CASE REPORT

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Vena cava inferior agenesis recognized by incidentally in a patient under cholecystectomy

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Abstract

Inferior Vena cava (IVC) agenesis is a rare congenital anomaly, which may cause significant consequences with regard to morbidity and mortality. In this case report, a patient diagnosed with IVC agenesis which was seen intraabdominal intensive venous collateral during surgery for cholecystectomy is presented.

Keywords: Inferior Vena Cava, Agenesis, Venous Anomalies, Abnormal Varicosities.

INTRODUCTION

Inferior vena cava (IVC) is the major venous collecting system that brings venous blood from the legs, pelvis, and abdominal organs to the right atrium. Changes in the development process of the IVC between the 6th and 8th weeks of intrauterine life give rise to some developmental anomalies (1). The congenital anomalies of the IVC affect approximately 0.5% of the general population (2). Congenital IVC agenesis is a rare anomaly, and its incidence is 0.0005%-1% (3).

In patients with IVC agenesis, venous drainage from the lower extremities is provided via the well-developed ascending lumbar veins within the azygos and hemiazygos system (4, 5). When preoperatively undiagnosed anomalies are found incidentally intraoperatively, the management of the surgical procedure is vital. Intraoperative bleeding caused by venous anomalies can be life threatening. The bleeding complications increase up to 40% when abnormal venous formations are recognized intraoperatively (6).

IVC agenesis may be associated with cardiovascular, tracheobronchial, and other visceral malformations (7), but may also show a silent course symptom until advanced ages (3). In cardiovascular surgery, this anomaly often causes pelvic congestion symptoms, deep vein thrombosis (3,8), and rarely pulmonary thromboembolism (3). In addition, it may be accidentally detected during abdominal surgeries (6) or may occur as an unexpected technical problem at cannulation during thoracic cardiovascular surgery. Thus, IVC agenesis may be a cause of morbidity, and mortality and negatively affect the surgical results. We present a patient who underwent surgery for cholecystectomy with intense venous collateral vascularization in the abdominal region.

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CASE REPORT

A 60-year-old female patient with a history of cholecystectomy was admitted to our clinic. Her patient file indicated that intense vascular formations were incidentally observed in the intraabdominal area during the operation for cholecystectomy. The surgery was initially planned laparoscopically. Open surgery was performed due to bleeding due to traumatization of venous collaterals during the laparoscopic procedure, which disrupted the surgical procedures. The gallbladder was reached by tying or clipping the venous collaterals observed during exploration. There were no additional complications except for non-major bleeding. Physical examination of the patient showed intense venous collaterals development on the right side of the abdominal area. Contrast-Enhanced Computed Tomography (CECT) scanning in venous phase for further examination was performed. The scanning revealed the complete absence of IVC. IVC was partially observed at the infra-renal level and was not observed at more proximal levels. Then, the IVC was observed normally at the diaphragm level and it opens into the right atrium. There were widespread collateral venous structures in the abdomen. The superior and inferior mesenteric veins drain through the collaterals to the portal vein, from there to the hepatic veins, and then to the IVC. IVC diameter was measured as 16-17mm at the crus diaphragm level. The portal vein diameter was measured as 22.2mm at the hepatic hilus level and it was larger than normal. Hepatic veins were dilated. Ovarian veins drained into the right and left renal veins on both sides, and the left renal vein was dilated. Widespread, dilated vascular structures were noted at the base of the superior and inferior mesenteric veins, in the left lower quadrant, at the pelvic level, in the subcutaneous fatty tissue on the abdominal wall. Heterogeneity was observed in the uterine parenchyma, and widespread vascularization in the myometrium was noted (Figures1- 4).

Informed consent was obtained from the patient.

