Case Report

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Unusual coexistence of double inferior vena cava with nutcracker syndrome–a case report and review of the literature

Anna Waśniewska[1], Kacper Ruzik[1], Łukasz Olewnik[1], Ludomir Stefańczyk[2] and Michał Polguj[1]

Abstract

Knowledge of vessel anomalies is significant for all specialists in clinical practice and may prevent serious complications following medical interventions. Here, the rare coexistence of a duplicated inferior vena cava (IVC) and nutcracker syndrome in a 42-year-old female patient with atypical abdominal pain is presented, using two complementary radiological techniques (colour Doppler ultrasonography and computed tomography angiography). The right renal vein was found to be compressed when passing between the superior mesenteric artery and the abdominal aorta. The lumen dimensions (width × height) of the right IVC and left IVC at the level of termination were 15.8 × 17.7 mm and 13.4 × 12.4 mm, respectively. Ultrasonography revealed low blood flow in the left IVC that was reversed, and thus blood travelled in the same caudal direction as in the aorta. In the right IVC, however, flow travelled in the cranial direction. The simultaneous existence of a duplicated IVC and nutcracker syndrome is an extremely rare vessel anomaly; nevertheless, this dual presence may result in clinical symptoms and would have an impact on medical operations and even minor medical procedures.

Keywords

Duplication, inferior vena cava, nutcracker syndrome, left renal vein entrapment, colour doppler ultrasonography, computed tomography angiography

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[1]Department of Normal and Clinical Anatomy, Chair of Anatomy and Histology, Medical University of Łódź, Łódź, Poland
[2]Department of Radiology and Diagnostic Imaging, Medical University of Łódź, Łódź, Poland

Corresponding author:
Anna Waśniewska, Department of Normal and Clinical Anatomy, Chair of Anatomy and Histology, Medical University of Lodz, Żeligowskiego 7/9, Łódź 90-752, Poland.
Email: ania.wasniewska@gmail.com

Introduction

The first case of duplication of the inferior vena cava (IVC), a congenital condition, was reported in 1916 by Lucas.[1] IVC duplication has a possible incidence rate of between 0.2% and 4.4%, and the anomaly is characterised by the presence of two veins situated bilaterally to the aorta: the left IVC and the right IVC, which are separate veins most often connected at the level of the renal veins.[2,3] The condition is usually asymptomatic and is diagnosed by chance during radiographic examination due to other causes.[4]

Left venous renal entrapment is commonly called the nutcracker phenomenon, and results in a decreased outflow of blood from the left renal vein (LRV) to the IVC.[5] The condition is usually caused by compression of the LRV between the superior mesenteric artery and the abdominal aorta, and entrapment results in dilatation of the lateral part of the LRV and a narrowing of the medial part.[6] The consequence of these changes is collateralization of the renal pelvis and hypertension in the vessels, which mainly promotes haematuria. Other common symptoms are albuminuria, left flank pain, lumbar pain and varicocele.[5,7] The nutcracker phenomenon along with the contemporaneous presence of symptoms is commonly called nutcracker syndrome, which can be categorized into a few subtypes depending on the course of the LRV. The most common nutcracker syndrome variants are anterior (involving compression of the LRV by the abdominal aorta and the superior mesenteric artery) or posterior (involving compression of the LRV between the abdominal aorta and the vertebral column). Vessel course has a significant influence on venous interventional radiologic procedures and retroperitoneal surgeries. Unrecognised anomalies prior to medical procedures may lead to interruption of veins and severe haemorrhage.[3,4,6,8,9] To the best of the authors' knowledge, the present case is a unique description of duplicated IVC coexisting with nutcracker syndrome, presented using two complementary radiological techniques.

Case report

A 42-year-old Caucasian female presented at Norbert Barlicki University Teaching Hospital No. 1, Lodz, Poland, in May 2015, reporting atypical abdominal pain, which was exacerbated by exertion and menstruation. She did not suffer any other symptoms, and laboratory parameters and urinalysis were normal. Initial abdominal ultrasonography indicated a vascular anomaly as there was an additional vein corresponding to the size of the IVC, but located on the left side of the body.

