

Two cases of inferior vena cava duplication with their CT findings and a review of the literature

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Duplication of the inferior vena cava (IVC) is the most common anomaly to affect the vena cava. Variations in the IVC are diagnosed in routine dissection studies, in retroperitoneal surgeries, or in computerised tomography (CT) sections ordered for various reasons. In this paper we present two cases of a double IVC together with the CT findings. The duplication might have occurred during embryological development. Although venous anomalies are rare, they have particular importance with respect to the interruptions that may occur during retroperitoneal and thoracic surgery or in the treatment of thrombo-embolic disease.

Key words: inferior vena cava, duplication, anomaly, variation, computed tomography

INTRODUCTION

Venous anomalies are frequently observed and result mainly from errors in embryological development. Venous anomalies of the retroperitoneal region have been described extensively by anatomists and radiologists. However, such anomalies should also be taken into consideration by surgeons operating in this region.

The development of the IVC is a complex and continually evolving process beginning from the 4th week after conception and lasting through the embryonic period until about the 8th week. There is a vast network of communicating veins, the most important of which are 3 pairs of parallel veins. The postcardinal veins appear first and eventually regress to form the iliac bifurcation. The subcardinal veins appear next, the right forming the suprarenal IVC while the left subcardinal vein regresses. The supracardinal veins are the last to appear. The right persists to form the infrarenal IVC, while the left portion completely regresses in foetal life. Anasto-

mosis of the subcardinal and supracardinal veins contributes to the formation of the renal veins.

Duplication of the IVC is the most commonly observed anomaly and is estimated to occur in 2% to 3% of the population [1].

CASE REPORTS

We detected two cases of double IVC on CT images during routine radiological examination. Computed tomography was performed by Philips AV-E1 spiral CT (Rotterdam, Netherlands). In our first case a 66-year-old man presented with long-term lumbar pain and both IVC were formed from the respective common iliac veins. Both of the IVC ran upwards bilaterally, lateral to the abdominal aorta as far as the level of the left renal vein (Fig. 1A). At the level of the left renal vein the left IVC joined to form a common trunk with the left renal vein and crossed the midline (Fig. 1B). With the entrance of the right IVC to this common trunk, the IVC ran upwards as a single vein (Fig. 1C) and left the abdomen in its normal

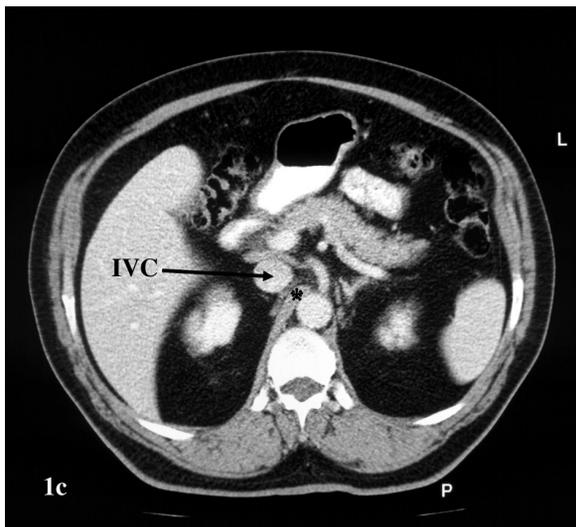
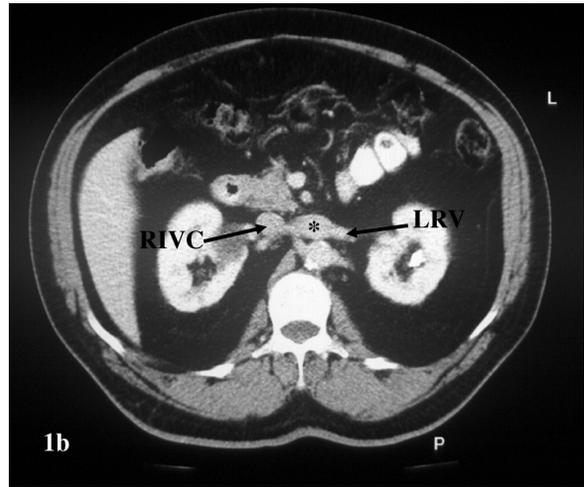
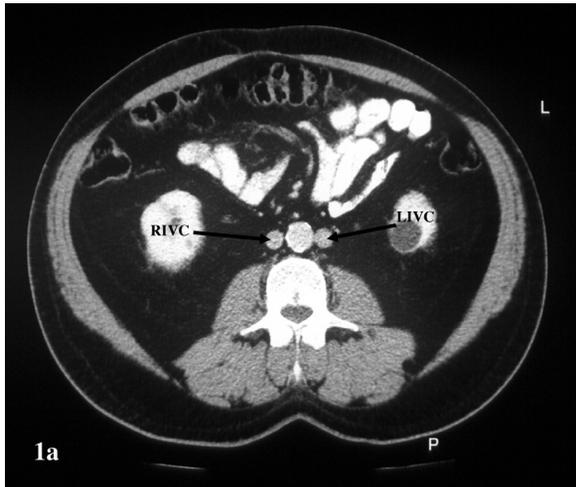


