

Inferior Vena Cava Anomalies: Duplication And Atypical Course

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Abstract

IVC duplication refers to the presence of both a right and left inferior vena cava, explained by abnormal development during embryonic life. This study presents three clinical cases illustrating the diversity of congenital IVC anomalies discovered incidentally during routine CT examinations. Two cases demonstrated classic infrarenal IVC duplication with normal-caliber right IVC and smaller left para-aortic IVC, while the third case presented an atypical retroaortic course. IVC duplication occurs in 0.08 to 3% of the population and may constitute a diagnostic trap, potentially being confused with aortic aneurysm or lymph node enlargement. Clinically, unrecognized anomaly can cause massive hemorrhage during surgery and ineffective pulmonary embolism prophylaxis with single caval filters, necessitating dual filter placement. Recognition of IVC anomalies is crucial due to their significant implications in diagnostic and therapeutic management, particularly in surgical intervention and thromboembolic pathology.

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I. Introduction

The IVC is the main collecting trunk for veins from the subdiaphragmatic portion of the body. Formed by the confluence of the right and left common iliac veins at the L5 level, it is located almost entirely within the abdominal cavity, traverses the diaphragm, and drains into the right atrium after a short thoracic course.

Inferior vena cava duplication refers to the presence of both a right IVC and a left IVC. These malformations are explained by abnormal development and regression of certain segments of the venous system during embryonic life.

II. Case Reports

Clinical Case 1

This involves a 51-year-old male with a medical history of ischemic stroke treated 6 years ago, dyspnea at rest, and chronic lower extremity edema without history of deep vein thrombosis (DVT), presenting with abdominal pain for which he underwent abdominal ultrasonography. This revealed a renal malformation.

A contrast-enhanced abdominal-pelvic CT scan performed in spiral mode without and with intravenous iodinated contrast material was therefore ordered for more precise evaluation:

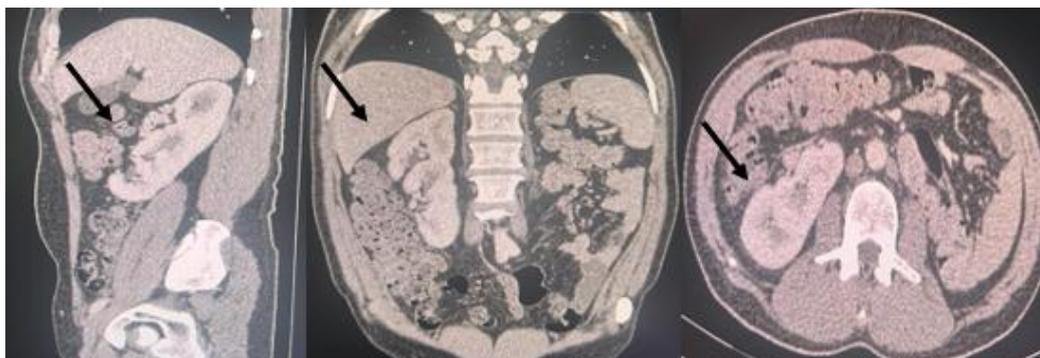


Fig. 1. Contrast-enhanced abdominal-pelvic CT scan with iodinated contrast material in three spatial planes (axial, sagittal, and coronal): sigmoid kidney in the right renal fossa.

The abdominal-pelvic CT scan section (Figure 1) demonstrates a malformed kidney or sigmoid kidney, located in the right renal fossa and extending to the right para-aortic region at the level of the right flank in the

form of two connected kidneys with dysrotation of the inferior portion, containing two renal pelves and two non-dilated ureters without obstruction and no visible lithiasis.

