Anomalies of the inferior vena cava and renal veins and implications for renal surgery

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KEY WORDS

inferior vena cava ◗ renal veins ◗ venous anomalies

ABSTRACT

Abnormalities of the inferior vena cava (IVC) and renal veins are extremely rare. However, with the increasing use of computed tomography (CT), these anomalies are more frequently diagnosed. The majority of venous anomalies are asymptomatic and they include left sided IVC, duplicated IVC, absent IVC as well as retro-aortic and circumaortic renal veins. The embryological development of the IVC is complex and involves the development and regression of three sets of paired veins. During renal surgery, undiagnosed venous anomalies may lead to major complications. There may be significant hemorrhage or damage to vascular structures. In addition, aberrant vessels may be mistaken for lymphadenopathy and may be biopsied. In this review we discuss the embryology of the IVC and the possible anomalies of IVC and its tributaries paying particular attention to diagnosis and implications for renal surgery.

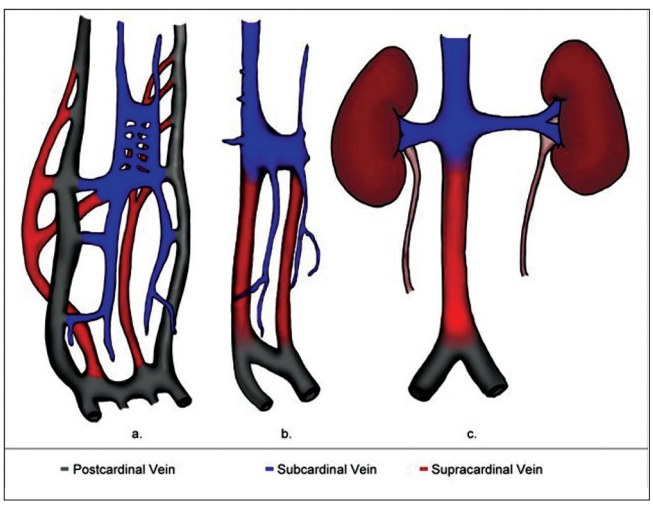

Fig. 1a. Subcardinal and supracardinal veins. Figure 1a demonstrates the three sets of paired veins that develop in the embryo, namely the subcardinal (blue), supracardinal (red) and postcardinal veins (grey). **1b.** The right subcardinal vein becomes dominant. Figure 1b demonstrates regression of the postcardinal veins; the supracardinal veins become dominant infrarenally, while the subcardinal veins become dominant above the renal veins. **1c.** Final structure of the IVC. Figure 1c demonstrates the final composition of the inferior vena cava.

INTRODUCTION

The first reported inferior vena cava (IVC) anomaly was in 1793 by Abernethy, when he described a congenital mesocaval shunt and azygos continuation of the IVC in a 10-month old infant with dextrocardia [1]. The most accepted classification of IVC anomalies is based on the embryonic veins that the anomaly is derived from [2]. IVC anomalies are uncommon and are usually discovered incidentally during cross sectional imaging in otherwise healthy individuals. The IVC develops between the 6th and 8th weeks of intrauterine life by a complex process involving the development and regression of 3 sets of paired cardinal veins [3].

Undiagnosed IVC anomalies may lead to significant complications during renal surgery. Fortunately, the widespread use of computed tomography (CT) scans with 3-dimensional reconstructions during preoperative planning enables identification of the aberrant anatomy prior to surgery. Hence the risk of inadvertent damage to anomalous venous structures with resultant hemorrhage may be avoided [4].

We aim to review the embryology of the IVC, reported anomalies and their identification and implications for renal surgery.

Embryology

The IVC and its tributaries start to develop by the 6th week of fetal life. Development is completed by the 8th week. Three pairs of cardinal veins contribute to the IVC and its tributaries and they are illustrated in figure 1a [3].