The second step of the diagnostic procedure involved colour Doppler ultrasonography, performed using the GE Vivid 7 PRO ultrasound unit (GE Healthcare, Chicago, IL, USA) and computed tomography (CT) angiography, performed using the GE LightSpeed 64 VCT scanner (GE Healthcare). Colour Doppler ultrasonography confirmed the presence of two IVCs (Figure 1), and investigation revealed differences in the direction of blood flow between the left and right IVC. An ultrasound scan at the level of the umbilical region showed that blood flow in the left IVC was weak (Figure 1a), whereas blood flow in the right IVC was normal (Figure 1b). Additionally, the low flow in the left IVC (Figure 1c) was reversed, occurring in the same caudal direction as in the aorta, while flow in the right IVC was observed to occur in the cranial direction (Figure 1b). In the left IVC, the direction of blood flow depended on the respiratory phase. During inhalation, the crura of diaphragm decreased the angle between the superior mesenteric artery and abdominal aorta. Moreover, the decrease of pressure in the chest exaggerated blood flow

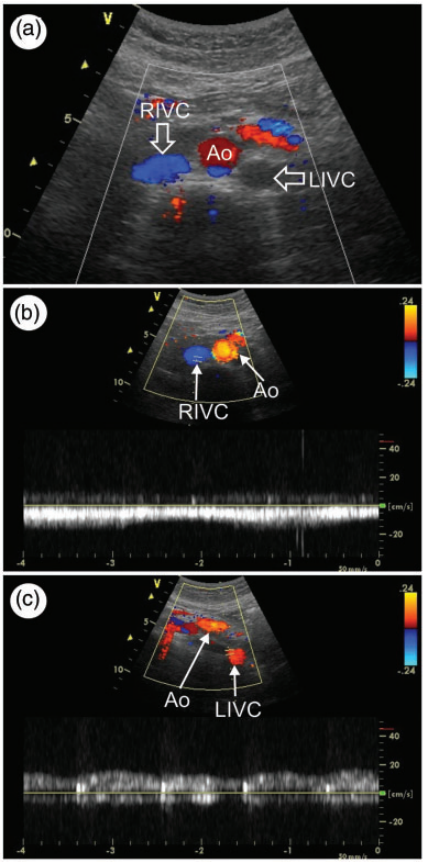

Figure 1. Colour Doppler ultrasonography images of the abdomen, scanned at the L2 level, showing: (a) the abdominal aorta (Ao), the right inferior vena cava (RIVC) typically located on the right side, with normal blood flow (indicated by blue colour, cranial direction), and the left inferior vena cava (LIVC) on the left side, with weaker blood flow (indicated by no visible colour change); (b) the spectrum of blood flow velocity of the typical RIVC (blue colour, cranial direction), and opposite blood flow direction of the Ao (red colour, caudal direction); and (c) the spectrum of blood flow velocity in the atypical LIVC (red colour, caudal direction, similar to the Ao).

in the right IVC and reduced blood flow in the left IVC. CT angiography (set to a 0.625-mm layer width and a 0.6-mm pitch) further confirmed the presence of a duplicated IVC (Figure 2). Additionally, the LRV was shown to be compressed when passing between the superior mesenteric artery and the abdominal aorta (Figure 3).

According to CT angiography measurements, the kidneys were of normal size; the length of the right kidney was 107.0 mm and the left was 108.0 mm. The lumen of the IVC at the level of the hepatic veins was 20.0×23.0 mm (always width $\times$ height, respectively), the right IVC at the level of termination was 15.8×17.7 mm, and the left IVC at the level of termination was 13.4×12.4 mm (Figure 4). The right renal vein (RRV) was measured to be 12.6×9.8 mm and the LRV varied from 8.8×12.8 mm (before narrowing) to 10.0×3.8 mm (at the narrowest place). In addition, the angle between the superior mesenteric artery and the abdominal aorta was 32°. The lumen diameter of the left ovarian vein before opening into the LRV was 5.0 mm. Comparatively, the lumen diameter of the right ovarian vein before opening into the RRV was 4.0 mm (Figure 4).

