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Case Report

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A Rare Renal Anomaly: Nutracker Syndrome

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Abstract

Nutcracker syndrome is a rare condition that manifests itself with abdominal pain and hematuria, usually resulting in compression of left renal vein between the aorta and the superior mesenteric artery. It is mostly seen in young women and the treatment should be individualized on a patient basis. A 17-year-old female patient was admitted to our clinic for 2 months of ongoing side pain, followed up with the diagnosis of Nutracker's syndrome after doppler ultrasonography and treated symptomatically.

Keywords: Flank pain, hematuria, Nutcracker syndrome

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INTRODUCTION

Nutcracker syndrome is a clinical condition that develops as a result of the compression of the left renal vein between the aorta and the superior mesenteric artery, in which the left renal vein expands due to impaired blood flow in it, and in which various symptoms and signs accompanying these morphological findings occur (1). This clinical condition was first reported by El-Sadr and Mina in 1950 (2), and the term Nutcracker was first used by de Schepper in 1972 (3). The terms Nutcracker phenomenon and Nutcracker syndrome can be used interchangeably in clinical practice. Nutcracker phenomenon is used for morphological findings due to compression of the left renal vein, whereas Nutcracker syndrome is used for cases where these symptoms are accompanied by clinical signs and symptoms (4). Clinical findings of Nutcracker syndrome vary from asymptomatic hematuria to severe pelvic congestion. General symptoms include hematuria, pain, varicocele, and orthostatic hypotension (5). Diagnosis is made by demonstrating the changes specific to Nutcracker syndrome in clinical findings and by imaging methods. Imaging methods used are Doppler ultrasonography (USG), computed tomography (CT), angiography, magnetic resonance (MR) angiography, and retrograde venography, but definitive radiological diagnostic criteria are not available (6). While conservative treatment is recommended in young patients with asymptomatic hematuria, various surgical, and intravascular interventional methods are applied in cases with severe renal congestion, side pain, and hematuria (1).

We present a rare case of a patient with "Nutcracker syndrome" who was admitted due to abdominal pain. Written informed consent was obtained from patients.

CASE PRESENTATION

A 17-year-old female patient was admitted to our nephrology outpatient clinic with complaints of left side pain that lasted for 2 months. On physical examination, her vital signs, abdominal examination, and urogenital examination were all normal. Laboratory tests revealed

white blood cell as 5.250 hc/mm^3, hemoglobin as 13.5 g/dL, platelet count as 194,000 hc/mm^3, and serum creatinine as 0.57 mg/dL (N=0.2–1.3). Two erythrocytes were seen in each area of magnification in complete urine analysis, and spot urine protein/creatinine value was found as 58 mg/g. Urinary infection was not considered with the existing findings. Since kidney size was normal in the abdominal USG and no stone or other pathology was detected, renal Doppler USG was performed to evaluate the renal vascular structures. On renal Doppler USG, the left renal vein was observed to be enlarged to the level where it crossed the aorta, and its diameter was measured as 13 mm. The diameter of the renal vein was significantly narrowed after crossing, and the diameter was measured as 3.5 mm. The clinical findings were confirmed by a different radiologist. Based on the current findings, the patient was diagnosed with Nutcracker syndrome. Since the patient had no serious symptoms and no hematuria, she was informed, and conservative treatment (spasmolytic, analgesic, and hydration) was applied by planning hematuria and clinical follow-up.

DISCUSSION

We present Nutcracker syndrome, which is a very rare condition in nephrology practice, in our case report.

There are different types of Nutcracker syndrome. The *anterior nutcracker* is the most common type that develops due to the compression of the left renal vein between the aorta and the superior mesenteric artery. The *posterior nutcracker* develops due to the compression of the retroaortic or circumaortic renal vein between the aorta and the vertebral corpus (7). In addition to classical types, there are many different causes, such as pancreatic tumors and para-aortic lymphadenopathy, leading to Nutcracker syndrome (1). Although this syndrome can be seen in all ages and genders, it is more common in the second and third decades of life and in women (5); our case study is also a young woman.

