Combined Anomaly of the Right Hepatic Lobe Agenesis and Absence of the Inferior Vena Cava: a Case Report

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The absence of the inferior vena cava is an uncommon congenital anomaly that has recently been identified as an important risk factor contributing to the development of deep venous thrombosis. Congenital agenesis of the right hepatic lobe is a rare anomaly which is found incidentally in radiologic examinations. We present a case of a congenital absence of the infrarenal inferior vena cava, combined with agenesis of the right hepatic lobe in a 62-year-old man presented with symptoms of deep venous thrombosis.

Index terms:
Inferior vena cava abnormalities
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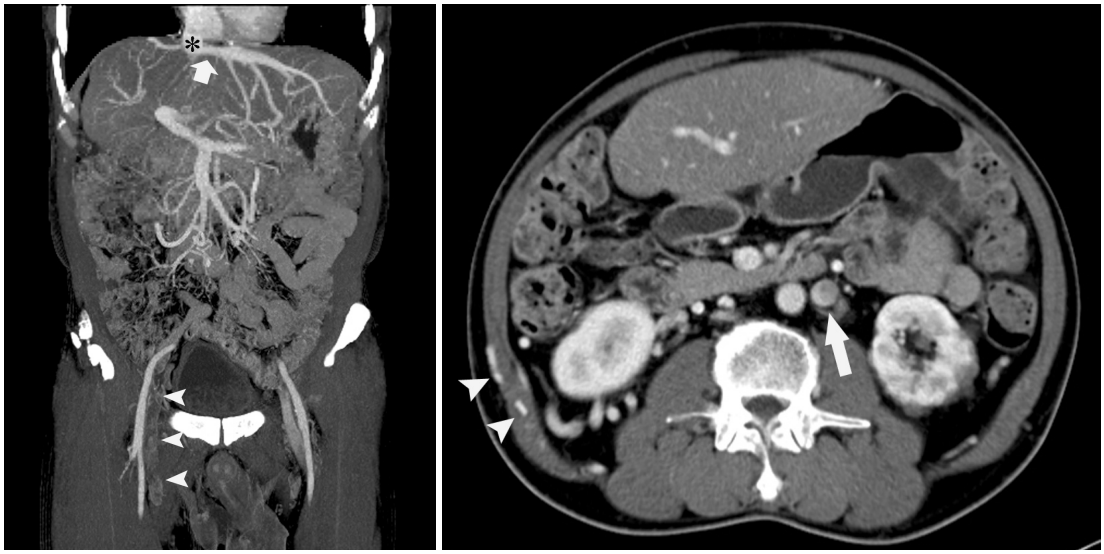
Congenital absence of the inferior vena cava (IVC) is a rare vascular anomaly resulting from the aberrant development during embryogenesis (1, 2). A recent report suggests that an anomaly of the IVC is a strong predisposing factor for the development of deep venous thrombosis (DVT), which presumably occurs due to venous stasis (3).

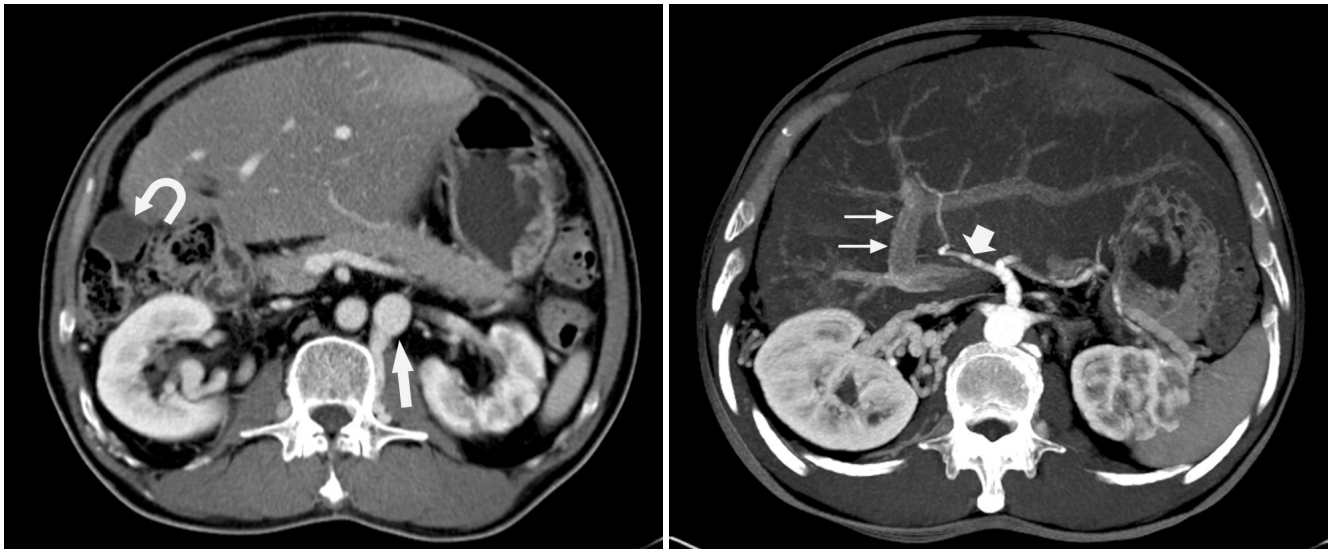
Agenesis of the right hepatic lobe is an extremely rare congenital anomaly (4, 5) which is usually accompanied by a retrohepatic gallbladder, partial or complete absence of the right side of the diaphragm, and intestinal malrotation (4, 5). Because no characteristic symptom has been identified, most patients with agenesis of the right hepatic lobe usually remain clinically latent. Agenesis of the right lobe of the liver is most easily diagnosed by computed tomography (CT) and magnetic resonance imaging (MRI) when no liver parenchyma is present on the right side of the gallbladder fossa (4, 5).