In the present case, there was no significant narrowing of the LRV at the level of crossing the abdominal aorta, nevertheless, difficulties in blood outflow due to the high volume of blood from the lower limbs and left kidney led to reversed blood flow in the left IVC.

Ethics approval was not deemed necessary for this retrospective study. Images were provided by the Department of Radiology and Diagnostic Imaging, Medical University of Lodz, following permission from the Head of the Norbert Barlicki Memorial Teaching Hospital, Lodz, and verbal informed consent to publish the case was provided by the patient.

Discussion

A search of the scientific literature during the present study revealed 14 different variations of the IVC, with IVC duplication being the most frequently reported anomaly

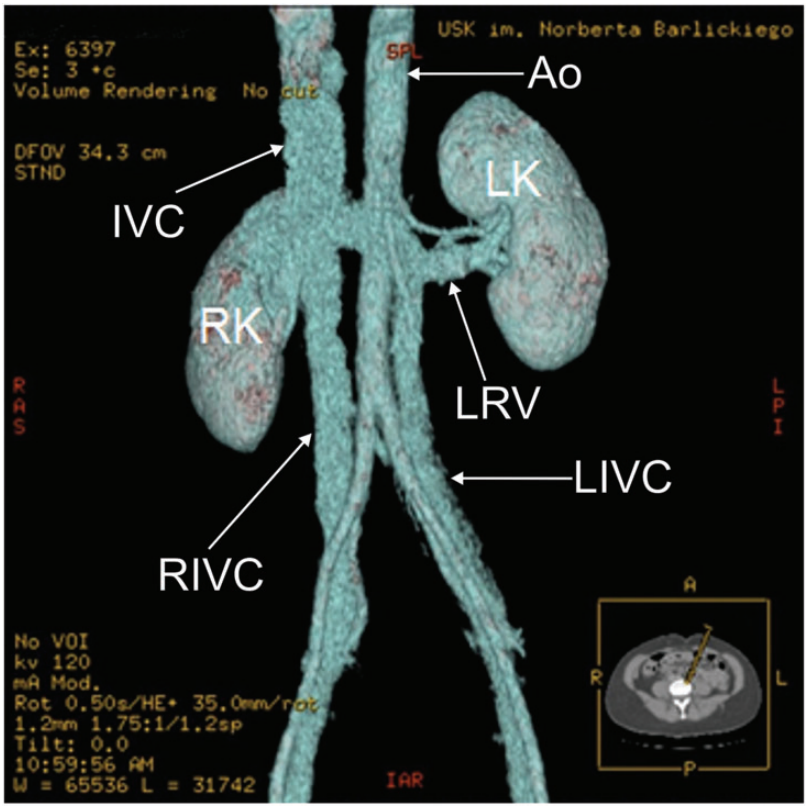

Figure 2. Three-dimensional computed tomography (CT) reconstruction of the abdomen vessels based on late arterial phase CT, showing the inferior vena cava (IVC), abdominal aorta (Ao), right inferior vena cava (RIVC), left inferior vena cava (LIVC), left renal vein (LRV), left kidney (LK), and right kidney (RK).

of this vessel.[10–12] Duplication of the IVC may be explained by numerous theories, such as persistence of the left IVC as a result of failure of the caudal left supracardinal vein to regress, or possibly a lack of anastomosis between primitive cardinal veins.[3,10] The development of renal veins is strongly connected with development of the IVC.[13]

