Bilateral inferior vena cava in a patient with lymphoma – clinical anatomy and embryology

Bruce A. Schoneboom¹, Padmanabhan Rengasamy²

¹- Graduate School of Nursing, Uniformed Services University of Health Sciences (USUHS)†, Bethesda, MD
²- Division of Basic Medical Sciences, Mercer University School of Medicine, Macon, Georgia 31207, USA

SUMMARY

The development of the inferior vena cava (IVC) includes contributions from diverse embryonic venous precursors and involves major cross connections and left-to right-shunting. This complex process is vulnerable to errors. However, congenital malformations of the IVC are reported to be rare. Most of these malformations are accidental findings in chest and abdomen imaging. Anomalies of the IVC are compatible with life but are of major clinical importance. This paper describes a case of bilateral IVC in a male cadaver observed by dissection. The common iliac veins were found to continue superiorly as the IVC and fused together slightly cranial to the level of the origin of the superior mesenteric artery from the aorta. The left IVC received the left gonadal vein and, slightly superior to its union with the renal vein, it received the left suprarenal vein. It fused side-to-side with the left renal vein, crossed to the right side anterior to the aorta and fused with the right IVC to form the common IVC. The embryology and clinical relevance of this anomalous IVC are discussed. Currently there is little emphasis on the clinical implications of IVC anomalies in medical school anatomy and embryology teaching. The most widely used textbooks dismiss vena cava malformations with the indication “they are not common”. A lack of knowledge of possible vascular malformations might result in a misinterpretation of the radiologic images of anomalous veins, leading to surgical errors such as bleeding during retroperitoneal lymph node dissection. Anatomy and embryology textbooks need to incorporate the findings that have emerged recently and are summarized here.

Key words: Bilateral inferior vena cava – Embryology of vena cava – Anatomy of double IVC – Clinical relevance of double IVC

INTRODUCTION

In its most common presentation the inferior vena cava (IVC) is formed by the union of the common iliac veins anterior to the fifth lumbar vertebral body, slightly to the right of the median plane. From here it ascends on the right side of the abdominal aorta. It then grooves the posterior surface of the liver, perforates the central tendon of the diaphragm at the T8 vertebral level, immediately pierces the fibrous pericardium, and opens into the lower posterior part of the right atrium. Its atrial opening is guarded by a valve, which in...
the fetus guides the blood flow to the left atrium. Embryological development of the IVC is complex, with contributions coming from bilaterally symmetrical primordial veins that emerge from a plexiform veins of the posterior body wall. The developmental mechanisms include delicately balanced processes of selective regression of certain parts and the persistence of the remaining parts of the precursor veins, as well as a series of complex anastomoses between them. Developmental errors such as failure to regress or incomplete regression result in abnormal patterns of IVC presentation. Until recently, several types of anomaly of the systemic venous system were discovered during cardiac catheterization for congenital heart diseases, abdominal surgery for other reasons, and cadaver dissections in anatomy laboratories or at autopsy (Mayo et al., 1980). However, with the advent of advances in imaging technology, malformations of the IVC have been recognized, often in asymptomatic patients who are imaged for other reasons and they are now reported more frequently than before (Cuevas et al., 2006). A lack of knowledge of vascular anomalies might result in misinterpretation of the radiologic images of anomalous veins as retroperitoneal growths or tumors, and in surgical errors with serious consequences, such as bleeding during retroperitoneal lymph node dissection (Nevruz et al., 2007). In the present report, the authors present a case of bilateral IVC and describe the embryology and clinical relevance of this anomalous structure.

CASE REPORT

This was one of the 14 adult cadavers used for the MSN program at the Graduate School of Nursing, Uniformed Services University of Health Sciences, Bethesda, MD. The inferior vena cava (IVC) was found to be of bilateral nature. On each side the IVC started off as the superior continuation of the common iliac vein formed at the corresponding sacroiliac joint. The internal iliac veins appeared short and convoluted. Each IVC ran retroperitoneally medial to the corresponding ureters, and anterior to the corresponding genitofemoral nerve, psoas major and sympathetic chain. The right one resembled the normal IVC in all its anterior relationships with the root of the mesentery, gonadal vessels, 1st and 3rd parts of the duodenum, and the head of the pancreas. It received the testicular vein slightly inferior to the point of union of the right renal vein. The right suprarenal vein was seen to unite with the right IVC immediately superior to the renal vein. After a course of about 2.5 cm from here, it was joined by the left IVC. The left IVC was found to course superiorly on the lateral side of the left common iliac artery and abdominal aorta but medial to the left ureter. It ran slightly obliquely away from the median plane as high as the L2 vertebra and then turned superomedially, crossing anterior to the aorta and posterior to the pancreas, and joined the right IVC to form the common IVC (Fig. 1). Anteriorly, the left IVC had the left gonadal vessels, the left colic vessels and coils of small intestine in its caudal portion and the body of the pancreas in its superior course. It was joined by the left testicular vein about 2.5 cm inferior to the level of its union with left renal vein. Slightly superior to this union, the left IVC received the opening of the left suprarenal vein. The left renal vein, being significantly longer than its counterpart, fused extensively with the left IVC and common IVC, the fusion being one of side-to-side rather than end-to-side. The side-to-side union is possibly the reason why the suprarenal vein appeared to unite with the left IVC. Both right and left renal veins described an oblique course superiorly before their union with the corresponding IVC. The common IVC coursed superiorly for about 2.5 cm, grooved the posterior surface of the liver, perforated the central tendon of the diaphragm and fibrous pericardium, and opened immediately into the lower posterior portion of the right atrium. Huge cysts were found to occupy the superior half of the left kidney. The right kidney was hypoplastic and about a third of the left kidney in size. The lymph nodes of the abdomen were numerous, large in size and firm to the touch. So were the nodes in the rest of the body.