The first veins to develop are the postcardinal veins, which drain the caudal half of the body. The postcardinal veins drain into the common cardinal veins, which drain into the *sinus venosus*. Medial to the postcardinal veins, the subcardinal veins start to develop. Multiple anastomoses form between the subcardinal veins as well as between the subcardinal and postcardinal veins on each side. Gradually the venous drainage of the caudal part of the body is shifted to the subcardinal veins and the postcardinal veins start to regress. The supracardinal veins gradually develop to take over the venous drainage of the caudal body and the subcardinal veins start to degenerate. Anastomoses develop between the supracardinal veins and postcardinal veins on each side and the right subcardinal vein becomes dominant as illustrated in figure 1b [4, 5].

Figure 1c displays the final structure of the IVC. The caudal ends of the postcardinal vein persist and form the common iliac veins. The cranial end of the right subcardinal vein forms the

Table 1. Classification of IVC anomalies

Anomalies of the postcardinal veins	Retrocaval/circumcaval ureter
Anomalies of the subcardinal veins	Interruption of the inferior vena cava with azygous/hemiazygous continuation
Anomalies of the supracardinal veins	Persistence of the left supracardinal vein – Left inferior vena cava
	Persistence of both left and right supracardinal veins – Double inferior vena cava
Anomalies of the renal segment	Circumaortic venous ring
	Retroaortic renal vein
	Multiple renal veins

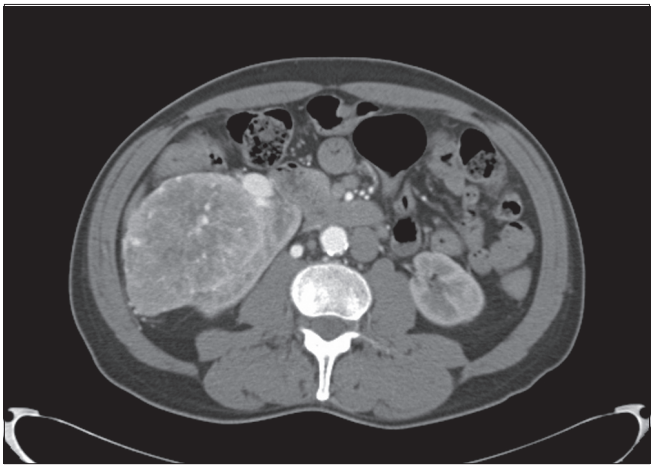

Fig. 2a. Transverse CT image of the left IVC.

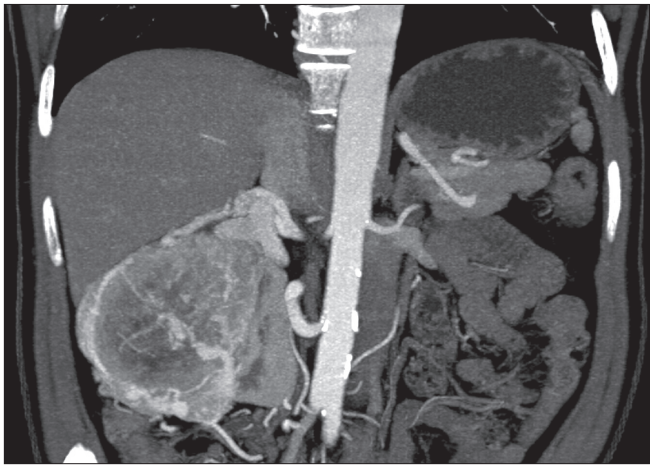

Fig. 2b. Coronal reconstruction of a CT image of left IVC.

hepatic segment of the IVC. The cranial part of the right subcardinal vein forms the suprarenal part of the IVC. The cranial part of the left subcardinal vein forms the left adrenal vein. The caudal segments of the subcardinal veins form the gonadal veins. The right supracardinal vein forms the infrarenal segment of the IVC. The cranial part of the right supracardinal veins forms the azygos vein. The anastomosis between the supracardinal and subcardinal veins forms the dorsal arch of the aortic collar, while the intersubcardinal anastomosis forms the ventral arch. From the aortic collar the renal veins develop (ventral and dorsal); the dorsal vein regresses and the ventral vein remains as the renal vein [4, 5].

