

Congenital agenesis of the inferior vena cava presenting with acute renal failure

Akut böbrek yetmezliği ile seyreden doğuştan inferior vena cava yokluğu

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Agenesis of the inferior vena cava (IVC) is a rare congenital disorder. Herein, we present a 35-year-old male case of agenesis of the IVC and disseminated pelvic venous congestion with complete extrinsic ureteral obstruction due to compression who was admitted with acute renal failure following contrast injection for a computed tomography scan. Renal functions of the patient returned to normal after hemodialysis. However, during follow-up, malabsorption occurred due to intra-abdominal congestion and collateral vein formation. Subsequently, he developed malnutrition and infection and died despite, a full supportive therapy.

Key words: Acute renal failure; agenesis of inferior vena cava; deep vein thrombosis; magnetic resonance imaging.

Agenesis of the inferior vena cava (IVC) is a rare congenital disorder that is usually found incidentally in asymptomatic patients who undergo intraabdominal interventions for other reasons. The most common clinical feature is deep vein thrombosis (DVT), and patients are also at risk for acute renal failure due to these abnormalities. In addition, some other congenital malformations, primarily congenital heart diseases, have been known to accompany this disorder. Treatment options include venotonic agents, elastic compression, anticoagulant therapy, and surgery.

CASE REPORT

A 35-year-old male patient was admitted to the hospital because of nausea and vomiting following abdominal contrast computed tomography (CT). He subsequently presented with a decrease in urine output and elevated blood urea and creatinine levels.

Inferior vena cava (IVC) yokluğu nadir görülen bir doğuştan bozukluktur. Bu yazıda IVC yokluğunun oluşturduğu, pelvik venöz konjesyona bağlı gelişen komplet eksternal üreter obstrüksiyonu ve bilgisayarlı tomografi görüntüleme için kullanılan kontrast maddenin neden olduğu böbrek yetmezliği ile hastaneye kabul edilen, 35 yaşında erkek hasta sunuldu. Hastanın renal fonksiyonları hemodiyaliz sonrası normale döndü. Ancak takiplerinde intraabdominal konjesyon ve kollateral ven oluşumuna bağlı emilim bozukluğu gelişti. Takiben, beslenme bozukluğunu takip eden enfeksiyonla birlikte tam destek tedavisine rağmen hasta kaybedildi.

Anahtar sözcükler: Akut böbrek yetmezliği; inferior vena cava yokluğu; derin ven trombozu; manyetik rezonans görüntüleme.

The patient suffered from disseminated varicosities in both legs at the age of 15 and underwent two stripping operations for the varices in his legs at the age of 29 and 30. Unfortunately, they later reappeared. Two months prior to coming to our facility, he had been admitted to another hospital with abdominal pain and underwent immediate surgery after being diagnosed with acute appendicitis; however, his intermittent abdominal pain continued for two months postoperatively. An abdominal contrast enhanced CT scan was then performed, and the patient was sent to our hospital because of elevated blood urea nitrogen and creatinine values.

A physical examination revealed desquamative, dark-colored skin alterations which were probably related to chronic venous insufficiency and intense subcutaneous varices on both lower extremities along with diffuse abdominal subcutaneous venous collaterals.



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In addition, hepatosplenomegaly was also observed on abdominal palpation.

In laboratory assessments, the serum urea, creatinine, and potassium levels were elevated, but all other parameters were normal. Furthermore, endoscopic gastrointestinal system investigations, transthoracic echocardiography (TTE), and a chest X-ray revealed nothing abnormal.

Renal ultrasonography (USG) showed grade 2 hydronephrosis in the right kidney and grade 4 hydronephrosis with decreased parenchymal thickness in the left kidney while contrast-enhanced CT in the portal phase revealed a discontinuity of contrast in the left portal vein that was compatible with portal vein thrombosis (PVT). Due to thrombotic occlusion, the splenic vein was not enhanced (Figure 1a). At this level, we also observed peripancreatic collateral vessels that reached the splenic hilus and found that the left kidney was longer than normal (140x80 mm) because of extremely dilated calices together with very thin (a few millimeters only) compressed parenchyma (Figure 1b).