A research study concerning IVC anomalies, by Shin et al.,[14] based the classification of double IVC subtypes on a previous study by Morita et al.[15] Both studies suggested that there are five types of double IVC (Figure 5). The most frequent type in the study by Morita et al. was duplicated IVC without communication between the iliac veins (type I, in 11/28 cases [39%]), compared with 6/23 cases (26%) in the study by Shin et al.[14,15] Interiliac branch from the left common iliac vein (type II) was the most common subtype (10/23 cases [43%]) in the study by Shin et al., compared with 5/28 cases (18%) in the Morita study. Other possible subtypes include: double IVC with communication from the left internal iliac vein (type III, in 6/28 cases [21%] and 2/23 cases [9%], Morita versus Shin study, respectively); double IVC with communication from the right internal iliac vein (type IV, in 5/28 cases [18%] and 3/23 [13%], Morita versus Shin study, respectively); and the least common type in both studies, double IVC

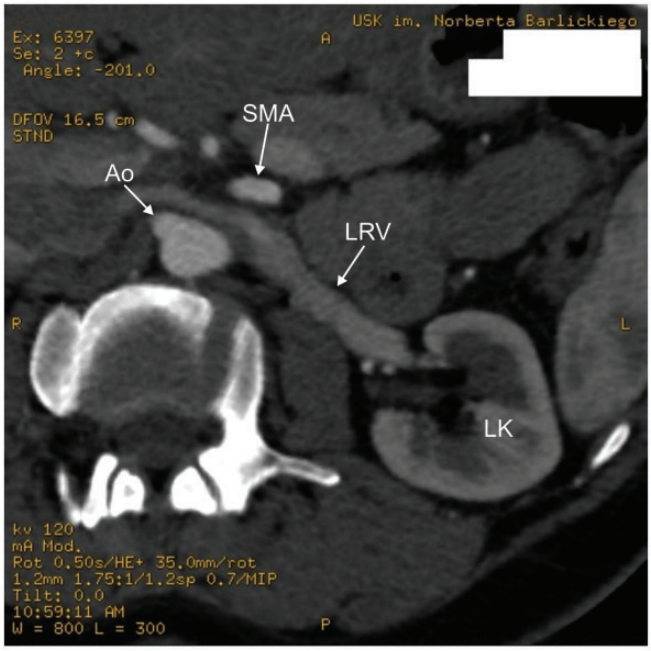

Figure 3. Helical computed tomography (CT) angiography, transverse scan at the L2 level, showing the left renal vein (LRV) compressed between the abdominal aorta (Ao) and the superior mesenteric artery (SMA); the left kidney (LK) is also shown.

with interiliac communication from the right common iliac vein (type V, in 1/28 cases [4%] in the Morita study and 2/23 cases [9%] in the Shin study).[14,15] The present case may be classified as type I (duplicated IVC without communication between the iliac veins). Although the presence of double IVC is usually asymptomatic, patients with this anomaly have an increased tendency to develop thromboembolic events.[16,17]

The nutcracker phenomenon is a condition involving pressure on the LRV by another anatomical structure, that results in dilatation of the vein before the compression, and narrowing behind the compression.[6] Entrapment of the LRV is usually asymptomatic, which is why it goes mostly undiagnosed or is diagnosed by chance during medical imaging examinations for other causes. This vessel anomaly accompanied by symptoms (the most common of which are haematuria, left flank pain, varicoceles in men or congestion syndrome in women) is called nutcracker syndrome.[6] Nutcracker syndrome may also manifest along with pelvic pain, as was reported by the present patient.[6,7] Nutcracker syndrome may be confirmed during the following medical imagining examinations: Doppler ultrasonography, CT, magnetic resonance imaging (MRI), phlebography, or intravascular ultrasound. CT or MRI may reveal a reduced angle between the abdominal aorta and the superior mesenteric artery, but for a definitive diagnosis, an angle less than 35° is required.[7] In the present description of the coexistence of duplicated IVC and nutcracker syndrome, the angle between the superior mesenteric artery and the abdominal aorta was 32°, fulfilling the previously published definition of the required angle for a diagnosis of nutcracker syndrome.[7]

