Right Double Inferior Vena Cava with Preaortic Iliac Confluence – Case Report and Review of Literature

C.S.RAMESH BABU¹, REKHA LALWANI², INDRA KUMAR³

ABSTRACT
Anomalies of the inferior vena cava (IVC) are uncommon and most of them remain asymptomatic. Though rare, anomalies of IVC can lead to severe hemorrhagic complications especially during aortoiliac surgery. Prior knowledge of these variations facilitates proper interpretation of radiological images and safe performance of interventional procedures and surgeries. During routine anatomical dissection of abdomen in a female cadaver we observed the presence of right sided duplication of inferior vena cava. Both IVCs were present on the right side of the abdominal aorta, one more ventrally and medially and the other more dorsally and laterally. These two IVCs were designated as ventral right IVC (VRIVC) and dorsal right IVC (DRIVC). The wider VRIVC, which appeared to be the main IVC, was formed by the union of two common iliac veins anterior to the right common iliac artery [Table/Fig-1]. This pattern of formation of IVC in front of right common iliac artery or aortic bifurcation is called “Preaortic iliac confluence” - “Marsupial Cava”). The right external iliac vein continued as the more dorsally and laterally placed dorsal right IVC. The right internal iliac vein after receiving a transverse anastomotic vein from the external iliac continued as the right common iliac vein. This transverse anastomosis was present behind the right common iliac artery. The narrower dorsal right IVC joined the wider ventral right IVC just below the level of renal veins to form a single IVC. The abdominal aorta presented a convexity to the left.

CASE REPORT
During routine anatomical dissection of retroperitoneum, a very rare anomaly of right sided duplication of IVC with preaortic iliac venous confluence was observed in a female cadaver. Both IVCs were present on the right side of the abdominal aorta, one more ventrally and medially and the other more dorsally and laterally. These two IVCs were designated as ventral right IVC (VRIVC) and dorsal right IVC (DRIVC). The wider VRIVC, which appeared to be the main IVC, was formed by the union of two common iliac veins anterior to the right common iliac artery [Table/Fig-1]. This pattern of formation of IVC in front of right common iliac artery or aortic bifurcation is called “Preaortic iliac confluence” or “Marsupial Cava”). Both common iliac veins were present posteromedial to the respective common iliac arteries. The right common iliac vein was formed by the union of internal iliac vein and an anastomotic vein from external iliac vein passing posterior to the right common iliac artery. The right external iliac vein continued lateral to right common iliac artery as the dorsally and laterally placed DRIVC. Both IVCs joined to form a single trunk just below the level of renal veins [Table/Fig-2]. It appeared that the right common iliac artery was passing through a venous ring [Table/Fig-3]. Right ovarian vein drained into VRIVC. The left renal vein received left ovarian and left suprarenal veins. The abdominal aorta presented a slight convexity to the left. Right ureter was normal in its position and relations lying lateral to both vena caval channels and crossing the bifurcation of common iliac artery [Table/Fig-3].

DISCUSSION
Congenital anomalies of the inferior vena cava (IVC) occur in approximately 2% to 3% of patients [1] and are detected incidentally since most of them are clinically silent. The incidence of double IVC or duplication of IVC was reported to be 0.2 % - 3 % [2]. Ipsilateral duplication of IVC is extremely rare. Review of 

Keywords: Double inferior vena cava, Marsupial cava, Inferior vena caval anomalies
Among the anomalies of IVC, the most common is double IVC with an incidence of 0.2% - 3% [2]. Classically the double IVC indicates the presence of right and left IVC on either side of the abdominal aorta representing the persistence of both right and left supracardinal veins. Ipsilateral duplication of IVC is very rare. Right double IVC is defined as the presence of two infrarenal IVCs lying as dorsally placed IVC without any interiliac anastomosis. Ng and Zajko [22] described five cases of right double IVC with a ventral and dorsal relationship between the two IVCs. All five cases also exhibited preaortic left common iliac vein which continued as ventrally placed IVC and the right common iliac vein continued as dorsally placed IVC without any interiliac anastomosis. Ng and Ng [23] reported a right sided double IVC by MR imaging with the

manifestations of a complex process of embryogenesis involving anastomosis between three paired venous channels (posterior cardinal, supracardinal and subcardinal) with enlargement and consolidation of some vessels due to hemodynamic changes and regression of others.

Normally the IVC is formed by the union of the two common iliac veins posterior to right common iliac artery. Formation of IVC anterior to right common iliac artery or aortic bifurcation is named as Preaortic iliac confluence [4]. Earliest reports of such anomalous formation appeared in the year 1929 [5,6]. This anomaly is very rare in humans but normal in Marsupial animals and hence the term “Marsupial Cava” [7]. Many cases of this anomaly reported in the literature were detected incidentally during abdominal aortic aneurysm surgery [8-13]. Natsis et al., [14] described an anatomical finding of a case of preaortic iliac confluence and Rocha et al., [15] detected four such cases and including them counted 17 such cases till 2008. Later 3 more cases were reported thus making a total of 20 cases [16,17]. In most of these cases preaortic iliac confluence formed a normal orthotopic IVC in a right lateroaoortic position [Table/Fig-4].

Tagliafico et al., [18] described a case of double right IVC in which a venous ring encircled the right common iliac artery. Though the authors did not use the term preaortic iliac confluence, the formation of ventral IVC appeared to be preaortic iliac venous confluence. To the best of our knowledge this is the first case report of right double IVC associated with preaortic iliac confluence.