Clinical findings may range from asymptomatic hematuria to severe pelvic congestion. The most common symptom is hematuria, and patients usually have microscopic hematuria. Macroscopic hematuria is seen in approximately 20% of the cases (8). The second most common symptom is pain. Pain is usually side pain or lower abdominal pain and is associated with inflammatory processes developing secondary to venous hypertension (9). If side pain is of colic type, it should be considered that it may be due to blood clots developing due to hematuria in the left ureter (6). Varicocele develops due to hypertension of the left renal vein and affects 9.5% of men. Pelvic congestion develops in patients, and congestion-induced dyspareunia, dysuria, and dysmenorrhea can be seen (5). In addition, orthostatic proteinuria and orthostatic hypotension may cause serious morbidity in some patients. Previous studies show it to be more common in people with low body mass index (8). Our patient complained of side pain for 2 months, and no findings of other complications were observed.

It is important to examine the image of the left renal vein and gonadal vein anatomy while making the diagnosis of Nutcracker syndrome. The normal length of the left renal vein is 6-10 cm, and its normal diameter is 4-5 mm (10). However, the diameter of the left renal vein may vary in proximal and distal regions in healthy individuals, as well. Buscehi et al. showed that the ratio of the proximal-to-distal renal vein diameter may be as high as 4:1 in some healthy individuals (11). The normal pressure gradient between the left renal vein and the inferior vena cava is 0-1 mm Hg. The normal diameter of the left gonadal vein, which is found to be enlarged in Nutcracker syndrome, is 3 mm on average (12). The angle of the anterior mesenteric artery to the aorta is also important in the diagnosis because it is thought that the pressure on the left renal vein increases as this angle narrows. The angle of the aortic–mesenteric artery, which is almost 90° in healthy individuals, was found to be 38°-56° in these patients (6).

Doppler USG, CT angiography, standard MR, MR angiography, renal angiography, and retrograde left renal vein catheterization methods can be used to evaluate the anatomical structure and the inferior pressure difference of the left renal vein-vena cava. Renal Doppler USG is useful as the first imaging method because it is non-invasive. Although Doppler USG diagnostic criteria for Nutcracker syndrome have been tried to be established, it has not been successful yet. In various studies, the sensitivity of renal Doppler USG was found to be between 69% and 90%, and its specificity was between 89% and 100% (6). Multiphasic CT urography, which is one of the other imaging modalities, is valuable because it can show renal tumors and other causes of hematuria, such as arteriovenous malformations and urothelial tumors. Retrograde venography is the most detailed method for Nutcracker syndrome, but it is not used frequently because its results are misleading, and it is interventional (1). Doppler USG findings were found to be adequate for the diagnosis, and invasive procedures were not needed in our patient.

Different treatment methods can be used to treat Nutcracker syndrome, ranging from conservative treatment to intravascular intervention and open surgical procedures. Since hematuria is completely cured with conservative treatment in 75% of the patients <18 years, conservative treatment is recommended in this group of patients (6). Angiotensin inhibitors can also be used in patients with orthostatic proteinuria (13). The aim was to eliminate left renal vein hypertension in most of the interventional therapies. However, it is difficult to understand the extent to which imaging results are associated with left renal vein hypertension. Therefore, interventional therapies should be considered for patients with severe pain, severe hematuria, and renal dysfunction. Applicable surgical methods include left renal vein transposition, graft bypass of the left renal vein, renal autotransplantation, superior mesenteric artery transposition, gonadal bypass, and external stent placement to the left renal vein. In addition, a stent can be placed into the left renal vein as an intravascular intervention. Among these methods, external

stenting and intravascular stenting in the left renal vein have recently become prominent, and some studies suggested that these methods have serious advantages over the other methods (14). Since our patient was young and had no additional problems other than pain, only conservative treatment was applied, and the patient was informed that if symptoms could not be relieved or new complications would develop, interventional treatment would be needed.

CONCLUSION

Nutcracker syndrome is diagnosed through imaging methods upon clinical suspicion. The treatment of the patient should be planned and applied according to the patient's symptom status, anatomical structure, and degree of organ dysfunction.

Informed Consent: Written informed consent was obtained from patient.

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