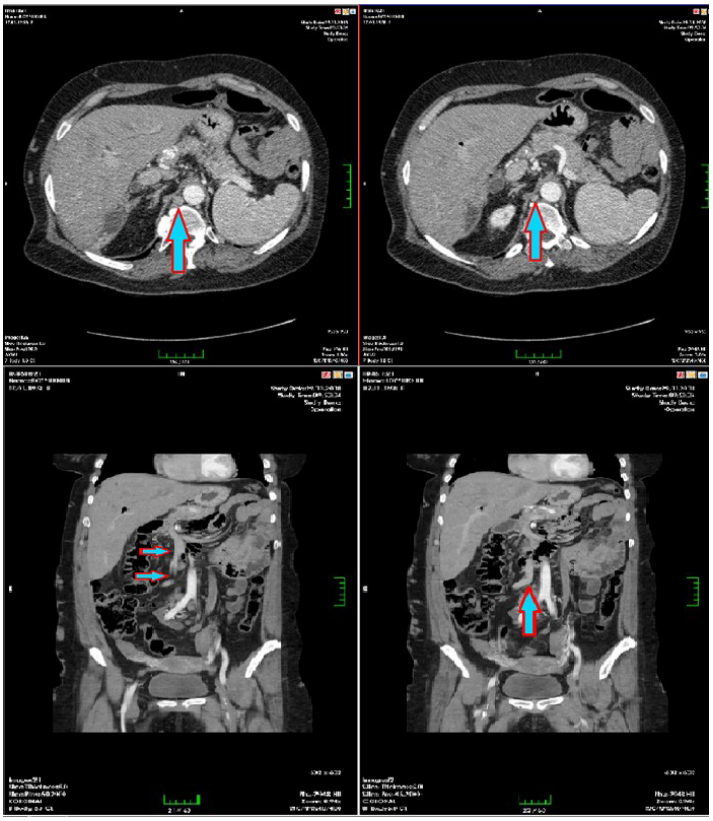

Figure 1: In I.V. contrast-enhanced CT sections; The IVC calibration narrows starting from below the diaphragm level; Its lumen below the level of the right vena renalis shows completely stenotic atresia (Partial IVC Agenesis).

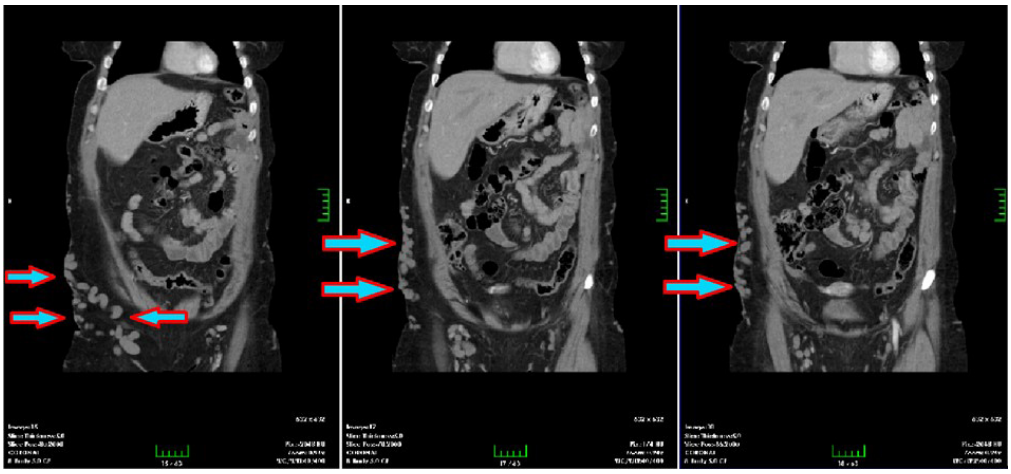

Figure 2: In I.V. contrast-enhanced CT sections taken in the axial plane; In order to provide venous system drainage secondary to infra-renal IVC partial agenesis, widespread tortuous venous vascular structures with increasing calibration and ectatic dilatation attract attention within subcutaneous adipose tissue planes in the right abdominal lower quadrant and pelvic level abdominal walls.