Classification

The most accepted classification of IVC anomalies is based on the embryonic vein that the anomaly is derived from [2]. The various possible anomalies are listed in Table 1 and will be discussed in turn. Anomalies of the supracardinal veins and the renal segment are most relevant to urologists.

Anomalies of the supracardinal veins

Left IVC

The infrarenal part of the IVC develops from the supracardinal vein. Regression of the right supracardinal vein and persistence of the left supracardinal vein lead to the development of the infrarenal IVC on the left side and the suprarenal IVC on the right side [6]. The IVC crosses to the right side at the level of the renal veins anterior to the aorta. Crossing of the IVC posterior to the aorta has also been reported [5]. The incidence of left IVC is 0.2–0.5% [7]. In the presence of a left IVC, the normal venous anatomy may be reversed. The left gonadal and adrenal veins drain directly into the left IVC while the right gonadal and adrenal veins drain into the right renal vein, which is then joined by the left IVC as it crosses the midline to lie on the right side at the level of the renal veins. The anomaly is usually silent and is found on preoperative imaging. The identification of a left IVC is critical prior to vascular procedures, particularly nephrectomy or adrenalectomy. In addition, a left IVC has been associated with other anomalies such as multiple renal veins and care must be taken during hilar dissection [8]. The risk of vascular injury is even higher if not identified prior to surgery or when the right renal vein crosses posterior to the aorta [9]. A left IVC can also be misdiagnosed as lymphadenopathy and cases exist in the literature where lymph node dissection has been attempted or chemotherapy administered [10]. Figures 2a and 2b display transverse and coronal CT images of a patient with left IVC.

Double IVC

The persistence of both the right and left supracardinal veins leads to the development of a double IVC up to the level of the left renal vein. The duplicated left IVC joins the left renal vein, which in turn runs to the right to drain into the right IVC. The incidence of double IVC is 0.2-3.0% [4]. The condition is asymptomatic and usually diagnosed incidentally following abdominal imaging. Double IVC can be misdiagnosed as lymphadenopathy especially in patients being evaluated for renal surgery or neoplasm. Patients with double IVC are at risk of vascular injury during retroperitoneal procedures therefore identification prior to surgery is essential to avoid such complications. Some reports suggest that patients with double IVC have a higher risk of developing thromboembolic events [11-13]. Recurrent pulmonary embolism after the insertion of an IVC filter should also raise the suspicion of a double IVC. Figures 3a and 3b display coronal and axial CT images of a double IVC.

Absent infrarenal IVC

Absence of the infrarenal segment of the IVC is a very rare anomaly. The etiopathogenesis of absent infrarenal IVC is controversial. Embryonic maldevelopment may be caused by maldevelopment of the right supracardinal vein, which results in preservation of the suprarenal IVC and absence of the infrarenal part [14]. The controversy is whether an infrarenal IVC is acquired or a true congenital anomaly. Thrombosis and fibrosis of the IVC is a possibility [14, 15]. Wax et al. reported a case of a patient with absent infrarenal IVC who in earlier life had a normal venous system [14]. Venous return is carried from the external and internal iliac veins to the lumbar veins, which communicate with the azygos and hemiazygos system of veins. Patients with an absent infrarenal IVC may present with symptoms of lower extremity venous insufficiency and venous thrombosis [4]. Dilated collateral veins can also be mistaken for paraspinal masses in which percutaneous biopsy would have disastrous consequences [16].

Anomalies of the aortic collar

Retroaortic left renal vein

The incidence of retroaortic left renal veins is 3.2% [17]. Retroaortic renal veins develop when the ventral arch of the aortic collar regresses and the dorsal arch remains to form the left renal vein [4]. The left retroaortic renal vein may join the IVC at the orthotopic position or more caudally at L4-5. The left renal vein may also course caudally, posterior to the aorta to join the left common iliac vein [18]. The condition is mostly asymptomatic, but can present with flank pain, hematuria, or varicocele.

