



Case Report

Uncommon Complex Anomaly of Inferior Vena Cava and Left Iliac Vein Demonstrated by Multidetector-Row CT Angiography

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Retroperitoneal venous anomalies have clinical importance in retroperitoneal and pelvic surgery. Multidetector-row computed tomography (CT) angiography is an important imaging method to be preferred in evaluating vascular structures in this locality. We describe a complex retroperitoneal venous anomaly with a multidetector-row CT angiography.

Key words: Complex venous anomaly – Inferior vena cava – Iliac vein – Multidetector-row CT imaging

Congenital venous abnormalities in the retroperitoneal space are relatively infrequent, and are usually recognized incidentally. These anomalies have clinical importance in retroperitoneal and pelvic surgery.¹ The inferior vena cava (IVC) is formed by the junction of the common iliac veins. Congenital anomalies of the IVC are uncommon, having an incidence of 0.5% to 3%.² The most common major venous anomalies occurring in the retroperitoneum include transposition of the IVC, the double IVC, the circumaortic renal collar, and the retroaortic renal vein. Less common anomalies include the absence of the IVC, the absence of the hepatic segment of the IVC with azygos continuation, and the absence of the infrarenal segment of

the IVC with preservation of the suprarenal segment.

The position of the iliac veins is described according to the corresponding arteries. The common iliac veins are formed by the junction of the internal and external iliac veins. The anatomic variations and the congenital anomalies of these vessels have been well explained.³ We reported a complex anatomic variation of retroperitoneal veins, which, to the best of our knowledge, has not been mentioned before in the literature.

Case Report

A 20-year-old man had a history of abdominal bloating for several months and recent occasional

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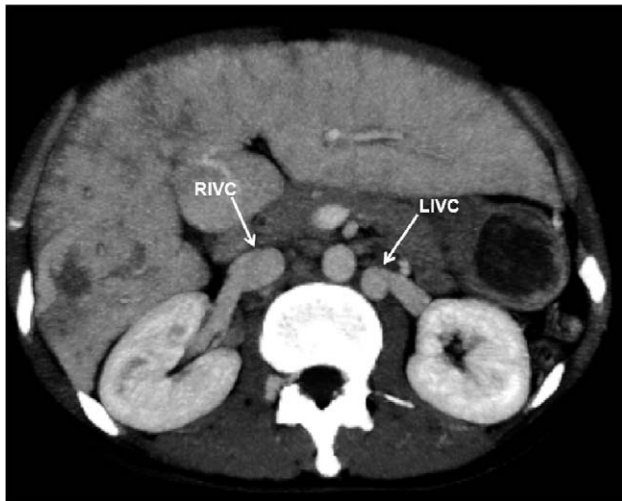


Fig. 1 On the level of the renal hilum, CT scan shows the left renal vein opening into the left IVC (arrow). RIVC, right IVC; LIVC, left IVC.

pain in the right upper abdomen. Physical examination showed upper abdominal tenderness and a palpable mass in the right upper abdomen. Laboratory data showed slightly elevated alkaline phosphatase levels. Tests for tumor markers were negative. As a part of routine parasite panel, a serologic test (enzyme-linked immunosorbent assay) was positive for echinococcosis. Other biochemical/serologic tests and medical screening findings were normal.

Multidetector-row computed tomography (MDCT) scan protocol was performed with a 16-MDCT scanner (Aquilion, Toshiba Medical Systems, Tokyo, Japan). A total of 100 mL of 300 mg I/mL iodinated contrast material (Omnipaque, Amersham Health, Cork, Ireland) was injected through the antecubital vein at a rate of 3.5 mL/s. A late arterial and early venous phase volumetric data set was acquired at 30 and 65 seconds, respectively, from the start of the intravenous injection of the contrast material. Abdominal MDCT demonstrated a hypoattenuating mass with central calcifications in the right lobe of liver. A variant drainage pattern of retroperitoneal veins was also incidentally found on contrast-enhanced axial maximum-intensity projection (MIP) images (Fig. 1). Then, coronal reconstruction and 3-dimensional volume-rendered images were performed for accurate visualization of the iliac veins, renal veins, and IVC. The abdominal aorta was in normal localization, with a normal branching pattern. The right IVC and right renal vein were in normal location. There was a normal