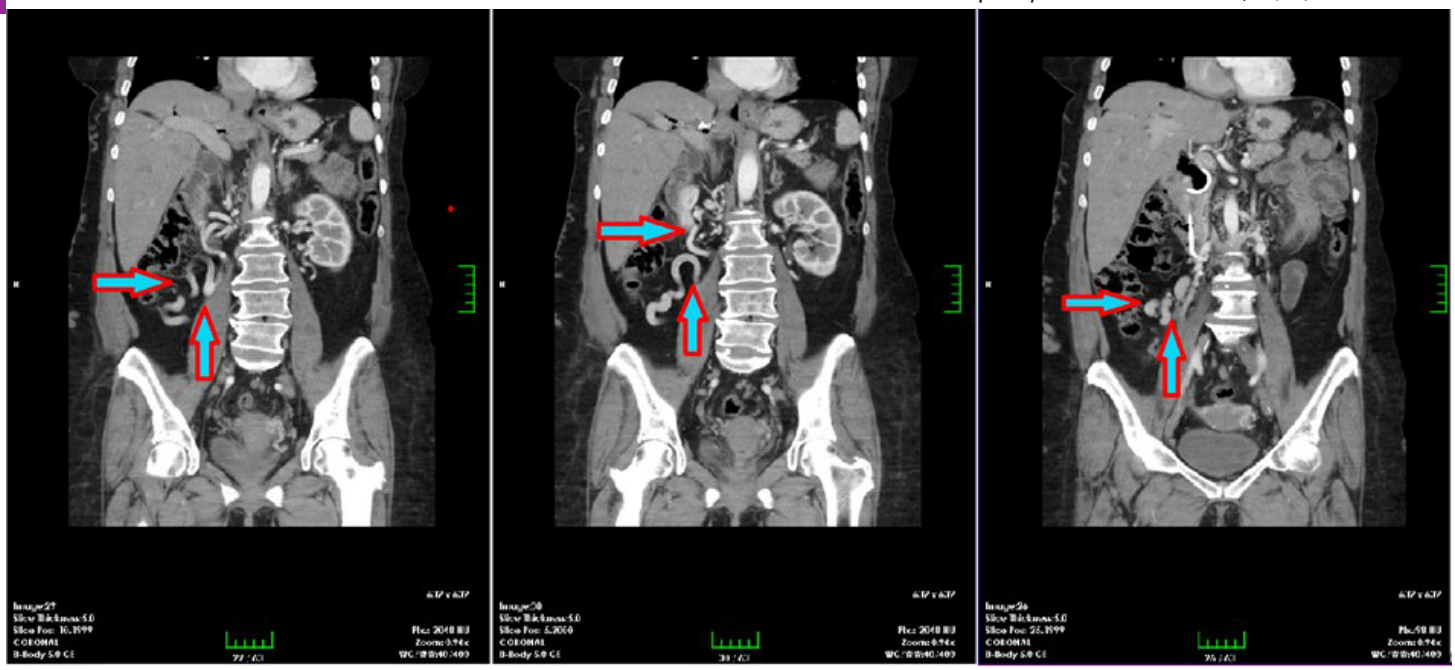

Figure 3: In I.V. contrast-enhanced CT sections taken in the axial plane; Venous vascular structures with ectatic dilatation increasing calibration to provide venous system drainage secondary to infra-renal IVC agenesis/atresia and diffusely tortuous varicose appearance at the levels of SMV and IMV roots are shown with arrows. The superior and inferior mesenteric veins drain into the vena porta via collaterals, then into the hepatic veins and then into the IVC.