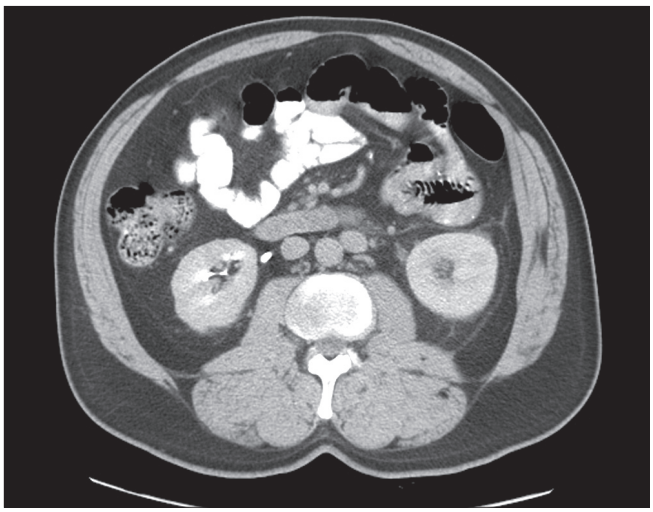

Fig. 3a. Transverse CT image of duplicated IVC.

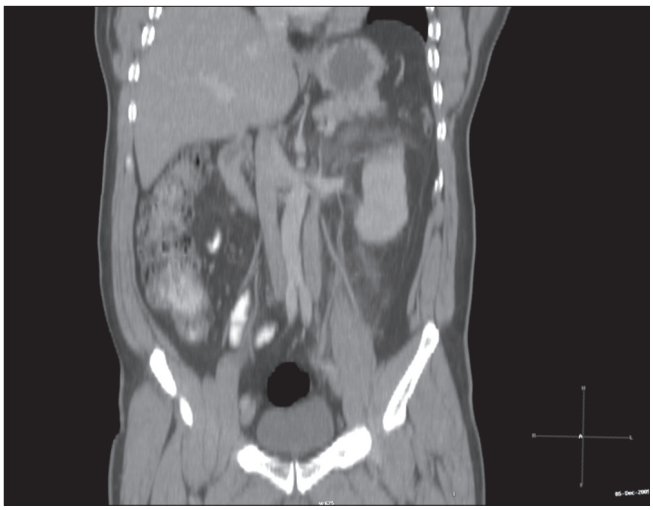

Fig. 3b. Coronal reconstruction of a CT showing a duplicated IVC.

Identification prior to surgery minimizes the risk of vascular injury and bleeding [18].

Circumaortic renal vein

In contrast to the retroaortic renal vein, a circumaortic renal vein develops when both the ventral and dorsal arch of the aortic collar persist [4]. It is usually asymptomatic and the incidence varies from 1.6-14.0% [17, 19]. A circumaortic renal vein is of great significance during donor nephrectomy [20]. In some centers, circumaortic and retroaortic renal veins are relative contraindications for donor nephrectomy [21].

Anomalies of the postcardinal veins

Retrocaval ureter (circumcaval ureter)

The embryonic ureter passes behind the postcardinal veins. When the postcardinal vein fails to regress, the ureter is left posterior to the IVC as illustrated in figure 4. Retrocaval ureter is also referred to as circumcaval ureter and has an incidence of <1% [22]. Patients with retrocaval ureter may present with symptoms of ureteric obstruction [23]. Calculi and hematuria have also been reported [5, 24]. Retrocaval ureter is diagnosed with retrograde pyelogram, intravenous pyelogram or CT/magnetic resonance (MR) urography. The proximal segment is usually dilated and lateral to the IVC. The distal segment is of normal caliber, but runs medially in the aortocaval groove prior to running laterally over the IVC. CT/MR urography with three dimensional (3D) reconstruction are both excellent tools for diagnosing and evaluating retrocaval ureter as they provide information about the IVC and other abdominal structures in addition to the urogram.

Renal function tests are recommended prior to intervention. Management of retrocaval ureter is by division and reanastomosis of the ureter anterior to the IVC. The procedure was classically performed with open surgery however laparoscopic and robotic techniques have also been successful [25, 26].