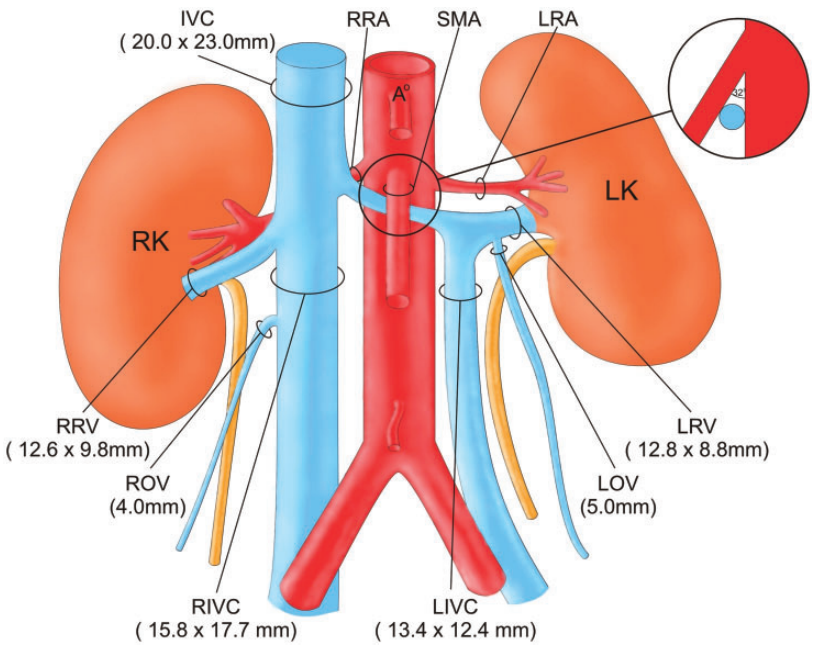

Figure 4. Schematic of vessel arrangements and dimensions in the present case, obtained during computed tomography angiography, showing the inferior vena cava (IVC) above the level of the renal veins, abdominal aorta (A°), right renal artery (RRA), left renal artery (LRA), superior mesenteric artery (SMA), right kidney (RK), left kidney (LK), right renal vein (RRV), left renal vein (LRV), right ovarian vein (ROV), left ovarian vein (LOV), and the right inferior vena cava (RIVC) and left inferior vena cava (LIVC) below the level of the renal veins.

The most frequent types of nutcracker syndrome are described as anterior, compression of the LRV between the superior mesenteric artery and the abdominal aorta, and posterior, a retroaortic LRV or a circumaortic double LRV entrapped by the abdominal aorta and the vertebral column. However, there are multiple pathologies and conditions that may lead to compression of the LRV, and nutcracker syndrome can also appear on the right side.[6,18] Treatment of nutcracker syndrome depends on a person's age and degree of symptoms. For example, patients aged under 18 years should simply be observed over a 24-month period before considering surgical treatment.[19,20] In paediatric patients, compression of the LRV by the superior mesenteric artery may be relieved by an increase in BMI.[21] Adults are recommended to be observed for at least 6 months prior to implementing any treatment. Adult patients with symptoms that persist for more than 6 months, or patients under 18 whose symptoms persist after 24 months of observation, are also recommended for surgical treatment.[19,20] Possible surgical treatment options may involve open surgery or endovascular treatment. Open surgery methods include the transposition of vessels (LRV, superior mesenteric artery, left gonadal vein), left kidney autotransplantation, nephropexy, nephrectomy, or renocaval bypass, however, endovascular surgery is currently more popular in treating nutcracker syndrome because it is a less invasive approach.[7,18,22]