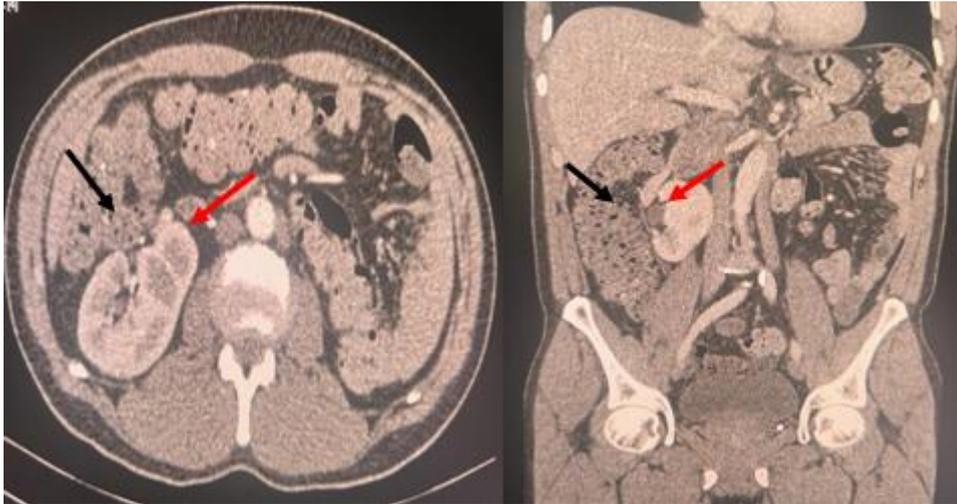


Fig. 2. Contrast-enhanced abdominal-pelvic CT scan in axial plane with coronal reconstruction: inferior vena cava duplication.

The abdominal-pelvic CT scan section (Figure 2) demonstrates the presence of two vena cavae: a main one (black arrow) with normal location and caliber showing low-lying bifurcation (right and left common iliac arteries) at the pelvic level, and a second left para-aortic one (red arrow) with smaller caliber showing no visible division down to the inguinal fold (left external iliac artery).

Clinical Case 2

The second patient, an 83-year-old male with no significant medical history, presented with dysenteric syndrome suggesting intestinal parasitosis. An abdominal CT scan performed in spiral mode without and with intravenous iodinated contrast material was therefore performed for precise etiological diagnosis:

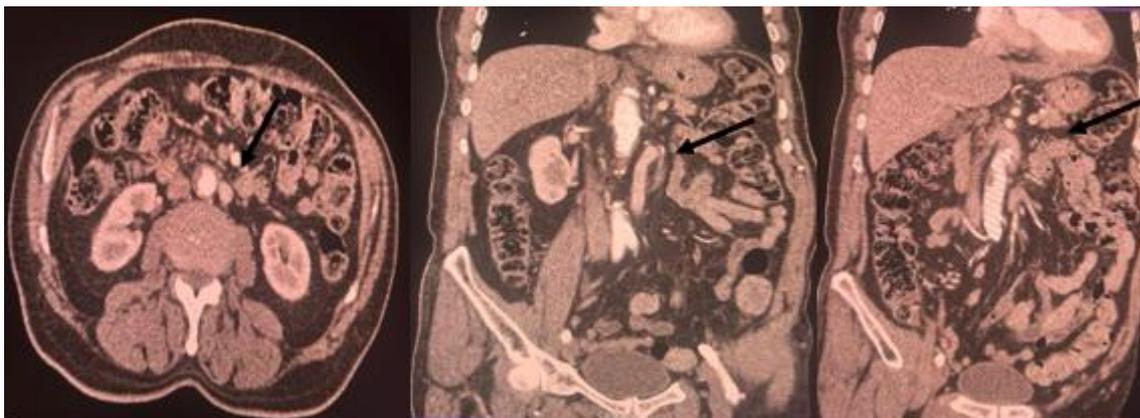


Fig. 3. Contrast-enhanced abdominal-pelvic CT scan in axial plane with coronal reconstructions: inferior vena cava duplication.

The abdominal CT scan section (Figure 3) demonstrates the presence of a vessel suggestive of a second vena cava (black arrow) located in the left para-aortic position, draining into the lumen of the normally positioned IVC at the retrohepatic level.

Clinical Case 3

The third patient, a 78-year-old female with main medical history of Parkinson's disease, hypertension, and cholecystectomy, presented for a follow-up abdominal-pelvic CT scan, performed in spiral mode without and with intravenous iodinated contrast material, which demonstrated:

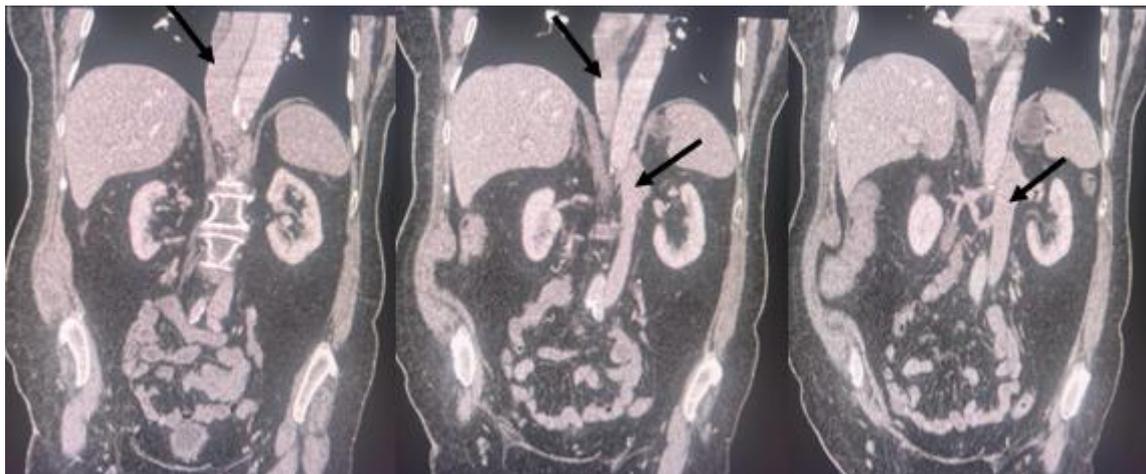


Fig. 4. Contrast-enhanced abdominal-pelvic CT scan with iodinated contrast material in coronal reconstructions: inferior vena cava course variant.

The abdominal-pelvic CT scan section (Figure 4) demonstrates an atypical course of the inferior vena cava (black arrow) which crosses the aorta to become left para-aortic.

III. Discussion

Prevalence and Classification of IVC Anomalies

Our three clinical observations illustrate the diversity of congenital inferior vena cava anomalies encountered in routine radiological practice. IVC duplication, although rare, is one of the most frequent congenital anomalies of this vein, occurring in 0.08 to 3% of the population [1]. Our cases 1 and 2 correspond to classic infrarenal duplications, while case 3 presents a particularly rare retroaortic course variant.

Inferior vena cava duplication is found in approximately 1 to 3% of autopsies [1] and very rarely during cadaveric dissections. While IVC duplication is a well-known anomaly, it usually presents as two venous trunks located on either side of the abdominal aorta [2]. Our observations confirm this classic presentation in cases 1 and 2, with a normal caliber right IVC and a smaller caliber left para-aortic IVC.

The existence of a double inferior vena cava with both channels lying on the right side of the aorta is exceptionally rare, as only two radiological observations of this type have been published [2]. This underscores the importance of recognizing the various forms of IVC duplication in clinical practice.

Embryological Basis

From an embryological perspective, the inferior caval system results from the evolution of three paired and symmetrical venous currents: the posterior cardinal system, the subcardinal system, and the supracardinal system [1]. IVC duplication would be due to the persistence of both right and left supracardinal veins that develop in parallel fashion.

The embryology of the inferior vena cava is complex and still controversial. Nevertheless, it has been accepted since MacClure that the infrarenal segment of the normal inferior vena cava derives from the right supracardinal vein after regression of the infrarenal segment of the subcardinal vein and the ipsilateral posterior cardinal vein [2,4]. The abnormal persistence of these left embryonic structures explains the anatomical variations observed in our cases.

The generally accepted pathogenesis is the persistence of one of the three left venous lines of Reagan, represented by the left supracardinal vein, the left subcardinal vein, and the left posterior or inferior cardinal vein, which normally disappear in the course of events whereas their right homologues take part in the formation of the normal right inferior vena cava [4].

Diagnostic Implications in Imaging

Increasingly, the diagnosis is made preoperatively for various pathologies: thromboembolic, abdominal aortic aneurysm, tumoral, and miscellaneous conditions. Radiological detection methods include ultrasonography, computed tomography examination, and cavography [1]. All three of our observations were discovered incidentally during abdominal CT examinations performed for other indications.

IVC duplication may constitute a trap in sectional abdominal imaging and may be confused with several alternative diagnoses: saccular aortic aneurysm, lumbar para-aortic lymph node enlargement, or left pyeloureteral dilatation [4]. In our first observation, the association with a renal malformation (sigmoid kidney) illustrates the possibility of association with genitourinary malformations, such as renal fusion and retrocaval ureter.

The duplication may not be detected and may constitute a trap in the interpretation of ultrasound or scanographic images, as shown in the literature [4]. In the absence of reference phlebography, the inferior vena caval duplication may not be detected and may be confused with lymph node enlargement or other retroperitoneal structures.

IV. Clinical And Therapeutic Implications

Thromboembolic Risk

In case of unrecognized anomaly, injury to the collateral tributaries of the double IVC can cause massive hemorrhage, and inadvertent ligation of one of the two venous chains can lead to thrombosis with downstream edema and other sequelae of venous hypertension [1].

