

Anomalies of the inferior vena cava: a report of two cases and a short review of the literature

S. Kapetanakis¹, C. Papadopoulos¹, P. Galani², G. Dimitrakopoulou², A. Fiska¹

¹Department of Anatomy, Medical School of Alexandroupolis, Greece

²Department of Radiology, General Hospital Amalia Fleming, Athens, Greece

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The inferior vena cava, also known as the posterior vena cava, is the large vein that carries de-oxygenated blood from the lower half of the body into the right atrium of the heart. Congenital anomalies of the inferior vena cava result from the persistence of the embryonic venous system. The majority of cases are clinically silent and are diagnosed in routine dissection studies, in retroperitoneal surgeries, or through imaging for other reasons. Although these anomalies are rare, they are of great importance during operations in the abdominal area or in the treatment of thromboembolic diseases. We report two cases of double vena cava and left vena cava, respectively, and a short review of the relevant literature. (Folia Morphol 2010; 69, 3: 123–127)

Key words: inferior vena cava, anomalies, double, left

INTRODUCTION

Anomalies of the inferior vena cava are rare and result mainly from abnormal embryological development. Among these, the most common are double inferior vena cava (IVC), with an incidence of 0.2–3% [2, 11], and left sided IVC with an incidence of 0.69% [20]. They are mostly incidentally diagnosed on imaging of the area, usually with computed tomography (CT scanning) or magnetic resonance imaging (MRI) done for other reasons. However, it is important to recognise these anomalies as they can have significant clinical implications, especially for surgeons and for the treatment of thromboembolic diseases.

In this particular study we present two rare cases. In the first case we report an 80 year-old man with acute renal failure and urine infection, in whom a double vena cava was noted in routine CT imaging of the area. In the second case we report a 66 year-old man with acute pain in the abdomen,

in whom CT imaging showed a left-sided IVC. Finally, we discuss the clinical importance of recognising the anomalies of the IVC and we review the relevant literature.

CASE 1

A case of double IVC on both sides of the aorta was observed during routine imaging with CT scanning (Fig. 1) in an 80 year-old man with past medical history of ischaemic heart disease, atrial fibrillation, and cancer of the urine bladder which appeared 4 months previously. The patient presented symptoms of urine infection, haematuria, and total oedema of the body. From the laboratory tests we found deterioration of the renal function/acute renal failure and low levels of albumin in the blood. There were positive urine cultures for *Klebsiella pneumoniae* $> 10^5$ and we started medication with amoxicillin and garamycin. We proceeded with CT scanning of the lower abdomen, due to the poor images of



Figure 1. Double inferior vena cava, bilateral of the aorta.

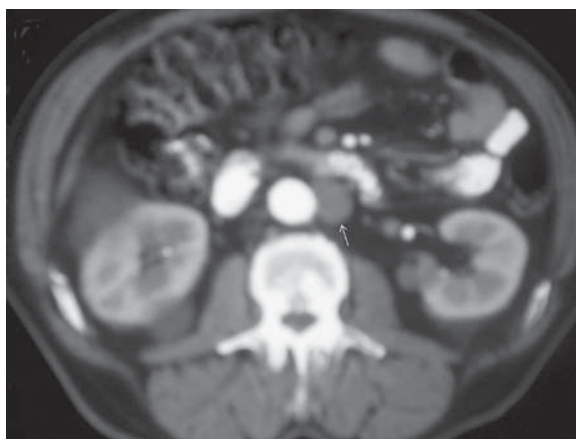


Figure 3. Left-sided inferior vena cava.

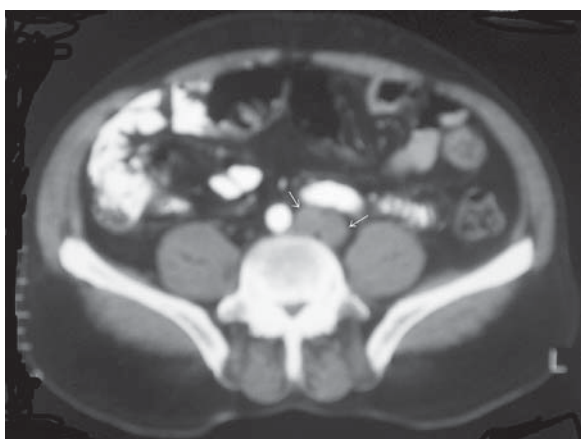


Figure 2. Bifurcation of the left-sided inferior vena cava to the common iliac veins.