In this report, we describe a 62-year-old male patient with right hepatic lobe agenesis, combined with DVT which was caused by congenital absence of the IVC. To the best of our knowledge, no previous descriptions of this malformation exist.

CASE REPORT

A 62-year-old man was admitted to our hospital with advanced stages of swelling in the right lower extremity. The patient had been treated at another hospital for DVT of the lower extremity seventeen years earlier. The patient had no apparent risk factors for any thromboembolic diseases, including recent trauma, surgery, immobilization or familial history of thromboembolism. A physical examination revealed the patient's edematous swelling and redness in the right lower extremity. The patient's peripheral arterial pulses were normal. A color Doppler ultrasonography (US) revealed the complete thrombosis of the right external iliac and femoral vein. We performed a contrast enhanced abdomino-pelvic CT and CT angiography using a multi-detector row CT (Somatom Sensation 64, Siemens, Forchheim, Germany) to define the extent of the disease, which revealed the low density thrombosis of the right femoral and





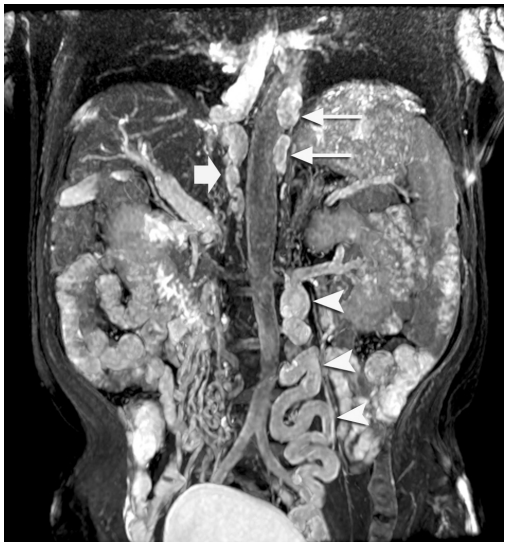


Fig. 1. 62-years-old patient with right hepatic lobe agenesis, combined with deep venous thrombosis caused by congenital absence of inferior vena cava.
A. Coronal thick (30 mm) slab maximum intensity projection CT image showed thrombus (arrowheads) occupying entire lumen of right femoral and external iliac vein. Left hepatic vein (arrow) drains into inferior vena cava (asterisk).
B. Transverse CT scan obtained inferior to kidney shows missing IVC. Enlarged left testicular vein (arrow) and prominent anterior abdominal wall collateral veins (arrowheads) are present.
C. Transverse contrast-enhanced CT scan obtained at hilus level of left kidney shows missing infrarenal segment belonging to IVC. Enlarged ascending lumbar vein and prominent left testicular vein (arrow) are present. Right hepatic lobe is absent and gallbladder (curved arrow) is identified in retrohepatic position.
D. Transverse thick (30 mm) slab maximum intensity projection CT image of arterial phase shows left hepatic artery (large arrow) arising from celiac axis. Complete absence of right hepatic artery branch is noted, along with left portal vein (small arrows).
E. Coronal MIP of two-dimensional time-of-flight MR image shows absence of infrarenal IVC, with enlarged azygos (large arrow) and hemiazygos vein (small arrows). Tortuous and enlarged left testicular vein (arrowheads) is also noted.