This case exemplifies the duplication of the infrarenal segment of the IVC. Despite technological advances in ultrasound, MR and CT imaging, autopsy and cadaveric dissections still remain the time-honored opportunity, whereby extensive exploration and documentation of the anatomical disposition of normal and anomalous blood vessels and their relationship to adjacent organs are possible. This dissection-based study also vividly illustrates the presentation and gross relationships of the bilateral IVC. Congenital IVC anomalies
reported in the literature include agenesis of IVC, hypoplastic IVC, interrupted or stenotic IVC, unilateral left IVC with or without *situs inversus*, duplication of the IVC etc. (Koc and Oguzkurt, 2007). About 90% of the malformations of the IVC have been reported to involve the suprarenal and hepatic segments (Kondo et al., 2009). Double IVC is reported to occur in about 0.5-3% of individuals and may be associated with other complex vascular malformations or pathologies, such as abdominal aortic aneurysm and deep vein thrombosis of lower limbs (Nishibe et al., 2004). Our case largely resembles the one described by Bass et al. (2000) in their CT observation on a 53-year-old woman with lymphoma. Contrary to textbook descriptions of the left IVC (Moore and Persaud, 2008), our cadaver had both IVC of comparable size, but individually both had about half the diameter of otherwise normal right-sided IVC, resembling the Type 2 of the description by Natsis et al. (2010). Both IVC were found to be located in a plane anterior to the ureters, suggestive of an embryonic subcardinal vein origin (Edwards, 1951). The right kidney was hypoplastic while the left kidney contained large cysts. In our case, the renal veins emptied into the IVC of the corresponding side. The left renal vein fused side-to-side with the left IVC, the union continuing into the infra-hepatic segment of the common IVC. The renal veins described a rather oblique and upward course to unite with the IVC of the corresponding side (Fig. 1).

If not suspected, the presence of such aberrant vascular structures might be misdiagnosed as lymph adenopathy (Evans et al., 2001) in abdomen scans. Double IVC has been reported to be present in association with congenital portal-systemic shunt and portal vein hypoplasia, requiring surgical intervention in the pediatric population (Chiu et al., 2008).

Fig. 1. The posterior abdominal wall showing the formation of the bilateral IVC as the superior continuation of the common iliac vein of either side. Note that the left renal vein fuses side-to-side with the left and common IVC. Abbreviations: CL, Caudate lobe; PH, Porta hepatis; CIVC, Common inferior vena cava; RIVC and LIVC, right and left inferior vena cavae; RSRV and LSRV, right and left suprarenal veins; RK and LK, right and left kidneys; RGV and LGV, right and left gonadal veins; RU and LU, right and left ureters respectively; AO, aorta.
The presence of an IVC on the left side significantly shortens the length of the left renal vein and therefore in live donor renal transplantation procedures (Gupta et al., 2002), a cuff of left IVC is taken along with renal vein to facilitate venous anastomosis in the recipient. Malformations of the IVC are increasingly recognized as a risk factor for deep vein thrombosis (DVT) (Nishibe et al., 2004; Kondo et al., 2009). In patients with an indwelling IVC filter to prevent recurrent pulmonary embolism the possible presence of IVC with azygos continuations or double IVC should be suspected because the anomalous IVC might be an alternate route for leg DVT (Nanda et al., 2008). The presence, size-symmetric or asymmetric of unusual connections (retroaortic left renal vein, azygos continuation, portal and mesenteric venous connection, etc.) should be evaluated before choosing the size and site of placement of IVC filters (Nanda et al., 2008; Kondo et al., 2009). Currently, little emphasis is placed on the clinical implications of IVC anomalies in medical school anatomy and embryology teaching, and the most commonly used textbooks (Moore and Persaud, 2008) dismiss vena cava malformations with the indication “they are not common”. Thus, findings that have emerged recently need to be incorporated into the body of medical knowledge, as we suggest here.