Anomalies of the subcardinal veins

Interruption of IVC with azygous continuation

Lack of development of the anastomosis between the right subcardinal vein and hepatic sinusoids results in a lack of union between the hepatic veins and the subcardinal vein [3]. The hepatic veins do not drain into the IVC. Instead, they drain directly into the right atrium [5]. Blood from the lower half of the body is shunted to the subsupracardinal anastomosis, which later forms the azygos and hemiazygos veins [5]. This azygos continuation may be associated with cardiovascular and renal anomalies [27]. However, the majority of patients with this anomaly are asymptomatic and the diagnosis is made incidentally [4]. The dilated azygos vein can be misdiagnosed as a dissecting aortic aneurysm, mediastinal mass, or lymphadenopathy [27]. Identification of the anomaly prior to cardiac catheterization or cardiothoracic procedures is crucial to avoid vascular injury [28].

Implications for renal surgery

Radical nephrectomy is the treatment of choice for localized renal cancer. It can be undertaken via open, laparoscopic or robotic approaches. A clear picture of retroperitoneal vascular anatomy is required especially with laparoscopic/robotic approaches as the field of view may be narrow and it may be difficult to appreciate aberrant venous anatomy. With open surgery, a midline or Chevron incision may provide better access to the great vessels compared to the traditional flank incision. Complete identification of the

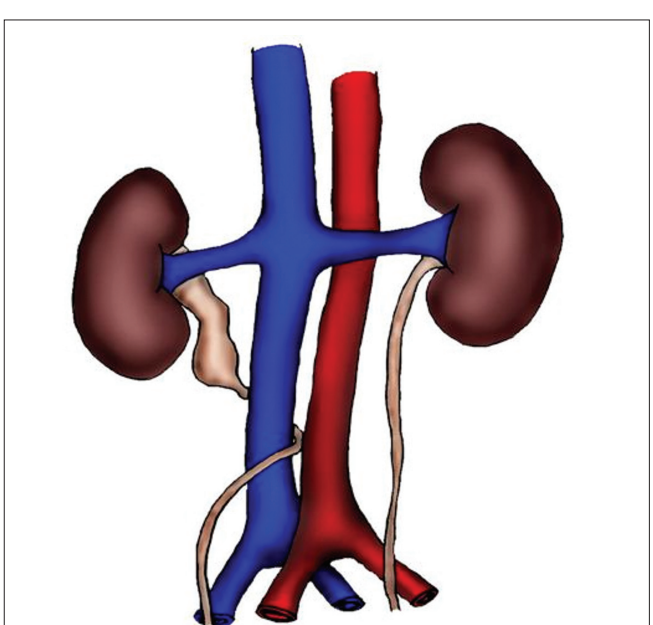

Fig. 4. Retrocaval ureter is seen.

renal vasculature is critical for safe nephrectomy. Careful planning is required and CT/MRI with 3D reconstruction is essential. Laparoscopic/robotic nephrectomy with the presence of left IVC requires more medial dissection to identify the lateral edge of the IVC prior to ligation of the renal vein. Similarly, left nephrectomy with double IVC also requires more medial dissection to the level of the aorta or left limb of the double IVC for safe ligation of the renal vein. The left limb of the double IVC must not be injured as, aside from hemorrhage, it may result in lower limb edema and/or thrombosis. Other anomalies like retro/circumaortic renal vein or multiple renal veins have lesser implications for ablative surgery, however, medial dissection is again required. Care must be taken to identify lumbar veins, which may bleed profusely if not properly controlled.

Laparoscopic donor nephrectomy is already a challenging procedure and venous anomalies add to its technical difficulty. Medial dissection is already required to maximize vessel length. Skeletonization of the renal artery and vein are required. This facilitates identification of the entire renal artery and vein as well as tributaries such as the gonadal, lumbar, and adrenal veins. Retroaortic and circumaortic renal veins may also be relative contraindications to donor nephrectomy. However, the medial dissection techniques used in donor surgery are essential for performing ablative laparoscopic/robotic nephrectomy in the setting of venous anomalies.