external iliac vein (Fig. 1A), as well as the nonexistent infrarenal segment of the IVC and the common iliac veins (Fig. 1B). The suprarenal IVC originates from the confluence of the renal veins. In the study patient, the hepatic vein was connected to the infrahepatic segment of the IVC and the IVC drained normally into the right atrium. Furthermore, a prominent azygos vein, hemiazygos vein, paravertebral venous plexus, lumbar vein, and left testicular vein were seen. The left renal vein was connected to the left testicular vein and also drained into the dilated lumbar vein (Fig. 1C). The bilateral, external, and internal iliac veins were drained to enlarge the azygos and hemiazygos veins via the ascending lumbar vein and anterior paravertebral venous plexus.

Incidentally, an abdominal CT scan revealed the nonexistence of the right hepatic lobe and compensatory hypertrophy of the left hepatic segment. The gallbladder was identified in a retrohepatic position, coursing laterally and horizeontally (Figs. 1C, D). The liver parenchyma was homogeneous in appearance and of normal attenuation. We used a computer workstation for image analysis, and the axial and coronal maximum intensity projection (MIP) images revealed the absence of the right hepatic vein, right portal vein, and its branches (Fig. 1D).

To evaluate the combined congenital cardiac and visceral anomalies, MRI and MR angiographies (MRA) were performed using a 1.5 Tesla magnet MR unit (Signa HD, GE Medical system, Milwaukee, WI). The MRA revealed the agenesis of the infrarenal IVC with a dilatation of the azygos, hemiazygos vein, left testicular vein, and numerous pelvic collaterals (Fig. 1E). The absence of the right hepatic artery and right portal vein was also described.

After the diagnosis was made, the patient was treated low-molecular heparin therapy, which resulted in the gradual reduction in swelling of the right lower leg. No further evaluation was performed beyond the incidental finding of the right hepatic lobe agenesis.

DISCUSSION

Due to the development of a complex embryologic system, several congenital anomalies of the IVC exist (1, 2). Furthermore, the embryogenesis of the IVC is a complex process, primarily involving the development, regression, and fusion of three pairs of embryonic venous channels: the posterior cardinal, subcardinal, and supracardinal veins. The aberrant development of these venous systems causes anomalies of the IVC system. Furthermore, the common IVC anomalies include the left IVC, double IVC, azygos continuation of the IVC, circumaortic left renal vein, retroaortic left renal vein, circumcaval ureter, and absence of the infrarenal IVC with preservation of the suprarenal segment (1). In our case study, the patient's infrarenal segment of the IVC was absent, whereas the suprarenal segment was preserved. In the case of the absent infrarenal IVC, the external and internal iliac veins merged to form enlarged ascending lumbar veins, which subsequently transported blood, from the lower extremities to the azygos and hemiazygos veins, via the anterior paravertebral collateral veins (1, 2). A normal suprarenal IVC is formed by the confluence of the renal veins. The absence of the infrarenal IVC causes the failure in the development of the posterior cardinal and supracardinal veins (1, 2). Since a single embryonic event does not fully explain this anomaly, some authors have suggested that the absence of the postrenal IVC results from the intrauterine or perinatal thrombosis of the IVC and is not embryonic in origin (1, 6).

Venous thrombosis is an illness of clinical interest, due to the associated morbidity and mortality. DVT has a multifactorial etiology, involving both acquired and genetic factors. The acquired risk factors include surgery, trauma, pregnancy, tumors and oral contraceptives, whereas the genetic factors include protein C deficiency, protein S deficiency, antithrombin III deficiency, and hyperhomocysteinemia. Recently, anomalies of the IVC have been recognized as possible risk factors contributing to DVT (1, 2). Previous studies have described that the congenital IVC anomaly is associated with a recurrent and bilateral DVT, especially in young patients (3). This may be a consequence of inadequate blood return, through collaterals, which may increase the venous blood pressure in the veins of the legs, hence favoring venous stasis (1, 2, 6).

A congenital IVC anomaly is frequently associated with congenital heart disease, malrotation of the intestine, dysgenesis of the lungs, polysplenia or asplenia, and renal agenesis (1, 3, 7, 8). These associated anomalies were not found in our out patient; however, an abdominal CT scan revealed the absence of the right hepatic lobe and the incidental retrohepatic position of the gallbladder.