A double inferior vena cava associated with recurrent unilateral phlebitis requires a specific therapeutic approach [3]. Unilateral recurrence of DVT may lead to searching for a local cause. In such cases, it is essential to evaluate the presence of transverse anastomoses and their patency. The recurrence of unilateral DVT led to searching for a local cause, and the association with IVC duplication and stenosis of the right primitive iliac vein explained the clinical presentation [3].

Pulmonary Embolism Prophylaxis

Placement of a single caval filter may be ineffective for preventing recurrent pulmonary embolism in case of double IVC; it is then necessary to place a filter in each IVC [1]. A filter must be inserted in each inferior vena cava if pulmonary embolism prophylaxis is to be effective [4]. This consideration is crucial for managing patients at high thromboembolic risk.

In view of the infrarenal caval duplication with a risk of migration of clots on both sides, and the need to place the filter immediately below the entry of the renal veins, it may be necessary to insert two filters, one placed conventionally in the right inferior vena cava and the other implanted in the supplementary left vessel [4].

Vascular Investigations

During treatment of left deep vein thrombosis, it is vital to know about this duplication. Therefore, both femoral veins must be systematically punctured during cavography to avoid missing the left IVC and its possible embolic content [1]. Both femoral veins must be punctured routinely for cavography; a single injection route is unacceptable, even if numbered, due to the risk of not detecting the left IVC and its possible thrombotic content [4].

Association with Abdominal Aortic Aneurysm

The association of abdominal aortic aneurysm (AAA) with IVC anomalies can pose problems [5]. Vascular variations can sometimes be the cause of complications during surgical reconstruction of vessels, particularly in retroperitoneal and pelvic vascular surgery [5]. The frequency of AAA is 2% in male subjects over 65 years of age, and in other risk groups, this frequency reaches 10% [5].

When there is suspicion of IVC duplication, one should consider injecting iodinated contrast material through both femoral veins to detect an accessory IVC and possibly to demonstrate the clots it contains [5]. The association of an AAA with IVC variation has been considered a danger in surgery [5].

Surgical Implications

Surgically, in case of IVC duplicity, the problem is twofold: on one hand, exposure of the subrenal aorta because the anastomosis is usually pre-aortic, and on the other hand, recognition of the anomaly [1]. To solve the exposure problem during abdominal aortic intervention, three possibilities are available: ligating the inter-iliac anastomosis, carefully mobilizing these IVCs before clamping, or limiting aortic dissection to the anterior and lateral faces without approaching the posterior face.

During laparotomy performed for treatment of an abdominal aortic aneurysm, in addition to the pre-aortic plexuses and the right gonadal vein, an accessory IVC must be sought. Duplications of the renal veins may be the cause of injury during aortic dissection in the course of aneurysm repair [5].

Retroperitoneal surgery, whether involving interventions on the abdominal aorta, lumbar spine, kidneys, or adrenals, must take into account the existence of this anomaly like any variation of retroperitoneal veins, or risk exposure to obvious perioperative or postoperative complications [2].

Practical Recommendations

The evolution of imaging allows preoperative diagnosis of inferior vena cava anomalies. The surgeon must have the rudiments to solve potential problems of anomalies of this vein, particularly in abdominal aortic surgery, nephrectomies, adrenalectomies, and sympathectomies [1].

A routine search for renal anomalies is essential, either by taking a delayed pyelographic film in the late stages of each cavography or by ultrasound exploration [4]. This recommendation takes on full meaning in our first observation where the IVC anomaly was associated with a complex renal malformation.

The relative frequency of duplication of the inferior vena cava calls for perfect familiarity with the different imaging appearances of this anomaly. In view of the practical and therapeutic implications of this anomaly, it must always be included in the range of possible diagnoses when interpreting a channeled or nodular structure situated to the left of or in front of the abdominal aorta and interrenal or infrarenal in position [4].

V. Conclusion

Our three observations illustrate the diversity of inferior vena cava anomalies and emphasize the importance of their recognition in routine radiological practice. The relative frequency of inferior vena cava duplication requires perfect familiarity with the different imaging aspects of this anomaly. Although often asymptomatic and discovered incidentally, these anatomical variations can have significant implications in the diagnostic and therapeutic management of patients, particularly in cases of surgical intervention or thromboembolic pathology. Recognition of inferior vena caval duplication not only raises diagnostic problems but may influence therapeutic programs, especially in the context of abdominal aortic aneurysm repair and thromboembolic disease management.

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