Figure 1 A. In the first case the right IVC (RIVC) and left IVC (LIVC) both ran upwards bilaterally, lateral to the abdominal aorta as far as the level of the left renal vein; **B.** At the level of the left renal vein (LRV) the LIVC joined to form a common trunk (*) with the left renal vein and crossed the midline. The RIVC was slightly larger than the left; **C.** With the entrance of the RIVC to this common trunk the IVC ran upwards as a single vein and left the abdomen in its normal anatomical position at the level of the aortic hiatus (*) indicates the right crus of the diaphragm.

anatomical position. The right IVC was slightly larger than the left. We also observed an atheromatous aorta and multiple lesions which disrupted both kidney borders. In our second case a 55-year-old woman presented with abdominal pain of one week's duration. The CT images obtained revealed a double IVC formation which proceeded in the same way as the previous case (Fig. 2). There were also focal lesions at the right kidney.

DISCUSSION

Formation of the IVC involves the subsequent appearance and regression of 3 paired veins: the posterior cardinal veins, the subcardinal veins and the supracardinal veins. The cardinal veins constitute the main venous drainage system of the embryo. The posterior cardinal veins develop primarily as the vessels of the mesonephroi and largely disappear with these transitory kidneys. The only adult derivatives of the posterior cardinal veins are the root of the azygos vein and

the common iliac veins. In the 5th week, the subcardinal veins appear. These are connected with each oth-



Figure 2. In the second case both of the IVC ran upwards bilaterally, lateral to the abdominal aorta as in the first case.

er through the subcardinal anastomosis and with the posterior cardinal veins through the mesonephric sinusoids. The subcardinal veins form the stem of the left renal vein, the suprarenal (adrenal) veins, the gonadal veins and the prerenal segment of the IVC. The supracardinal veins are the last pair of vessels to develop. They are united by an anastomosis that is represented in the adult by the azygos and the hemiazygos veins. Caudal to the kidneys the left supracardinal vein degenerates but the right becomes the IVC. The final development of the "normal" right-sided IVC is a reflection of the persistence of the right subcardinal veins as the prerenal segment, the subsupracardinal vein (anastomosis between the subcardinal and supracardinal veins) as the renal segment, the right supracardinal vein as the postrenal segment and the posterior cardinals as the iliac bifurcation and iliac veins [1, 2, 11, 13, 14].