Due to patterns of embryological development, anomalies of the IVC usually

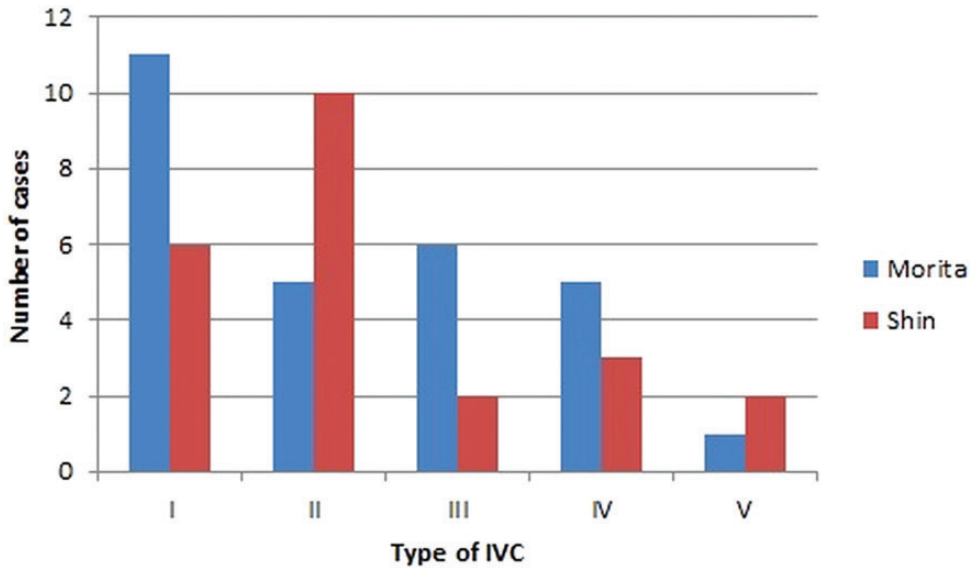

Figure 5. Classification of double inferior vena cava (IVC) subtypes, based on two previously published studies from Shin et al.,[14] and Morita et al.[15] Type I, duplicated IVC without communication between the iliac veins; Type II, communication via interiliac branch from the left common iliac vein; Type III, double IVC with communication from the left internal iliac vein; Type IV, double IVC with communication from the right internal iliac vein; and Type V, double IVC with interiliac communication from the right common iliac vein.

coexist with other vein anomalies.[23] For example, the case of a doubled IVC with retroaortic LRV, azygos continuation of the IVC, and the presence of a hepatic portion of the IVC that drained into the RRV has been reported.[8] Furthermore, a similar case in a 74-year-old male patient has been described, in which the patient was diagnosed due to an enlarged juxtarenal abdominal aortic aneurysm.[24] During imagery examination, the following anomalies in vessel course were noticed: the left IVC joined to the LRV, then crossed the abdominal aorta posteriorly, met the right IVC and continued cephalad as the azygos vein.[24] Finally, the case of a patient with suprapubic pain has been reported, in which an abdominal multi-row-detector CT revealed a double IVC, a retroaortic LRV, azygos continuation of the right IVC and a common trunk for the right and left iliac artery that drained into the right external iliac vein.[25] In all these cases, double IVC with retroaortic LRV were noticed, however, none of the cases were classified as nutcracker phenomenon or nutcracker syndrome, possibly because there is no accurate diagnostic definition for nutcracker phenomenon or nutcracker syndrome.[7]

In the present case, despite a wide space between the abdominal aorta and superior mesenteric artery, high blood flow from the lower limbs and left kidney through the LRV to the left IVC, caused reversed blood flow in left IVC. Changes in blood flow were confirmed by colour Doppler ultrasonography, which assesses dynamic blood flow under physiological conditions, and is more accurate than venography.[26] This is because in situations of low blood pressure, as in the present case, contrast injection applied before venography would have changed results. In the present case,

nutcracker syndrome in the left IVC occurred according to the respiratory phase, and that is why the present case is so exceptional. It is probable that the patient did not suffer from any symptoms, other than abdominal pain exacerbated by exertion and menstruation, because of the reversed blood flow in the left IVC.

In conclusion, duplication of the inferior vena cava and nutcracker syndrome are both rare vessel anomalies, and the coexistence of both is even more rare. Their presence may result in clinical symptoms, thus, knowledge of vessel variations is important for specialists in all clinical areas, and may prevent serious complications following medical interventions.

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Declaration of conflicting interest

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ORCID iD

Anna Waśniewska https://orcid.org/0000-0002-4845-6088

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