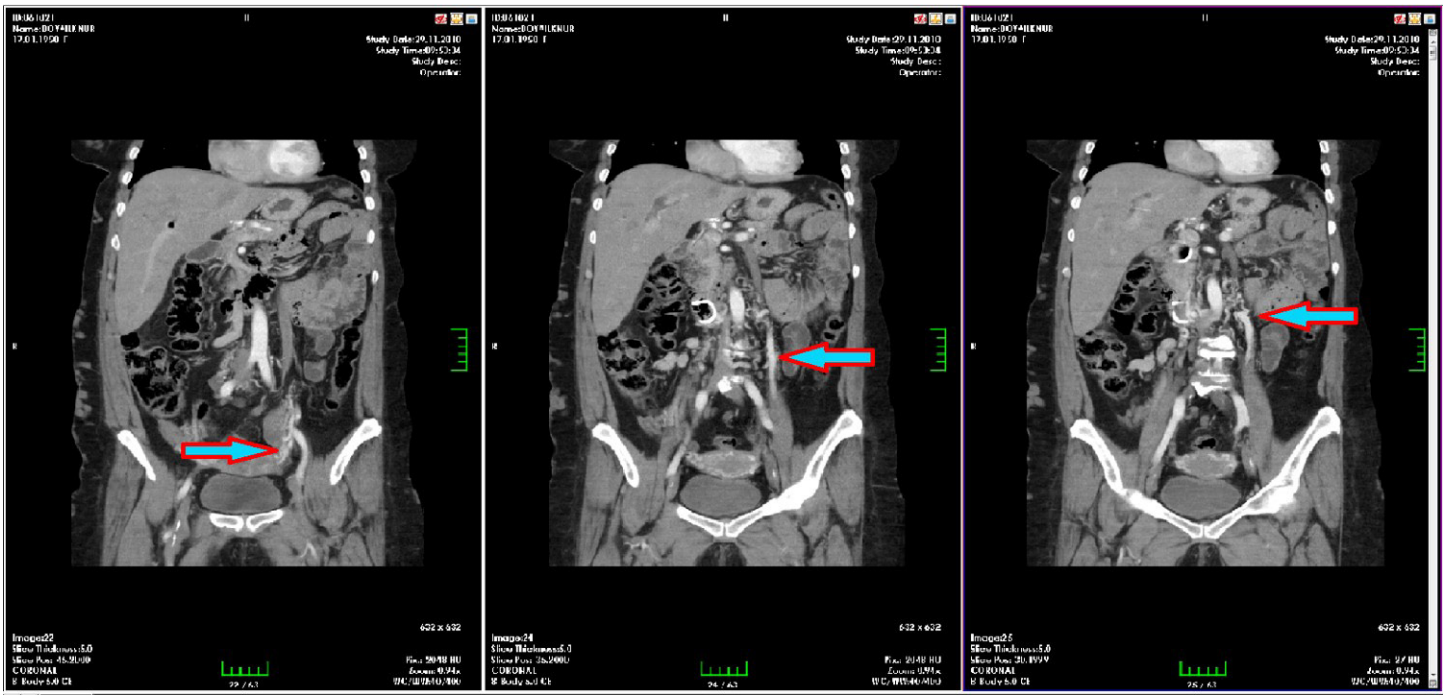

Figure 4: In Contrasted CT Sections; Secondary to infra-renal IVC agenesis, tortuous left ovarian vein, showing ectatic dilatation by increasing calibration to provide venous system drainage, and pouring into the left vena renal.

DISCUSSION

IVC agenesis is a rare anomaly which is usually asymptomatic until advanced ages and it is often incidentally detected during cross-sectional imaging in healthy subjects (4). In IVC agenesis, venous system drainage is usually provided the suprarenal cava, superior vena cava, portal vein, and subclavian veins through the gonadal venous system, paravertebral venous plexus, hemorrhoidal plexus, and the superficial pathway (2). Symptoms may vary according to the localization of these networks (9,10).