Agenesis of the right hepatic lobe is a rare congenital anomaly. Several reports have described this anomaly on a CT and US (4, 5, 9), and found the absence of the right hepatic lobe, compensatory enlargement of the left hepatic lobe, posterior interposition of the hepatic flexure of the colon, retrohepatic or suprahepatic position of the gallbladder, absence of the right hepatic artery and portal vein, and absence of the right intrahepatic ducts (4, 5). Chou et al. (4) described the criteria for diagnosis of agenesis of the right hepatic lobe on a CT as the absence of the right hepatic vein, right portal vein and its branches, as well as

dilated right intrahepatic ducts. Moreover, the diagnosis of agenesis of the right hepatic lobe of the liver is easily established with cross-sectional imaging when no liver parenchyma is found to the right of the gallbladder fossa; however, the differential diagnosis includes severe right hepatic lobe atrophy secondary to liver cirrhosis, cholangiocarcinoma, choledocholithiasis, idiopathic portal hypertension, prior fulminant hepatitis, Caroli's disease, and prior surgical resection.

The pathogenesis of this congenital anomaly is not fully explained; however, the abnormal development and thrombosis of the supplying portal venous segment during embryologic growth have been suggested as possible pathogenic mechanisms (9). Because most patients with agenesis of the right hepatic lobe usually remain clinically latent, the vast majority of cases are found incidentally. However, some patients have had symptoms due to atypical cholecystitis, choledocholithiasis, or portal hypertension (4, 5).

Advances and the increased use of noninvasive imaging techniques, such as US, CT, and MRI, have led to a more frequent detection of variant anatomy and anomalies of the IVC and liver. The Doppler US is usually the first imaging modality employed to evaluate patients with DVT; however, the detection of IVC anomalies by US is difficult (3). Other diagnostic imaging methods, including contrast-enhanced CT and MRI, are better tools for detecting anomalies of the IVC (1−4, 10). In particular, MRI is an excellent method to evaluate venous anomalies because of its multiplanar imaging. In addition, the advantages of MRI enhance the visualization of other associated congenital anomalies. The multi-detector row CT (MDCT) is another important method for noninvasively assessing the vascular system. We used a computer workstation for image analysis, as well as axial and coronal MIP images to provide characteristic details of the hepatic artery, portal vein, hepatic vein, and IVC. We were able to correctly diagnos of this congenital anomaly by means of CT and MRI.

In conclusion, the absence of the infrarenal IVC represents a potential risk factor for the development of DVT. Moreover, agenesis of the right hepatic lobe is a rare congenital anomaly which may be found incidentally in adults who undergo an abdominal CT. Lastly, MRI and MDCT are important imaging modalities which can provide accurately definition of these anomalies.

References

1. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics* 2000;20:639-652
2. Bass JE, Redwine MD, Kramer LA, Harris JH Jr. Absence of the infrarenal inferior vena cava with preservation of the suprarenal segment as revealed by CT and MR venography. *AJR Am J Roentgenol* 1999;172:1610-1612
3. Gayer G, Luboshitz J, Hertz M, Zissin R, Thaler M, Lubetsky A, et al. Congenital anomalies of the inferior vena cava revealed on CT in patients with deep vein thrombosis. *AJR Am J Roentgenol* 2003;180:729-732
4. Chou CK, Mak CW, Lin MB, Tzeng WS, Chang JM. CT of agenesis and atrophy of the right hepatic lobe. *Abdom Imaging* 1998;23:603-607
5. Inoue T, Ito Y, Matsuzaki Y, Okauchi Y, Kondo H, Horiuchi N, et al. Hypogenesis of right hepatic lobe accompanied by portal hypertension: case report and review of 31 Japanese cases. *J Gastroenterol* 1997;32:836-842
6. d'Archambeau O, Verguts L, Myle J. Congenital absence of inferior vena cava. *J Belge Radiol* 1990;73:516-517
7. Gayer G, Apter S, Jonas T, Amitai M, Zissin R, Sella T, et al. Polysplenia syndrome detected in adulthood: report of eight cases and review of the literature. *Abdom Imaging* 1999;24:178-184
8. Gayer G, Zissin R, Strauss S, Hertz M. IVC anomalies and right renal aplasia detected on CT: a possible link? *Abdom Imaging* 2003;28:395-399
9. Radin DR, Colletti PM, Ralls PW, Boswell WD Jr, Halls JM. Agenesis of the right lobe of the liver. *Radiology* 1987;164:639-642
10. Zhang L, Yang G, Shen W, Qi J. Spectrum of the inferior vena cava: MDCT findings. *Abdom Imaging* 2007;32:495-503