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NUTRACKER SYNDROME

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Abstract

In the nutcracker anatomy, the aortomesenteric angle is more acute, which leads to a compression of the left renal vein between the aorta and the superior mesenteric artery. As a result, there is an impended outflow of the left renal vein in the inferior vena cava. This is termed as nutcracker syndrome. Complex of clinical symptoms exists in the nutcracker syndrome (NS). Nutcracker phenomenon (NP) is the term used to describe an anatomic finding without any clinical symptoms. The aim of this case study was to show the use of a multiphase computed tomography as a diagnostic method of choice in dealing with cases of nutcracker anatomy in patients.

We present four cases of patients who underwent computed tomography of the abdomen. Two of the cases were with anterior nutcracker syndrome, one case was with posterior nutcracker syndrome and one case with nutcracker phenomenon.

Establishing of the exact diagnosis in these patients is difficult, because of the rarity and variety in symptoms. Existence of the clinical symptoms distinguishes whether the patient has NS or NP. Multiphase CT scan of the abdomen with additional reconstructions in maximum intensity projection (MIP), volume rendering (VR) and multiplanar reconstruction (MPR) as an imaging method is the method of choice in the diagnosis of NS and NP.

Keywords: nutcracker syndrome, nutcracker phenomenon, computed tomography, anatomic variations

Introduction

The normal position of the left renal vein is anterior to the abdominal aorta, in the space between the aorta and the superior mesenteric artery which has a normal angle of about 90° [1].

When this angle is more acute (less than 90°) the result is compression of the left renal vein between the aorta and the superior mesenteric artery [1]. As a result, there is an impended outflow of the left renal vein in the inferior vena cava [2].

This is termed as nutcracker syndrome. Also, if there is a retro-aortic course of the left renal vein (LRV), it will lead to compression of the LRV between the aorta and the vertebral column. This is termed as posterior nutcracker syndrome [1,2]. When there is anterior or posterior NS, there is a complex of symptoms with different level of clinical manifestation [2,3].

Nutcracker phenomenon (NP) is the term that is used to describe an anatomic feature like this without any clinical symptoms [2,3].

The prevalence of NS and NP is unknown [3,4]. The age group of patients can range from the first to the seventh decade, however the highest peak of occurrence is in the second and third decade or in the middle-aged adults [3,4].

As to the sex of the patients, females have been reported to be affected more often [3,4]. The aim of this case study was to show the use of a multiphase computed tomography (CT) as a diagnostic method of choice in dealing with cases of nutcracker anatomy in patients.

Case series

We present 4 cases of patients where a CT examination of the abdomen in non-contrast and post-contrast phases was performed. The examinations were made using a CT scanner ”Somatom go up”, with 32 rows of detectors. The recording is performed at a tube voltage of 120 KV. The rotation speed is 0.8 sec.

The pitch factor is 0.09 -1.5. Also, the recording is performed during automatic submission of mA (Auto mA) and usually it ranges from 13 mA - 240 mA. The cross-sectional thickness of the native series is 3 mm, and of the post-contrast series 1 mm. Additional reconstructions are made in maximum intensity projection (MIP), volume rendering (VR) and multiplanar reconstruction (MPR).

The Nois index is 13. The maximum wide field of view (FOV) is used. The reconstruction matrix is 512 x 513 pixels.

The first patient was a 30-year-old woman who was sent for a CT scan of the abdomen.

The working diagnosis was ectopic pregnancy. The clinical symptoms were pain in the left inguinal region and vaginal bleeding. There was anamnestic data for previous ectopic pregnancy and left unilateral salpingectomy. The laboratory analysis showed macroscopic hematuria and proteinuria. A CT scan was indicated and performed; it showed an enlarged cavity of the uterus with a thickened miometrium and a hyper-attenuated zone towards the cervical canal.

Ovarian cystic formations were present bilaterally. There was compression of the left renal vein between the abdominal aorta and the superior mesenteric artery with prominent left ovarian vein and marked as periuterine vascularization on the left. It was an incidental finding that went in favor of anterior NS.

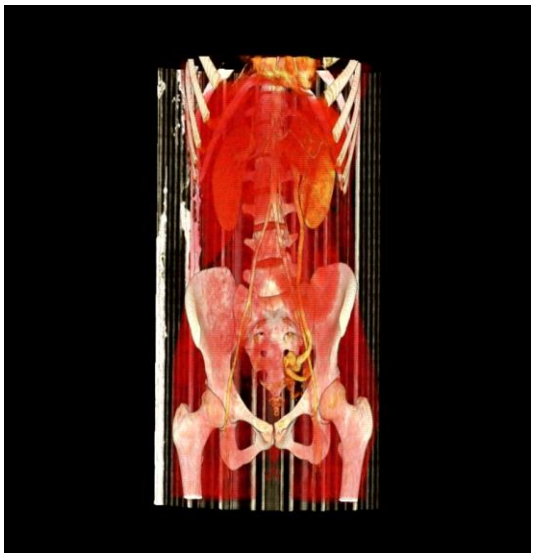

Fig. 1 Compressed LRV between AA and SMA, sagittal view, Case 1

Fig. 2 Left ovarian varices, coronal view, VR, Case 1

Fig. 3 Left ovarian varices, coronal view, Case 1

The second patient was a 43-year-old woman with repetitive epigastric pain. There was indication for further examinations, which included gastroscopy, abdominal ultrasound and laboratory analysis. All were with normal findings.

However, the epigastric pain persisted and was radiating towards the left side of the abdomen. A CT scan of the abdomen was performed where compression of the left renal vein between the abdominal aorta and the superior mesenteric artery was visualized, with prominent left ovarian vein, and pelvic venous congestion. Also, a small amount of fluid was present in the pelvis. Thus, an incidental finding of anterior NS was made.