Identification and complications

Misdiagnosis of venous anomalies can have disastrous consequences. Dilated azygous and hemiazygous vessels have been misdiagnosed as mediastinal masses and percutaneous biopsies have been attempted [29]. Retroaortic renal veins or double IVC can be mistaken for a retroperitoneal neoplasm or as lymphadenopathy, highlighting the need for coronal reconstructions [30]. Contrast enhanced CT or MRI is required to identify vascular structures. CT/MR urography is also useful to identify retrocaval/circumcaval ureter and must be performed prior to surgical correction. Suspicious lymphadenopathy, especially if extensive i.e. has significant cranial or caudal extension, requires careful examination of coronal and 3D reconstructions enhanced with contrast. Figures 3a and 3b illustrate this point in a patient with IVC duplication. Examination of transverse CT images alone suggests the presence of para-aortic lymphadenopathy. Examination of the venous phase using 3D and coronal reconstructions demonstrates IVC duplication.

Retrocaval ureters may present with obstruction, hematuria, or stones and diagnostic or therapeutic ureteroscopy may be attempted. CT scan with or without contrast is usually performed as preoperative evaluation. Any suspicion of aberrant anatomy must be fully evaluated using contrast enhanced CT/MRI with 3D reconstructions. Complications of ureteroscopy include ureteric perforation, which can lead to major hemorrhage if the IVC is injured, especially where the ureter travels around the IVC. Retro/circumcaval ureter is corrected with ureteroureterostomy, which can be performed open, laparoscopically or robotically. Careful preoperative planning is essential using CT/MR urography with 3D reconstruction.

Finally, abnormal veins can be dilated and tortuous; therefore blood flow can be altered. This increases the chance of thromboembolic events in these anomalous veins. Recurrence of emboli from deep vein thromboses of the lower extremities in the presence of an IVC filter may suggest the presence IVC duplication [31].

Thus a high index of suspicion coupled with high-quality contrast enhanced imaging with 3D reconstruction is critical to evaluate aberrant anatomy. It is also critical for preoperative planning to ensure good surgical outcomes.

Our experience

At our center we have had experience of renal surgery in two patients with venous anomalies. The first patient underwent radical nephrectomy for a 10-cm right renal tumor. Left sided IVC was identified on preoperative CT scan with 3D reconstructions. An open approach was selected. The suprarenal IVC was identified on the right side by complete mobilization of the liver. The infrarenal IVC was seen on the left side running parallel to the aorta. After joining the left renal vein, the IVC crossed the aorta anteriorly to receive the right renal vein. The right gonadal vein drained into the right renal vein, close to the confluence of the renal vein and IVC. There were two renal arteries arising separately from the aorta. Following identification of the vascular structures the nephrectomy was completed uneventfully. Transverse and coronal CT images are displayed in figures 2a and 2b.

The second patient underwent left laparoscopic radical nephrectomy for an 8-cm left renal tumor. Double IVC was identified on preoperative CT scan with 3D reconstruction. A transperitoneal approach was used. The bowel was mobilized very medially to expose the lateral aspect of the left IVC. This allowed identification of the origin of the left renal artery and vein. The lumbar, adrenal, gonadal, and renal veins coalesced with the left IVC to form a single crossing vessel that travelled anterior to the aorta to join the right IVC. Once the vessels were identified, the nephrectomy was completed uneventfully. Transverse and coronal CT images are displayed in figures 3a and 3b.

In both instances careful preoperative planning allowed the procedure to be performed safely and highlights the need for contrast enhanced CT/MRI with coronal and 3D reconstructions.

CONCLUSION

Anomalies of the IVC or renal veins are rare anatomical variants that result from abnormal development/regression of the fetal venous circulation. With the exception of retrocaval/circumcaval ureters, the majority of patients are asymptomatic and these variants are detected incidentally on CT/MRI. Their presence may have significant implications for surgical procedures, as careful preoprative planning is required. This is particularly the case for procedures such as laparoscopic radical nephrectomy or laparoscopic donor nephrectomy.

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