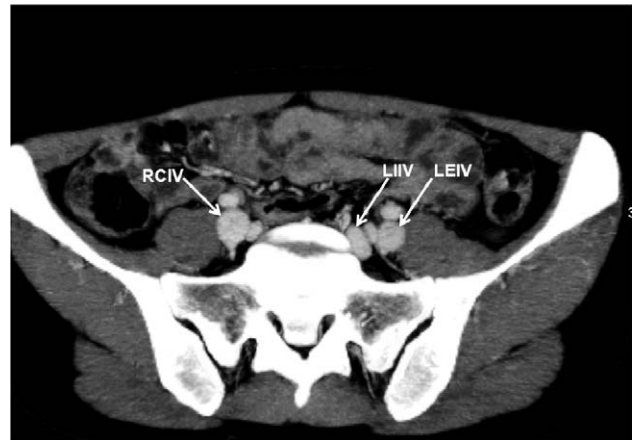


Fig. 2 The axial image at the iliac vessel level reveals both EIV and IIV on the left side of the pelvis, and CIV on the right side of the pelvis. RCIV, right common iliac vein; LIIV, left IIV; LEIV, left EIV.

confluence of the right common iliac vein into the right IVC. The left common iliac vein was absent (Fig. 2). The left external iliac vein (EIV) was showing continuity with the left IVC. There was a connection vein between the left EIV and distal segment of the right IVC (Fig. 3). The left renal vein was draining into the left IVC. At the infrarenal level, a thin connection vein with retroaortic course was observed between the right IVC and the left IVC (Fig. 4). Coronal MIP image clearly shows a persistent enlarged azygos vein (Fig. 5). The patient had no other anomaly detected by MDCT. He was also given a diagnosis of having a hepatic alveolar echinococcosis.

Discussion

Knowledge of the embryologic development of the retroperitoneal venous system is necessary to understand its congenital anomalies.¹ The caval system is derived from the modification of 3 pairs of primitive veins: the posterior cardinal, subcardinal, and supracardinal veins. These veins arise in a chronologic order between 4 and 8 weeks of fetal life. The posterior cardinal veins are the dominant system at 6 weeks of embryonic life, and they drain the caudal portion of the embryo. These veins do not form any part of the adult IVC, but abnormal development can lead to an anomaly.² In a 7-week-old embryo, the subcardinal veins become dominant. They are located in ventromedial to the posterior cardinal veins. During the eighth week of embryonic life, the supracardinal veins start to

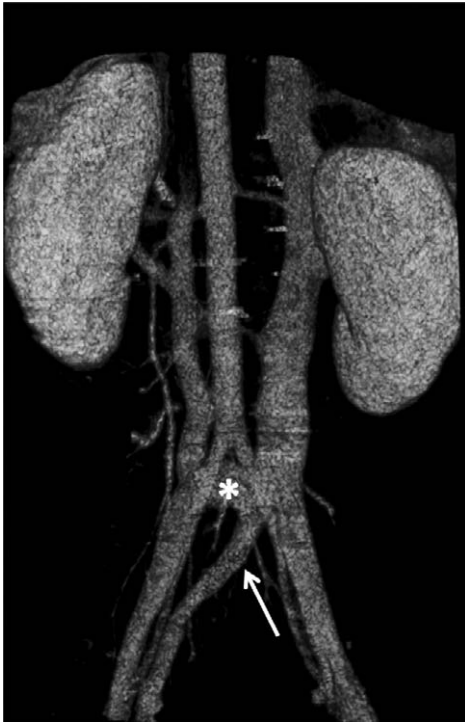


Fig. 3 Volume-rendered image viewed from a posterior coronal view shows a thick connection vein (asterisk) between the left EIV and the distal segment of the right IVC. It also shows the left IIV opening into the right common iliac vein (arrow).