Fig. 4 Reduced aortomesenteric angle with narrowing of the LRV, axial view, Case 2

The third patient was a 34-year-old woman who was sent for a CT scan of the abdomen due to a previous ultrasound finding of mild hydronephrosis in the left kidney.

The patient was without any clinical symptoms.

The CT scan showed parapelvic cyst with dimensions 47 x 23 mm in the left kidney which caused impression on the pyelon. The left ureter was slightly hypotonic.

There was a visible compression of the left renal vein between the abdominal aorta and the superior mesenteric artery and marked left periuterine vascularization. The secondary finding supported the presence of NP.

Fig. 5 Multiple pelvic left ovarian varices, sagittal view, Case 3

The fourth patient was a 50-year-old woman with a history of abdominal pain, meteorism and constipation in the previous months. She was sent for a CT scan of the abdomen for further investigation of the cause.

The CT scan showed a retro-aortic position of the left renal vein between the abdominal aorta and the vertebral column. There was marked left periuterine vascularization. An incidental finding of posterior NS was made.

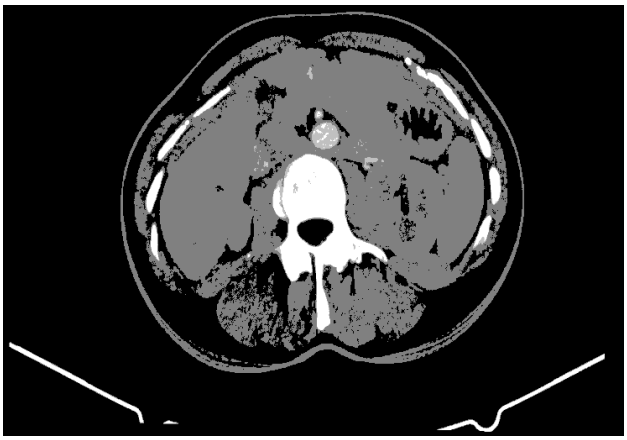

Fig. 6 Retro-aortic position of the LRV between the AA and the vertebral column, axial view, Case 4

Discussion

NS and NP are rare entities [5,6]. In majority of cases there is association with asthenic constitution of the patient and subsequently reduced retroperitoneal and mesenteric fat, also an abnormal route of the left renal vein and the superior mesenteric artery [7].

In NP, there are no clinical symptoms, but the anatomic variation exists [2,3].

On the contrary, in NS there is variety of clinical symptoms which include atypical left flank pain, hematuria, orthostatic proteinuria, varicocele and infertility, dyspareunia and other gynecological symptoms, varicose veins in the pelvis, buttocks or upper thighs, orthostatic hypotension, fatigue and abdominal pain [8].

The diagnosis of NS is difficult to be established clinically and because of that it is usually delayed [9]. Imaging methods as a multiphase CT of the abdomen are necessary for a definite diagnosis [6, 10].

Multiphase CT of the abdomen in three planes (axial, coronary and sagittal) can demonstrate the compression of the left renal vein in the fork formed between the superior mesenteric artery and the abdominal aorta, or between the vertebral column and the abdominal aorta, as well as left ovarian vein distension and pelvic congestion [7].

In our case series, three of the patients, the first, the second and the fourth case were with the diagnosis of NS. They presented with clinical symptoms like left inguinal pain, abdominal pain and hematuria. Clinically there was difficulty in establishing the diagnosis and therefore several other diagnostic methods were used.

Finally, the anatomical variant was detected when an examination with a multiphase CT scan of the abdomen in three planes was performed. Also, additional reconstructions of MIP, MPR and VR were used for better visualization of the vascular vessels. In all cases, there was also a left ovarian vein distension and pelvic venous congestion. On the other hand, in the third case NP was established, because the patient was asymptomatic. In all four cases, the multiphase CT scan of the abdomen was performed for another reason not linked to the anatomic variant. Consequently, the NS and the NP were detected as a secondary incidental finding.

These conditions are associated with a vascular disorder; however, the majority of patients has manifested urological or gynecological symptoms [1,8].

This was also the case with our patients. Therefore, clinicians should keep this condition in mind in patients presenting with recurrent flank pain or hematuria [1,8]. Empirically we know that the clinical course of this condition is characterized by repeated diagnostic procedures which may be avoided by early diagnosis [9].

An imaging method as a computed tomography scan of the abdomen is required to diagnose this condition [7,9]. The "Beak sign", the term used for the abrupt narrowing of the left renal vein with a triangular shape at the aortomesenteric portion in the nutcracker anatomy is the most useful finding among the various CT parameters, because it has shown sensitivity of 91.7% and specificity of 88.9% [2].

Also, there is prestenotic dilatation of the left renal vein together with perirenal and ovarian vein varices [2].

The treatment options are various and still under discussion [2,7,9]. Conservative treatment and follow-ups are recommended if the symptoms are mild [2,7,9].

Surgical or radiological interventions are indicated for severe symptoms like consistent and strong pain, gross hematuria and impairment of the renal function [2,7,9].

One of the most frequently used procedures is the transposition of the left renal vein [6,9].

Endovascular intervention is now considered the first line of therapy for the management of NS and it has been reported as a relatively effective technique [4,9].

Conclusion

The nutcracker anatomy can be presented as an anterior or posterior NS or NP. Establishing of the exact diagnosis is difficult, because of the rarity and variety in severity and symptoms. Existence of the clinical symptoms distinguishes whether the patient has NS or NP.

Multiphase CT scan of the abdomen with additional reconstructions like MIP, MPR and VR as an imaging method is the method of choice in the diagnosis of NS and NP.

This method provides a clear display of the anatomic variations of the blood vessels in question in patients with nutcracker anatomy. This provides the clinicians with the final diagnosis.

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