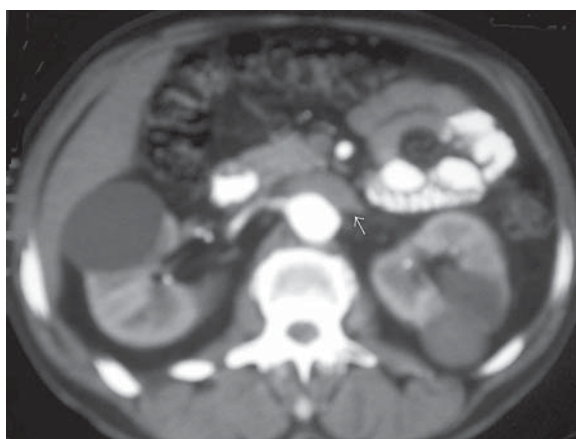


Figure 4. The left-sided inferior vena cava crosses anteriorly to the aorta at the level of the renal arteries.

the ultrasonography, to examine the kidneys and the urine bladder. During the imaging we incidentally observed a double IVC, which was on both sides of the aorta. We were not able to carry out further examination of the upper abdomen to see where these two venae cavae joined and formed one main IVC because the patient presented with a high fever and died five days later.

CASE 2

A 66 year-old, previously healthy man presented to our hospital with acute pain in the abdomen. All laboratory tests and ultrasonography of the area were negative for pathologic findings. Nevertheless, we proceeded with CT imaging of the abdomen because of the persistence of the symptoms. This examination was also normal, so we pinned the symptoms on a possible colic of the large intestine.

Apart from other findings, the CT imaging incidentally showed two common iliac veins that joined at the left side of the abdominal aorta (Fig. 2) and formed a left sided IVC (Fig. 3) that crossed anteriorly to the right side at the level of the renal arteries (Fig. 4). The patient took the appropriate medication with analgesics and went home with dietary instructions.

DISCUSSION

The IVC is formed by the right and left common iliac veins on the right anterior surface of the fifth lumbar vertebra, 2.5 cm to the right of the median plane, inferior to the bifurcation of the abdominal aorta, and brings blood to the right atrium of the heart from all parts of the body below the diaphragm [10]. The IVC is right-sided in nearly 98% of the population [15]. The abdominal part of the IVC

is 20 cm long and ascends on the right side of the bodies of the lumbar vertebrae and to the right of the aorta. It enters the thorax through the caval foramina in the central tendon of the diaphragm, which is at the level of the eighth thoracic vertebra. Unlike the superior vena cava, it has a substantial number of tributaries. It also anastomoses with the azygos vein system (which runs on the right side of the vertebral column). The tributaries include the veins that collect blood from the muscles and coverings of the loins, and from the walls of the abdomen, the reproductive organs, the kidneys, and the liver. The specific levels of the tributaries are as follows: the common iliac vein is at L5, lumbar veins are at the level between L1–L5, the gonadal vein is at L2, the renal and suprarenal veins are at L1, and finally the hepatic and inferior phrenic veins are at T8. Because the IVC is not centrally located, there are some asymmetries. The testicular (gonadal) veins and suprarenal veins drain into the IVC on the right side but into the renal vein on the left side, which in turn drains into the IVC. By contrast, all the lumbar veins and hepatic veins usually drain directly into the IVC.

The IVC develops in a complex process which begins on the fourth week of gestation and is completed approximately on the eighth week [21]. It involves the development, regression, anastomosis, and replacement of three pairs of embryonic veins (posterior cardinal, subcardinal, and supracardinal veins) [4]. The posterior cardinal veins appear first and regress, except for the distal aspects which become the iliac bifurcation. The subcardinal veins appear on the fifth week. They form the stem of the left renal vein, the suprarenal (adrenal) veins, and the prerenal segment of the IVC. The left subcardinal vein completely regresses. Subsequently, the supracardinal veins develop. The left regresses and the right forms the infrarenal IVC. The persistence of the left supracardinal vein is the cause of most of the anomalies of the IVC.

In this study we report two cases of anomalies of the IVC. In the first case a double vena cava was incidentally observed during imaging for other reasons. The IVC was situated bilaterally to the abdominal aorta and, according to the embryology, this resulted from the failure of regression of the left supracardinal vein. The occurrence of this variation of the IVC is about 0.2–3% in the population [11]. The duplicated left IVC usually drains into the left renal vein that crosses anterior to the aorta and joins the IVC. Unfortunately, we could not run further

imaging on our patient because of his bad medical condition and his eventual death. Thus, we can only assume the course of the duplicated IVC of our patient.

In the second case, a 66 year-old man presented abdominal pain, and the imaging of the area with CT scanning showed a left-sided IVC. It is one of the most common variations of the IVC with a prevalence of 0.69% [20]. A left sided IVC usually ascends to the left side of the abdominal aorta that crosses anteriorly to the right side at the level of the renal arteries [9].