The etiology and exact mechanisms by which anomalous IVC develop are not yet known. Several venous primordia appear to contribute to the development of the IVC. During the early phase of organogenesis, bilaterally symmetric veins, known as cardinal veins, develop from plexiform endothelial venous networks of the embryonic body wall (Hamilton et al., 1966). These are (1) the anterior and posterior cardinal veins (2) the subcardinal veins and (3) the supracardinal veins. The anterior cardinal vein, also called the precardinal vein, lengths caudally and unites with the posterior (or post) cardinal vein to form the common cardinal vein (duct of Cuvier), which opens into the sinus venosum of the embryonic heart. The subcardinal and supracardinal veins initially appear plexiform and are situated parallel and medial to the posterior cardinal vein. Subsequently, they both have their cranial and caudal ends terminating in the posterior cardinal vein of the corresponding side, which they reinforce functionally (Hamilton et al., 1966). The brachiocephalic (innominate) anastomosis between the anterior cardinal veins rostrally and the iliac anastomosis between the posterior cardinal veins caudally shunt the blood from left to right, as a result of which the left posterior cardinal veins between these two anastomoses become mostly regressed. The intersubcardinal anastomosis develops when the embryos are of 10 mm CR length and further shunts the blood from left to right, and hence the left supracardinal and subcardinal veins also largely disappear, leaving behind only some remnants, namely the suprarenal and gonadal veins, which empty via the left renal vein, developing largely from the inter-subcardinal anastomosis. With the development of ductus venosus, shunting blood from the left umbilical vein to the right hepatocardiac channel, the left hepatocardiac channel and the right hepatocardiac channel becomes the dominant vessel transporting blood to the sinus venosus of the embryonic heart. During this period, new anastomoses develop and successively connect the right (a) caudal part of the posterior cardinal vein with the supracardinal vein, (b) the supracardinal with the subcardinal and (c) the subcardinal with the hepatocardiac channel (Moore and Persaud, 2003). This results in regression of most of the right posterior cardinal vein. Thus the definitive IVC derives from the (1) caudal portion of the right posterior cardinal vein, (2) the posterior cardinal-supracardinal anastomosis, (3) the adjoining segment of the supracardinal vein (4) the supracardinal-subcardinal anastomosis, (5) the adjoining segment of the subcardinal vein (6) the anastomosis between the subcardinal and hepatocardiac channel and (7) the hepatocardiac channel itself. In our case, the left IVC possibly developed as a result of (1) failure of the caudal iliac anastomosis (2) a persistence of the left supracardinal and subcardinal veins and the anastomosis between them. The caudal portions of the subcardinal veins receive embryonic gonadal veins (McClure and Butler, 1925) and thus form part of the testicular/ovarian veins. Since the left subcardinal vein is incorporated into left IVC, the left gonadal vein opened into the left IVC of our case.

In addition to hemodynamic factors, which cellular and molecular mechanisms might be involved in the preferential regression of certain segments and the persistence of the remaining parts of the venous primordia and in the initiation and progression of several anastomoses between the primordial veins are not known. Multiple contributions - such as genetic factors and environmentally triggered alterations in the expression and regulation of
growth factors that control the epithelial–mesenchymal transformations necessary for the formation of the endothelium (e.g. TGF-β), along with vascular endothelial growth factors (VEGF) and several VEGF receptors, might also play crucial roles in these complex network of embryonic primordial blood vessels (Chua et al., 2009). Certain VEGF isoforms have been suggested to be involved in the early stages of normal vascular assembly, arterial and venous differentiation, vascular remodeling and pattern formation (Stalmans, 2005). Nevertheless, it is clear that errors (e.g. thombosis) in any stage of development of the precursor veins or in their subsequent reorganization might result in anomalies of the IVC. Anomalies of the IVC often coexist with other cardiovascular and visceral malformations. Isolated IVC malformations are asymptomatic, possibly because of extensive collateral circulation via the azygos veins, hemiazygos veins, lumbar veins, or the vertebral plexus of veins. Thus, contrary to reports stating that IVC anomalies are rare, the actual incidence possibly remains under reported. Sophistication in imaging techniques has resulted in relatively easy identification, accurate descriptions and an increased frequency of the reporting of venous anomalies. As long as clinicians become aware of the potential existence of IVC anomalies in conjunction with other malformations, the possibility for better prognosis will be expected to increase in future.

Acknowledgements

The authors are grateful to the staff of the Anatomy Teaching Laboratory of USUHS for their valuable technical assistance.

Conflicts of interest

The authors do not have conflicts of interest. The views expressed are those of the authors and do not reflect the official views of the University or the Department of Defense.

References


Edwards EA (1951) Clinical anatomy of lesser variations of the inferior vena cava; and a proposal for classifying the anomalies of this vessel. Angiology. 2: 85-99.


