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Diagnostic Imaging

Left inferior vena cava with nutcracker syndrome: A case report

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ABSTRACT

Nutcracker syndrome is a condition where there is compression of the left renal vein between the aorta and the superior mesenteric artery causing symptoms. Here, we describe the case of a female patient who had symptoms secondary to the compression of a left inferior vena cava between the aorta and the superior mesenteric artery causing a nutcracker syndrome. We will review the etiology, embryology, and imaging of the inferior vena cava and nutcracker syndrome.

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Introduction

Compression of the left renal vein between the aorta and the superior mesenteric artery is known as anterior nutcracker phenomenon [1–3]. If the compression occurs between the aorta and the vertebral body it is called posterior nutcracker phenomenon [1–3]. If the vascular compression becomes symptomatic the term syndrome is preferred over phenomenon [1].

The later entity was first described in 1950 by doctors El-Sadr and Mina, and nutcracker syndrome was first described by De Schepper but it was first used by Doctor Chait in 1971 [2].

In this report we present the case of a patient with the diagnosis of a nutcracker syndrome secondary to the compression of a left inferior vena cava (variant) between the superior mesenteric artery and the aorta.

Case presentation

This is the case of a 33-year-old female with long-term history of recurrent diffuse abdominal pain associated to sporadic episodes of diarrhea. The patient states that sometimes the pain occurs after the intake of food; however, she denies postprandial nausea or vomit. In the system reviews the patient refers

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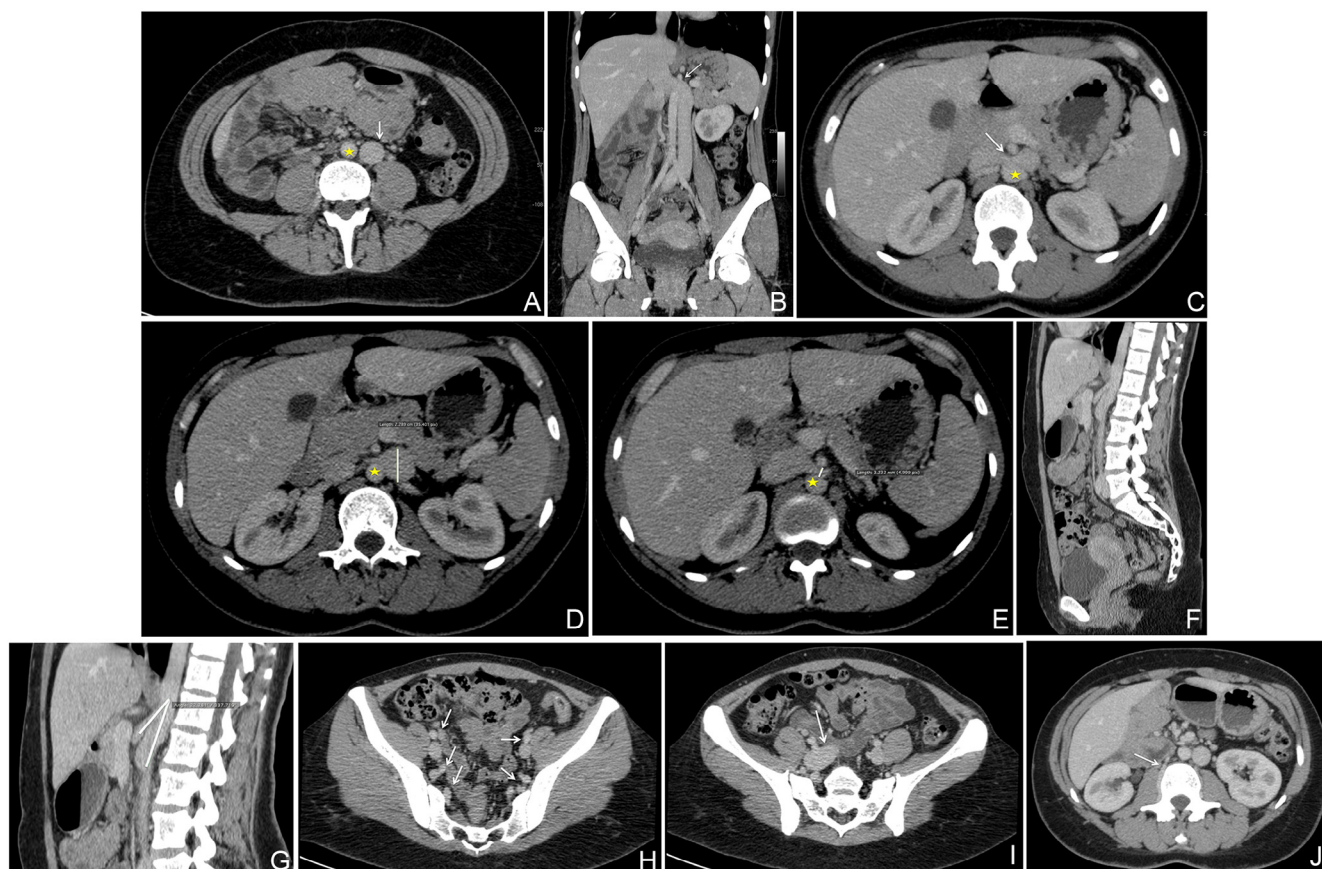


