovascular stenting is an alternative treatment option. It can be preferred to surgery because of the long period of renal congestion, additional anastomoses and extensive dissection requirement of the surgery.

CONCLUSION

NS is a rare entity. In a proportion of cases, more commonly in children, it can resolve spontaneously. For asymptomatic patients, the treatment is conservative. For patients symptomatic multiple techniques have been developed for the treatment of this condition.

Informed Consent

Written informed consent was obtained from patient who participated in this study.

Conflicts of Interest

The authors do not declare any conflict of interest.

Author Contributions

All the authors participated in the development and implementation of this work. They read and approved the final version of the manuscript.

References