Grade 4 hydronephrosis and a very large kidney with highly narrowed parenchyma and a prominent pelvicalyceal system were present on the right side, but there was no ureteral continuity (Figure 1c).

Magnetic resonance venography (MRV) determined that both renal veins were draining into an abnormally thin IVC, which after joining with the hepatic veins, finally reached the right atrium. Additionally, the azygos vein, originally formed by enlarged lumbar veins, joined with the left-sided hemiazygos vein at the level of the diaphragmatic crus and then drained into the superior vena cava (SVC) within the thorax (Figure 1d). The MRV also revealed an abnormally narrow IVC at the right renal hilus level with fusion of the left and the right renal veins along with collateral drainage. The azygos vein at this level joined with the paravertebral and lumbar veins. It then joined with the hemiazygos veins at the level of the diaphragmatic crus and traveled as a single vessel to arrive at the SVC on the right side of the mediastinum (Figure 1e).

The renal parenchyma tissue was narrow and damaged due to high-grade hydronephrosis, and the ureter along with the renal pelvis and calices were distended. Furthermore, there were several wide collaterals that were imposing serious pressure on the ureters in the abdomen. The highly narrowed parenchyma probably also caused the diminished excretion of the contrast media. We found normal blood urea and creatinine levels at the preoperative laboratory assessment, but after the administration

of the contrast media, they increased rapidly. All of these findings led to the diagnosis of contrast-induced nephropathy, and the patient underwent hemodialysis. After three sessions, the serum urea and creatinine levels, renal functions, and urine excretion had returned to normal. The patient was then discharged and follow-ups were scheduled. During the follow-up period, malabsorption and gastrointestinal tract compression occurred due to intraabdominal congestion and collateral vein formation. As a result, the patient developed malnutrition followed by a generalized infection. Although he received full supportive therapy, the patient died at the end of the third month after the hemodialysis treatments

DISCUSSION

Agenesis of the IVC with azygos continuation is a rare congenital disorder. It is usually asymptomatic and is diagnosed as a result of interventions and radiological examinations of the retroperitoneal region performed for other reasons. Congenital malformations of other cardiovascular, tracheobronchial, and visceral organs accompany this disorder.^[1]

In 1793, Abernethy first described an abnormality in the IVC when he reported on a 10-month-old child who had an IVC with a mesocaval shunt and azygos continuity as well as polysplenia and dextrocardia.^[2]

Following advances in imaging techniques, the condition has been reported in asymptomatic patients with increasing frequency and diversity at estimated rates ranging from 0.07-8.7% of the general population.^[3,4] However, the rate is higher among patients with congenital heart disease.

Other congenital diseases accompany IVC abnormalities at rates ranging from 0.6-2%^[5,6] and can coexist with congenital heart diseases such as dextrocardia as well as interatrial and interventricular septal defects. In addition, abnormalities including situs inversus, hypoplasia of the biliary tract, polysplenia, intestinal malformations, and portal vein agenesis have also been noted.^[5,6]

Anatomically, the IVC is comprised of the hepatic, suprarenal, renal, and infrarenal segments and is located to the right of the vertebral column. The IVC and its branches are formed between the fifth and eighth embryonic weeks by a complex combination involving the anastomosis of three vein pairs (posterior cardinal, subcardinal, and supracardinal). The posterior and subcardinal veins, which form in the fifth embryonic week, anastomose to the supracardinal vein in the sixth embryonic week. In the seventh week of embryonic