Recognition of IVC anomalies is important as they may cause complications related to accompanying anomalies

and possible complications during interventional procedures (7). This anomaly may cause retroperitoneal hemorrhage secondary to large venous aneurysms (10). Occasionally, however, this anomaly is incidentally recognized during abdominal surgery or retroperitoneal surgeries (6) and may affect the method or outcome of surgery by causing technical difficulties (6,11). It can cause major hemorrhages due to vascular injury during the surgical procedure (6,11). In addition, abnormally developing veins in the thoracic area may be mimicked for aortic dissection, lymphadenopathy, and mediastinal tumors and by mistake a percutaneous biopsy may be performed (12). In our case, no preoperative diagnosis was provided. This anomaly was diagnosed via triphasic CT angiography in the venous phase in the postoperative period, due to the suspicion of dense venous collateral formations observed during surgery. In our case, although the surgical procedure was started laparoscopically, it was converted to open surgery due to bleeding. Thus, major bleeding that could be life-threatening was avoided. In such cases, even if it was not diagnosed before the operation, it is essential to manage the process carefully during the operation to avoid an injury that could cause excessive bleeding.

In the literature, transjugular insertion of IVC filter caused difficulties during the process due to IVC anomaly (13). It should be kept in mind that IVC anomalies may cause various complications during the invasive procedure for the treatment of deep venous thrombosis (DVT) and may have predisposing effects for DVT (14,15). In addition, an undiagnosed IVC agenesis can be detected incidentally during IVC cannulation in a patient undergoing open heart surgery and may result in vascular injury (5). Therefore, it is important to diagnose this condition before the cardiopulmonary bypass, which is vital, and before other cardiothoracic procedures (4,5).

Pelvic congestion syndrome is a chronic pelvic pain event caused by increased congestion in the pelvic area due to impaired venous drainage as a result of insufficiency of the pelvic veins (3). A rare cause of this syndrome is IVC agenesis (3,16). IVC agenesis should be considered in patients presenting with pelvic pain. In addition, IVC agenesis can present with venous stasis in the lower limbs and may cause deep vein thrombosis due to increased venous pressure in the lower extremity veins (7,17). IVC anomalies are most commonly presented with DVT in the lower extremities. Congenital IVC anomalies are detected in 5% of patients with lower limb DVT below 30 years of age (17). Especially, in young patients, this anomaly should be taken into consideration during the investigation of the etiology of DVT (18). Although these patients can often present with lower extremity proximal DVT, the risk of pulmonary embolism is very low as the thrombus is difficult to reach the lungs through the extensive compensatory collateral circulation network (2). Besides all these, patients with IVC agenesis may present with symptoms such as paresthesia and neural deficits due to neural compression or obstructive pyelonephritis due to compression to surrounding tissues of highly dilated veins (2).

The CECT scanning and contrast-enhanced MRI in the venous phase provide information on the exact type of IVC anomaly and also describe the extent and compressive effect of deep venous thrombosis and hypertrophic varicose veins. The direct venography via femoral access is a widely used diagnostic test; however, it cannot describe the compressive effects of thrombosed and hypertrophic vessels (3). The treatment of these patients is evaluated individually. In addition to traditional venous insufficiency treatment in patients with signs of venous stasis, anticoagulation therapy can be performed in the treatment to prevent DVT (7, 8). Also, thrombectomy can be performed with invasive procedures in cases with acute DVT. There are cases undergoing endovascular reconstruction and prosthetic graft interposition for interrupted IVC anomaly (19,20).

CONCLUSION

IVC agenesis is a rare entity and clinical suspicion is important in the diagnosis of the disease. Triphasic CT angiography, contrast-enhanced MRI, and direct venography are useful for definitive diagnosis. Besides the use of anticoagulants in the treatment, surgical or endovascular interventions may come into prominence according to the patient's symptoms. The preoperative screening of vascular anomalies and considering IVC anomalies are important to be successful in planned operations and to prevent possible injuries and excessive bleeding. Pre-surgical screening for IVC anomalies can assist in planning safe abdominal and cardiovascular surgery as well as interventional or diagnostic procedures such as IVC filter placement.

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Conflict of Interest

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Ethical Declaration

Informed consent was obtained from the participant and Helsinki Declaration rules were followed to conduct this study.

Authorship Contributions

Concept: KT, Design: KT, Supervising: KT, Data collection

and entry: KT, Analysis and interpretation: KT, Literature search: KT, Writing: KT

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