

Chronic Pelvic Pain and Hematuria Associated with Posterior Nutcracker Syndrome

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ABSTRACT

The nutcracker syndrome is a rare syndrome in which the left renal vein is compressed between the aorta and the superior mesenteric artery or even more rarely between the aorta and vertebral body, in case of an anatomical variation of the left renal vein.

It is more likely discovered within young patients presenting pelvic pain, hematuria, or varicocele. Treatments vary from conservative, to surgical or endovascular.

Here we present the case of a young patient with chronic pelvic pain and hematuria, diagnosed as a posterior nutcracker syndrome.

KEYWORDS: pelvic pain-hematuria-left renal vein-posterior nutcracker syndrome.

Introduction

The nutcracker syndrome (NCS) is a rare syndrome defined by clinical symptoms occurring due to an anatomical compression of the left renal vein between the aorta and superior mesenteric artery in anterior NCS, or between the aorta and vertebral body in posterior NCS.

It can be found in young patients presenting hematuria.

We describe the case of a young patient consulting for chronic pelvic pain, and one episode of hematuria leading to the diagnosis of a posterior NCS.

Case Description

A 30 years old female patient, treated for breast cancer, consulted for chronic pelvic pain, with one episode of hematuria. The patient had lost lots of weight with her illness and had noticed that pain was most intense during effort, which led her to see her doctor. Her clinical and biological exams were normal. A thoraco-abdominal CT scan was performed, and revealed a retro-aortic left renal vein, compressed between the aorta and vertebral body, with a dilation of the left gonadal vein and pelvis varices, confirming the diagnosis of the posterior nutcracker syndrome.

In follow up, the patient only had occasional pelvic pain for which only a conservative treatment was chosen.



Figure 1: Computed tomography angiogram of abdomen showing a retro-aortic renal vein in axial view (A) and sagittal view (white arrow) (B) with a compression of LRV between the aorta and vertebral body (A)



Figure 2: Abdominal CT angiogram, coronal view in maximum intensity projection, showing dilation of left gonadal vein (white arrow) and pelvic varices.



Figure 3: Abdominal CT angiogram, in coronal volume rendering, showing retro-aortic left renal vein with dilation of left gonadal vein and pelvis varices.

Discussion

The nutcracker syndrome was first described by Grant in 1937 and first written in 1950 by El Sadr in patients presenting an anatomical compression of the left renal vein (LRV), most commonly the anterior one where the left renal vein is compressed between the aorta and the superior mesenteric artery, due to a narrow angle between them causing hypertension of the left renal vein. [1], [2]

The posterior nutcracker syndrome is rarer, with a little more than ten cases described in literature. [2] It is defined by the anatomical compression of the left renal vein between the aorta and the vertebral column caused by an anatomical variation of the left renal vein, seen in about 3% of the population [3] : either a retro-aortic LRV or circum-aortic one

(2 LRV passing anteriorly and posteriorly to the aorta). [3] [4]

The anatomical compression of LRV without clinical symptoms, is called « the nutcracker phenomenon ».

The detection of this syndrome is difficult through clinical and biochemical features alone. Thus, imaging is required and is based on Computed Tomography (CT), Doppler ultrasound and renal venography. Sometimes magnetic resonance angiography can be used instead of CT [5].

The most common clinical manifestations are: hematuria, postural proteinuria, pelvic pain or varicocele. They are related to venal hypertension. [4]

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The pressure in the left renal vein, will break glomerular capillaries and make red corpuscles and urinary protein pass through the thin walled septum into the collecting system, causing hematuria.

This pressure will eventually lead to vein reflux, dilation of left gonadal vein and pelvic congestion, with possibility of formation of pelvic varices or varicocele. [3], [4]

The best way to confirm the diagnosis is through imaging.

Doppler ultrasound can show the anatomical variation, pelvic varices with a dilation of renal and gonadal vein, but intestinal gas and the radiologist's experience may affect the results.

Venography measures the pressure gradient between the renal vein and inferior vena cava which is normally <1mmHg. If the gradient is >3mmHg it is considered as a gold standard to the diagnosis. [5] This method is very invasive, therefore CT imaging is the most reliable method to confirm the diagnosis, because it is simple, rapid and stable.

It allows an analysis of vessels anatomy and their variations, measurement of their diameter and allows a safety guideline for surgical treatment. [4]

It should be done in arterial and venous phases.

In anterior nutcracker syndrome, it will show an abrupt narrowing of the left renal vein between the aorta and vertebral body, with an acute aortomesenteric angle, called as the « beak sign ». [3]

However, in posterior nutcracker syndrome the angle between the aorta and vertebral body is relatively fixed. The normal distance between aortic wall and vertebral body is between 1-28mm and can decrease to 2-8mm in posterior NCS. [3]

The most effective evaluation method is the diameter ratio between the renal hilum and the stenosis of left renal vein, with a ratio>4 in this syndrome. [4]

CT can also show gonadal vein dilation and pelvic varices.

Treatment of this syndrome depends on age and severity of symptoms, in patients under the age of 18 with mild

symptoms, such as microscopic hematuria, a conservative treatment is preferred.

If the conservative treatment doesn't work or the symptoms are severe, a surgical treatment is required and the most common and effective method is the transposition of the left renal vein.

Endovascular stenting can also be used and is less invasive than surgery, especially because of its shorter time, quick recovery and less complications compared to surgery. [4], [2]

Conclusion

The nutcracker syndrome is rare and should be considered in young patients with hematuria, pelvic varices or varicocele. CT imaging is the most reliable modality to confirm diagnosis and endovascular stenting is a new alternative treatment of this syndrome to avoid the risks of surgical procedures.

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