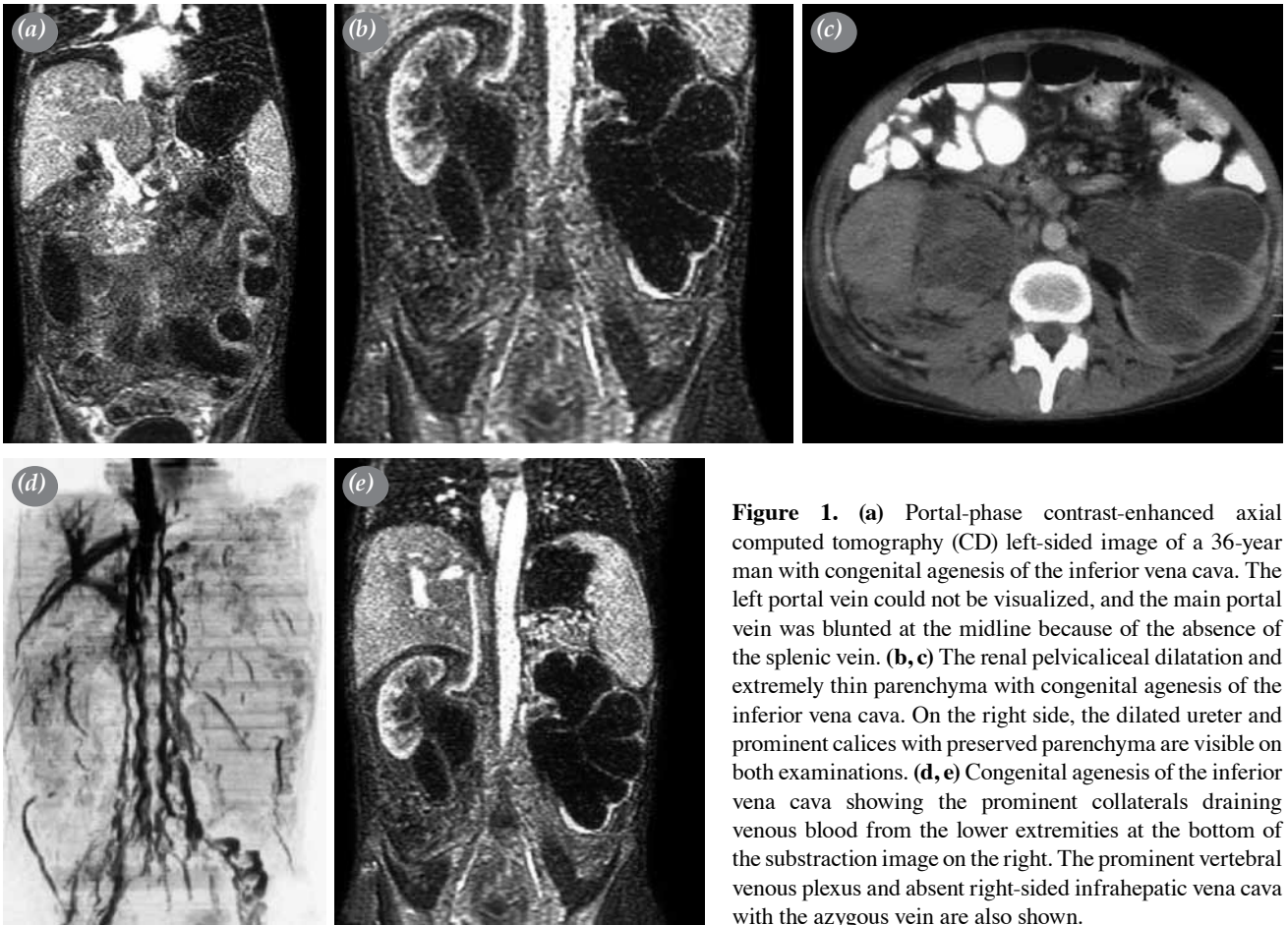


Figure 1. (a) Portal-phase contrast-enhanced axial computed tomography (CD) left-sided image of a 36-year man with congenital agenesis of the inferior vena cava. The left portal vein could not be visualized, and the main portal vein was blunted at the midline because of the absence of the splenic vein. (b, c) The renal pelvicaliceal dilatation and extremely thin parenchyma with congenital agenesis of the inferior vena cava. On the right side, the dilated ureter and prominent calices with preserved parenchyma are visible on both examinations. (d, e) Congenital agenesis of the inferior vena cava showing the prominent collaterals draining venous blood from the lower extremities at the bottom of the subtraction image on the right. The prominent vertebral venous plexus and absent right-sided infrahepatic vena cava with the azygos vein are also shown.