[Table/Fig-4]: Features of cases of preaortic iliac venous confluence

<table>
<thead>
<tr>
<th>Sl.No.</th>
<th>Name of author &amp; year</th>
<th>No. of cases</th>
<th>Sex</th>
<th>Comparison of size of two IVCs, (Venral = V Dorsal = D)</th>
<th>Level of confluence of the two IVCs from renal veins confluence</th>
<th>Drainage site of right gonadal vein</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Doyle et al., 1992 [19]</td>
<td>1</td>
<td>M</td>
<td>V &gt; D</td>
<td>At the level of renal veins</td>
<td>NA</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>Meyer et al., 1998 [20]</td>
<td>1</td>
<td>NA</td>
<td>V &gt; D</td>
<td>Caudal to renal veins</td>
<td>NA</td>
<td>Spiral CT.</td>
</tr>
<tr>
<td>3</td>
<td>Nagashima et al., 2006 [22]</td>
<td>5</td>
<td>M-2 F-3</td>
<td>V &gt; D – 3 V &lt; D – 1 V = D -1</td>
<td>Caudal to renal veins</td>
<td>VIVC – 4 IVC -1</td>
<td>CT. Left common iliac vein continued as Ventral IVC crossing ventral to aortic bifurcation or right common iliac artery. Right common iliac vein continued as Dorsal IVC. No interiliac communication.</td>
</tr>
<tr>
<td>4</td>
<td>Senecal et al., 2004 [21]</td>
<td>1</td>
<td>F</td>
<td>V &lt; D</td>
<td>Caudal</td>
<td>VIVC</td>
<td>CT. VIVC continuation of LCV. DVC formed by RCIV with an anastomotic branch from LCV.</td>
</tr>
<tr>
<td>5</td>
<td>Tagliafico et al., 2007 [18]</td>
<td>1</td>
<td>M</td>
<td>V &gt; D</td>
<td>Caudal to renal veins</td>
<td>VIVC</td>
<td>US, CT. Venous ring encircling right common iliac artery.</td>
</tr>
<tr>
<td>6</td>
<td>Ng and Ng, 2009 [23]</td>
<td>1</td>
<td>F</td>
<td>NA</td>
<td>At renal veins level</td>
<td>NA</td>
<td>US, MRI. Left common iliac vein passed behind aorta and ascended as double IVC.</td>
</tr>
<tr>
<td>7</td>
<td>Gong et al., 2011 [24]</td>
<td>1</td>
<td>F</td>
<td>NA</td>
<td>Midsegment duplication</td>
<td>NA</td>
<td>Partial right double IVC with circumcaval ureter.</td>
</tr>
<tr>
<td>8</td>
<td>Present Case, 2013</td>
<td>1</td>
<td>F</td>
<td>V &gt; D</td>
<td>Caudal to renal veins</td>
<td>VIVC</td>
<td>Ventral IVC showing Preaortic iliac venous confluence; Dorsal IVC continuation of right external iliac vein.</td>
</tr>
</tbody>
</table>

[Table/Fig-5]: Features of cases of right double inferior vena cava

1788 contrast enhanced spiral CT scans of abdomen revealed an incidence of 0.39 % for duplication of IVC and not a single case of preaortic iliac vein confluence and right sided duplication was found in this retrospective review [3] Anomalies of IVC are unusual
importance of awareness of these venous anomalies cannot be overemphasized. Aneurysm surgery, retroperitoneal surgeries, placement of IVC stents during preoperative evaluation, and radiologists and vascular surgeons. Since some of the venous anomalies are frequently overlooked during preoperative evaluation, these anomalies are important entities to the practitioners. Anomalies of IVC are rare and clinically asymptomatic and may remain undetected. These anomalies are important entities to the radiologists and vascular surgeons. Since some of the venous anomalies are frequently overlooked during preoperative evaluation, intraoperative awareness is most essential to avoid unexpected surgical hazards. Most important clinical consequences of double IVC with preaortic iliac confluence are observed in abdominal aortic aneurysm surgery, reoperation of tortuous iliac and renal veins, placement of IVC filters and surgical ligation of IVC. Thrombosed double IVC may be mistaken for lymphadenopathy or a retroperitoneal mass. The importance of awareness of these venous anomalies cannot be underestimated because of their rarity.

REFERENCES
[25] Tsuyoshi et al., [25] described the continuation of left external iliac vein as the left IVC passing through the split between the two IVCs. The partial double right IVC was due to the persistence of both right supracardinal and right subcardinal veins. Embryologically the preaortic iliac confluence probably represents the persistence of the ventral limb of circumumbilical venous ring which surrounds the future common iliac arteries on each side and regression of dorsal limb of the venous ring. Normally the ventral portion of the venous ring disappears. This conformation is similar to the normal formation of left renal vein but occurs in a more caudal location at the level of aortic bifurcation.

CONCLUSION
Anomalies of IVC are rare and clinically asymptomatic and may remain undetected. These anomalies are important entities to the radiologists and vascular surgeons. Since some of the venous anomalies are frequently overlooked during preoperative evaluation, intraoperative awareness is most essential to avoid unexpected surgical hazards. Most important clinical consequences of double IVC with preaortic iliac confluence are observed in abdominal aortic aneurysm surgery, reoperation of tortuous iliac and renal veins, placement of IVC filters and surgical ligation of IVC. Thrombosed double IVC may be mistaken for lymphadenopathy or a retroperitoneal mass. The importance of awareness of these venous anomalies cannot be underestimated because of their rarity.