predominate. These veins are located lateral to the subcardinal veins. Cranially, the supracardinal veins extend above the diaphragm to become azygos and hemiazygos veins. Caudally, the right supracardinal vein forms an anastomosis with the iliac veins, which are derived from the persistent posterior cardinal veins.² The interiliac communicating vein is considered to be originated from iliac anastomosis between posterior cardinal veins.³ The IVC is composed of 4 segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment is derived from the vitelline vein. The right subcardinal vein forms the suprarenal segment of the IVC. The infrarenal segment is believed to be derived from the right supracardinal vein. Anastomotic communications between the supracardinal and subcardinal veins form the intervening renal segment of the IVC. These anastomotic channels form a collar of veins encircling the aorta. If the dorsal portion of the circum-aortic collar persists, the left renal vein is located posterior to the aorta, forming a retroaortic left renal vein.⁴

Congenital anomalies of the IVC can be classified under the main headings of suprarenal segment,

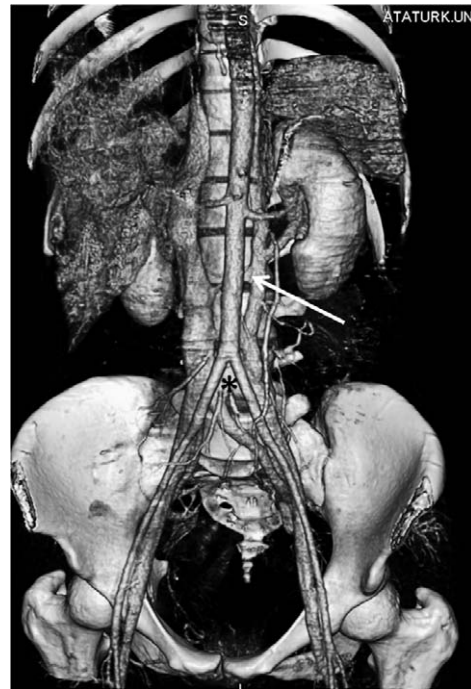


Fig. 4 Volume-rendered 3-dimensional image viewed from an anterior coronal view clearly shows the double-IVC anomaly, and a thick connection vein (asterisk) between the left EIV and the distal segment of the right IVC. It also shows a thin connection vein (arrow) with a retroaortic course between both the right IVC and the left IVC, as well as left iliac vein abnormality.

renal segment, and subrenal segment anomalies. Regarding double IVC, which is one of the important anomalies of the subrenal segment, there is a subject about persistence of the bilateral supra-cardinal veins.⁵ In this anomaly there are 2 vena cavae on both sides of the aorta, just inferior to the renal hilus. At the renal hilus level, the left IVC unites with the right IVC by passing in front of the aorta, with contribution from the left renal vein. The right suprarenal IVC has been formed with normal anatomic integrity and through a normal course. Among the suprarenal segment anomalies of IVC are azygos continuity of the IVC and hemiazygos continuity of the left IVC. Minniti *et al*⁵ classified the congenital anomalies of the vena cava according to embryologic origin and imaging features. In their articles, researchers defined 3 new variations, under the heading of complex anomalies, in addition to this classical classification. One of the complex anomalies that was reported as a new variation by the researchers is double-IVC anomaly, in which the left IVC shows continuity with hemiazygos and azygos veins. In this anomaly, at the suprarenal



Fig. 5 Coronal MIP image shows enlarged azygous vein (arrow). It also shows a thick connection vein (asterisk) between the left EIV and the distal segment of the right IVC.

level, whereas the right IVC continues its normal course, the left IVC shows hemiazygos vein continuity, passes to right side by crossing the aorta at the diaphragm level, and drains into the azygos vein. Azygos vein, which as increment at calibration has been drained into the right superior vena cava through its normal course. Similar to this complex anatomic variation, which was defined by Minniti *et al*,⁵ in our patient there was a double-IVC anomaly at the subrenal level, and hemiazygos and azygos vein continuity of the left IVC. But in our case additional complex anomalies also were present.

In a recent study, Morita *et al*⁴ examined the subject of pelvic venous variations in cases with congenital IVC anomaly. In this study, for a rare anatomic variation that was defined as type 2d, it was emphasized that the left lower extremity veins were continuing directly as left IVC to the left of the

aorta, without uniting with the internal iliac vein (IIV), and the left IIV was being drained into the right IVC, uniting with the right main iliac vein. Our case also was partially similar to this rare anatomic variation defined by Morita *et al*,⁴ but it differs from that variation in a few important ways. In our case, at the level of the left EIV beginning its course like the left IVC, a thick connection vein, which had an increment of calibration with the vein in question, was forming a connection with the right IVC to the slight upper of the left IIV connection level. Furthermore, the left IVC passage to the right side in front of the aorta at the renal level was not observed in our case. But we observed a thin connection vein that has a retroaortic course and has low calibration under the renal hilus level, between the left IVC and the right IVC (Fig. 6).