Except for the anomalies described above, there are many other mentioned. The four most common anomalies that also have clinical importance are the double IVC (0.2–3%) [11, 15], the isolated left-sided IVC (0.2–0.5%) [11, 15], the circumaortic left renal vein (1.5–8.7%), and the retro-aortic left renal vein (2.1%) [11]. The first two variations may be revealed by imaging with ultrasonography of the abdomen [9]. All other rare anomalies are only observed on CT imaging [9] of the area or in dissection studies and retroperitoneal surgeries. Rare anomalies of the IVC include the absence of a part of the IVC, azygos, and hemiazygos continuation of a duplicated IVC, double superior and double IVC, hypoplasia, or agenesis and interruption of the IVC [21]. In particular, all anomalies and anatomic variations are described in Table 1 where we report the relation between nationality and these anomalies. We should also note that there is a similar incidence of the anomalies of the IVC in Asiatic populations (mostly in India, China, and Japan) and in European and other Western countries. Anomalies of the IVC are observed mostly in males at a percentage of 62%. Additionally, we came to the conclusion that interrupted IVC is observed mostly in women, in a high percentage, while aplasia/agenesis of the IVC is observed in males and particularly in European and other Western countries. This is a fact that is probably due to gene mutations that appear in these populations. Further research and understanding of the molecular mechanisms may help us predict such anomalies in future in individuals and avoid many of the mistakes made during operations in the abdomen by surgeons who are unaware of the anatomic variations of the area.

The majority of cases of anomalies of the IVC are diagnosed incidentally during imaging or surgeries. Its occurrence is rare but has important clinical consequences. During retroperitoneal surgeries [1, 16] it is very useful to know the anatomy of the

Table 1. Major anomalies of the inferior vena cava (IVC) and the relation with the populations in Asiatic and Western countries from a short review of literature

A/A Anomalies	Populations	No. of cases
1 Double IVC	23 Asiatic, 26 Western	49 cases
2 Transposition (left IVC)	8 Asiatic, 11 Western	19 cases
3 Circumaortic renal vein	1 Asiatic	1 case
4 Retro-aortic (left) renal vein	1 Asiatic, 1 Western	2 cases
5 Absence of the hepatic portion of the IVC	1 Western	1 case
6 Right double IVC with azygos continuation of the posterior medial vein	1 Western	1 case
7 Double IVC with hemiazygos and azygos continuation of the left VC	1 Western	1 case
8 Azygos continuation of the IVC	5 Western	5 cases
9 Double IVC with recurrent right renal vein and hemiazygos continuation of the IVC	1 Western	1 case
10 Double IVC with retro-aortic left renal vein and azygos continuation of the IVC	1 Western	1 case
11 Absent infrarenal IVC with preservation of the suprarenal segment	1 Asiatic, 4 Western	5 cases
12 Duplication anomaly of the IVC with normal drainage of the right IVC and hemiazygos continuation of the left	1 Western	1 case
13 Double superior and double IVC	1 Asiatic, 1 Western	2 cases
14 Double right VC forming a periureteric venous ring	1 Asian, 2 Western	3 cases
15 Agenesis/aplasia of the IVC	9 Asiatic, 21 Western	30 cases
16 Interrupted IVC	5 Asiatic, 9 Western	14 cases
17 Double IVC with circum-aortic renal vein	1 Asiatic	1 case
18 Interrupted IVC with azygos continuation	5 Asiatic, 3 Western	8 cases
19 Hypoplasia of the IVC	1 Asiatic, 3 Western	4 cases
20 Anterior IVC	1 Western	1 case

area and recognise the venous anomalies so as to reduce the hazards of significant venous haemorrhage. Additionally, the surgeon must be aware of such variations during organ transplantation [12], radical nephrectomy [1], sympathectomy, or ureteral surgery [3, 17]. It could be fatal to mistakenly injure the IVC during an operation of the abdomen. Another implication of recognising the anomalies of the IVC is in patients with thromboembolic diseases in whom it is planned to place a filter in the vena cava. If there is an unrecognised double IVC, there may be reoccurrence of embolisms by thrombus from the left IVC [13, 14]. A higher occurrence of thromboembolic disease [7] and pulmonary embolism has also been observed in patients with anomalies of the IVC [9]. Radiologically, the presence of IVC anomalies such as a double IVC can be mistaken for lymphadenopathy [6, 8, 11] and metastatic disease of the lymph nodes in cases of testicular carcinoma [18] or as a retroperitoneal mass [5, 16]. Thus,

when an anomaly is suspected on CT or MRI, it is necessary to perform an inferior venogram in order to outline precisely the vena cava system [3, 19].

CONCLUSIONS

In conclusion, although the anatomical variations of the IVC are rare, they should be recognised by radiologists and surgeons in order to avoid mistakes during imaging of the area or surgeries and in cases with venous thromboembolic disease. Consequently, these anomalies would not be interpreted as pathological findings and the best treatment would be possible for each patient.

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