The major anomalies of the IVC, most frequently encountered are:

1. Duplication of the IVC [3, 5, 6, 9, 12, 15, 18, 19, 22, 24, 26, 28].
2. Transposition of the IVC (left IVC).
3. Circumaortic (left) renal vein.
4. Retro-aortic (left) renal vein.
5. Absence of the hepatic portion of the IVC [1, 14].
6. Right double IVC with azygos continuation of the posterior-medial vein [13].
7. Double IVC with hemiazygos and azygos continuation of the left vena cava [13].
8. Azygos continuation of the vena cava inferior [2].
9. Double IVC with recurrent right renal vein and hemiazygos continuation of the IVC [2].
10. Double IVC with retro-aortic left renal vein and azygos continuation of the IVC [2].
11. Absent infrarenal IVC with preservation of the suprarenal segment [2].
12. Duplication anomaly of the IVC with normal drainage of the right IVC and hemiazygos continuation of the left IVC [4].
13. Double superior vena cava with double IVC [24].
14. Double right vena cava forming a peri-ureteric venous ring [10, 16, 20].

Of these anomalies duplication of the IVC constitutes the majority, with a prevalence of 2–3% [1]. The double IVC in the case presented here is considered to be a persistence of both the right and left supracardinal veins.

Associated anomalies reported with duplication of the abdominal vena cava include:

- cloacal extrophy;
- congenital absence of the right kidney;

- renal ectopia with abdominal aortic aneurysm [23];
- right retrocaval ureter [10];
- left retrocaval ureter and congenital absence of the iliac anastomosis [1, 15, 20];
- anomalous drainage from the left arm [19];
- congenital heart disease: the association of ostium primum, ostium secundum and double vena cava inferior with abnormal left atrium drainage [27];
- transcaval ureter [7].

The formation of the venous drainage system of the human body is a complex process, since it involves succeeding structures forming and disappearing in a predefined order. Interruption of one of these steps results in the formation of congenital anomalies. Further understanding of the molecular mechanisms through analysis of how the cells of the developing human proliferate, migrate and self-destruct may in the future help us predict the anomalies in an individual.

The most important clinical consequences of the duplication of the vena cava are observed in retroperitoneal surgery [1, 23]. In order to reduce the hazards of significant venous haemorrhage, the surgeon should be aware of the embryogenesis and to be able to recognise and identify the major venous anomalies in this area [23]. Another surgical implication can be observed during whole organ transplantation or radical nephrectomy [1]. Moreover, during the staging of testicular tumours the radiologist should be aware of the possible venous anomalies in order to avoid possibly misleading reports such as para-aortical lymphadenopathy [8].

In addition, recognition of a double IVC is important when surgical ligation of the IVC or the placement of an IVC filter is to be performed for thromboembolic disease [21]. Failure to diagnose the double IVC may lead to recurrent embolism [23].

Major venous anomalies such as double IVC also lead to a further problem in lymph node dissection as the lymphatic drainage generally tends to follow the vascular pattern. Patients with an abnormal venous anatomy have unusual patterns of lymphatic drainage and lymph node metastases [6, 23].

An understanding of the anatomical variations of the IVC is, moreover, especially significant in the cross-sectional images, radionuclide venography or catheterisation and opacification of the IVC. For instance, a thrombosed double IVC may be misinterpreted as a paravertebral lymphadenopathy or a retroperitoneal mass [6].

There may also be misinterpretations of the anomalous venous drainage as mediastinal mass or paraver-

tebral lymphadenopathy. Thus, when an anomaly of the IVC is suspected as a result of a radiological finding on abdominal CT or ultrasound, it is necessary to perform an inferior venogram or MRI study in order to trace precisely the vena caval system [3, 23, 26].

Furthermore, the presence of a double IVC will dilute the left renal vein sampling for renin in the hypertensive patient because of the blood flow carried by the left vena cava [1].

When the venous return occurs in alternative pathways, the aberrant vessel may simulate a mediastinal mass or it may mimic an aortic dissection. More importantly, peri-operative death has been observed following inadvertent ligation of the hemiazygos-to-azygos continuation of a left IVC [2].

Finally, duplication of the IVC has been discussed with regard to the physiological asymmetries of the venous system. The embryological implications have been compared to the finding of genes that control the asymmetry of visceral organs [16].

In conclusion, although venous anomalies are rare, they have particular importance for the various interruptions that may take place during retroperitoneal and thoracic surgeries or in the treatment of thrombo-embolic disease. Last but not least, radiologists should be aware of the venous anomalies so that they are not interpreted as pathological findings.

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