In the detection of retroperitoneal vascular anatomy and anatomic variations, Doppler ultrasonography can be the primary preference in radiologic modality. Yet, that this technique is operator dependent, as well as the fact that its abdominal gas distention constitutes a serious limitation, restricts the use of this method.⁶ CT and magnetic resonance angiography are 2 important imaging methods to be preferred in evaluating vascular structures in this locality.⁷ We describe a complex vena cava anomaly with an MDCT angiography, which has not yet been described in the literature. We believe that this anomaly occurs as a result of some conditions, including the persistence of bilateral supracardinal veins (double IVC) in the subrenal tract, the persistence of the left supracardinal vein in the suprarenal tract, and the persistence of the subcardinal vein at the pelvic level.

The presence of IVC anomalies can affect the outcomes of surgical and interventional procedures, such as abdominal aortic aneurysm repair, nephrectomy, renal transplantation, therapeutic spermatic/ovarian vein embolization, renal/adrenal vein sampling, and IVC filter placement.⁸ First, performing a correct anatomic evaluation is very important before renal transplantation. In order to perform a successful renal transplantation, it is imperative to secure the optimal length of the donor renal vein and to isolate the recipient renal vein.⁹ For live-donor nephrectomy, the left kidney is normally preferred because it has a longer renal vein, but there are several reports of successful renal transplantation using the left kidney with its short renal vein from donors with a left-sided or duplicate IVC.¹⁰ In addition, the left-sided limb of a duplicate IVC was sacrificed to increase donor renal vein length, but

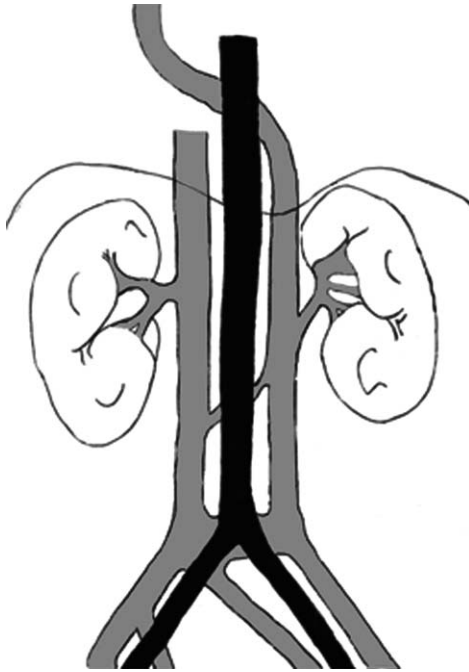


Fig. 6 Drawing illustrates the complex anomaly of the IVC and the left iliac veins.

this caused ipsilateral edema in the donor's pelvis and thigh, and is not to be recommended.¹¹ Second, the presence of IVC anomalies can cause technical difficulties during aortoiliac surgery, and patients are most likely to suffer severe bleeding; thus, the surgeon must be alert to the presence of these anomalies and to treat them correctly to avoid severe injuries.¹² In such cases, a transperitoneal rather than a retroperitoneal approach to the abdominal aorta and kidneys has been recommended in patients with a duplicate IVC to enable better visualization and control of aberrant vessels.¹⁰ Third, the presence of IVC anomalies can affect venous access, and the number and localization of filters to be used during the IVC filter placement procedure. Finally, left-sided and duplicate IVC may predispose a patient to thromboembolism because of changes in blood flow induced by the anomalous IVC, or compression of the vessel or its major tributaries.¹⁰

As a result, being unaware of the anatomic variations of retroperitoneal veins can lead to unfavorable results, especially in the cases in which surgery is planned in this region. With the common

use of CT, new anatomic variations are continuously being identified. Today, by means of the advances in CT technology, even the most complex vascular anomalies of this region can be successfully imaged with MDCT angiography, and thus the surgeon gains the upper hand before retroperitoneal and pelvic surgery.

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