development, regression is seen in some segments while the subcardinal and the supracardinal veins combine via transverse anastomosis to form the IVC, which then collects the diaphragmatic blood. The iliac tract, which spans the length of the body, then forms at the posterior cardinal vein, and the right subrenal tract is formed from the right supracardinal vein. In addition, the renal tract from the anastomosis between the right supracardinal and subcardinal veins, the suprarenal tract from the right subcardinal vein, and the hepatic tract from the hepatocardiac canal are also formed. Abnormal persistence or regression that occur at this time cause the development of vena cava abnormalities.^[7-9]

Several malformations of the IVC have been reported. Left IVCs^[10] are seen when the IVC ends at the left renal vein and passes from the anterior of the aorta to form a normal right IVC. The prevalence rate for this anomaly ranges from 0.2-0.5%.^[11]

Another malformation is the double IVC in which the left IVC ends at the left renal vein, crosses the

aorta anteriorly, and drains into the right IVC.^[10] It is formed by the persistence of both supracardinal veins and has a prevalence rate of between 0.2 and 3%.^[11] In Turkey, Apaydin et al.^[12] identified three cases of this abnormality in a series of 2,000 patients who were investigated for malignancy.

Azygos continuity of the IVC has a prevalence rate of approximately 0.6%.^[11] This is found when the IVC continues alongside the azygos vein in the absence of the hepatic segment and is caused by the atrophy of the right subcardinal vein due to the lack of the right subcardinal and hepatic anastomosis.

In addition, it is possible for there to be two left renal veins surrounding the aorta. In these cases, the left superior renal vein takes the left adrenal vein and crosses the aorta anteriorly while the left inferior renal vein crosses the aorta posteriorly 1-2 cm below the adrenal vein. This anomaly has a prevalence rate of 8.7%,^[11] making it the most common.

Other rare abnormalities that have been identified via novel cross-sectional radiological examinations

include a retroaortic renal vein in which the left renal vein crosses the aorta posteriorly, complete IVC agenesis, a circumcaval ureter, a double right IVC, segmental septation of the IVC, a double IVC, and continuity of the left-sided IVC with the azygos vein, and lipoma in the IVC. A prevalence rate of 2.1% has been reported for all of these anomalies.^[13-15]

Being aware of these abnormalities, which are often asymptomatic, is crucial for preventing complications. Furthermore, patients with decreased renal filtration due to insufficient venous return and mass effect on the ureters caused by dilated vessels run the risk of acute renal failure; therefore, contrast media injections should be used with extreme caution and only when absolutely necessary. In addition, surgeons and interventional radiologists should be warned about these potential complications.

The most common clinical feature in agenesis of the IVC is deep vein thrombosis (DVT), which is often found in young patients who have no predisposition. Insufficient collateral formation is the most important risk factor associated with DVT. In two studies by Obernosterer et al.,^[16,17] 16% of DVT patients, the majority of whom were young, had IVC abnormalities.

Impaired venous return of the lower body system results in the formation of wide collaterals in the pelvic and thoracic regions while an enlarged venous plexus at the paravertebral region causes mass effect. Since the venous return diminishes during activity, especially when the intrathoracic pressure increases, the mass effect of the vertebral venous collaterals becomes more severe. Another clinical problem that is sometimes seen on X-rays occurs when an image of an enlarged azygos vein may be misdiagnosed as aortic root dissection, mediastinal mass, or paratracheal lymphadenopathy. Other clinical issues that may be encountered are collaterals at the pelvic region, hematuria due to stasis, renal colic, acute abdominal disorder, and hemoptysis due to bronchial vein rupture.

Treatment options include venotonic agents and elastic compression. For patients with DVT, anticoagulant therapy may also be prescribed, with conservative doses usually being sufficient to provide relief. Since thrombotic recurrence is expected in agenesis of the IVC, lifelong oral anticoagulation should be considered. Elastic stocking support and leg elevation can also be used to prevent venous insufficiency, and patients should be advised to avoid additional risk factors, such as prolonged immobilization and oral contraceptive use.^[18] Surgical

interventions have rarely been reported in the literature,^[19-21] but they may be indicated for patients who show deterioration in the clinical stage of IVC or for acute thrombosis of the iliofemoral veins, which can lead to a complete collapse of the collateral system. Surgical treatment options consist of arteriovenous fistula (AVF) construction and either a thrombectomy or replacement of the IVC.^[22]

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