Fig. 1 – (A) Contrast abdominal computed tomography (CT) in the axial plane where the 2 main retroperitoneal vessels are seen; the aorta (yellow star) is located in the middle line and the inferior vena cava (white arrow) in the left. **(B)** Coronal abdominal CT where the inferior vena cava is on the left of the patient and it crosses to the right side just anterior to the aorta. **(C)** Axial abdominal CT. Note how the inferior vena cava (white arrow) is compressed by the aorta (yellow star) and the superior mesenteric artery while it crosses the midline. **(D and E)** Axial abdominal CT showing the caliber of the inferior vena cava before it crosses the midline, measuring up to 22 mm. Figure 1E shows the diameter of the inferior vena cava at the site of the compression with a diameter of 3 mm, creating the peak sign, thus a difference of more than 50% is estimated. **(F and G)** Sagittal reconstruction and zoom of the sagittal reconstruction showing the aortomesenteric angle of 22°. **(H)** Axial contrast-enhanced computed tomography (CECT) where there are multiple large vascular (white arrows) structures in the pelvis secondary to the compression of the left inferior vena cava. **(I)** Axial CECT where a dilated right iliac vein is observed. **(J)** Axial CECT showing an engorged right lumbar vein (white arrow).

dyspareunia and dysmenorrhea since many years ago. At physical examination mild abdominal tenderness is found. Given the bizarre case, the primary care physician orders abdominal computed tomography (CT) with contrast.

On the axial images the presence of a left inferior vena cava is noted (Fig. 1A and B). A transition point is noted below the superior mesentery artery, in the course where the abnormally located inferior vena cava crosses over to the right side to continue along a normal course to the right atrium (Fig. 1C) with a peak sign (Fig. 1D and E). In the sagittal reconstruction the aortomesenteric angle is 22° (Fig. 1F and G). There is prominence of the pelvic vasculature and bilateral enlargement of the iliac veins (Fig. 1H and I). The presence of engorged lumbar veins that drained into the inferior vena cava was also noted (Fig. 1J). The presence of thrombus, tumor, and masses was excluded.

Etiology

The prevalence of the nutcracker syndrome is not known at the time [1], because many patients are asymptomatic; therefore, in some cases it is never diagnosed. It is known that this pathology is more common in female patients between the third and fourth decades [1].

As to the etiology, it is known that there is a decrease in the angle between the superior mesenteric artery and the aorta (normal value: 38°–65°) [1], as well as a decrease in the distance between the aorta and superior mesenteric artery (aortomesenteric, normal value 10–14 mm) [1]. Among the causes that have been described to alter the aorto-mesenteric distance and the angle between the aorta and superior mesenteric artery is the loss of weight with decrease of retroperitoneal fat.

The most appealing fact about our case is that the patient had a left inferior vena cava (one of its multiple anatomic variants), which is compressed between the aorta and the superior mesenteric artery at the level of the drainage of the left renal vein.

The left inferior vena cava has been well documented previously in the literature and occurs in approximately 0.2%–0.5% of the general population. In most cases, the vena cava is left-sided up to the level of the left renal vein. At this point it crosses the midline in front of the aorta to continue with the inferior (intrahepatic) right vena cava.

The presence of a nutcracker syndrome caused by the compression of a left inferior vena cava by the superior mesenteric artery and the aorta is extremely rare and has only been described in the literature in 3 occasions, so its prevalence is unknown.

Embryology

The development of the inferior vena cava is a very complex process. It forms from 3 pairs of embryologic veins: posterior cardinal, subcardinal, and supracardinal. These veins form some anastomosis and then most of these veins regress, except the vein that forms the inferior vena cava. The mature inferior vena cava vein is a complex structure that originated from some parts of these veins and its anastomosis.

The anomalies of the inferior vena cava occur when there's an abnormal regression of the embryologic veins. The left inferior vena cava happens when the right supracardinal vein regresses and there is persistence of the left supracardinal vein [4].

Clinical manifestations

The patients can present with micro- or macrohematuria, orthostatic proteinuria, abdominal pain, varicocele or pelvic varices, and pelvic venous congestion syndrome (dyspareunia, dysmenorrhea, chronic pelvic pain) [1,2], all of which are secondary to venous hypertension [3]. The most characteristic feature is that these symptoms appear after exercise [1]. Some patients have diffuse abdominal pain that is similar to biliary pain [4] and lumbar pain [5].

Imaging findings

The diagnosis of the pathology can be made with abdominal CT, abdominal angioresonance, Doppler ultrasound, and venography [3].

On the CT, an abrupt compression of the left inferior vena cava or of the left renal vein between the superior mesenteric artery and aorta with dilatation of the proximal vein can be seen. This sign is called the “peak sign” [1], and it has clinical meaning when the difference of diameter between the vein before and after the compression is more than 50% [2]. Also, the aortomesenteric distance in the sagittal plane has been proposed for the diagnosis when it is less than 3 mm [1]. On the sagittal plane, the aorta-mesenteric angle has to be less than 16° [1].

The diagnosis with Doppler ultrasound is done when the ratio of systolic peak velocity between the site of the compression and the vein at the renal hilum is >4.7 [1]. This finding has a sensitivity of 100% and a specificity of 90% [1,3]. Although this finding was described in the original nutcracker, we assume that it may be extrapolated.

The gold standard for the diagnosis is the venography, which is diagnostic when the pressure difference between the left renal vein and inferior vena cava is more than 3 mm Hg [1].

Conclusion

The nutcracker syndrome secondary to the compression of the left inferior vena cava between the superior mesenteric artery and aorta is extremely rare and to our knowledge has only been described 3 times in the medical literature. The radiologist has to be familiar with the inferior vena cava variants and its complications to perform an early diagnosis and prevent long-term sequels.

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