

Double inferior vena cava: a report of three cases

Ng W T, Ng S S M

ABSTRACT

Double inferior vena cava (IVC) is a congenital anomaly resulting from the persistence of the embryonic venous system. The majority of cases are clinically silent and diagnosed incidentally on imaging for other reasons. However, these venous anomalies may have significant clinical implications, especially during retroperitoneal surgery and in the treatment of thromboembolic diseases. We report three cases of double IVC and review the relevant literature. The clinical importance of recognising double IVC is discussed.

Keywords: computed tomography, double inferior vena cava, inferior vena cava, venous anomaly

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INTRODUCTION

Double inferior vena cava (IVC) is a congenital anomaly resulting from the persistence of the embryonic venous system. It is a relatively uncommon condition with a reported incidence of 0.2%–3%.⁽¹⁾ The majority of cases are clinically silent and are diagnosed incidentally by imaging (including computed tomography [CT] and magnetic resonance [MR] imaging) done for other reasons. However, these venous anomalies may have significant clinical implications, especially during retroperitoneal surgery or in the treatment of thromboembolic diseases.⁽²⁻⁵⁾ Three cases of double IVC are being reported. We also review the relevant literature and discuss the clinical importance of recognising double IVC.

CASE REPORTS

Case 1

An 81-year-old woman with a past medical history of ischaemic heart disease and chronic obstructive airway disease presented with fever and jaundice. The liver function tests showed an obstructive pattern. Ultrasonography of the abdomen showed gallstones but no features of acute cholecystitis or cholangitis. Two tubular structures were incidentally noted on the right side of the abdominal aorta, representing venous

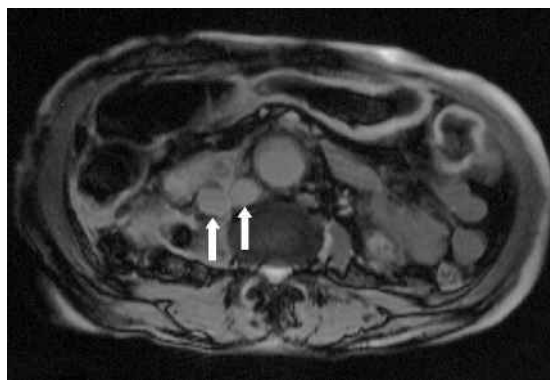


Fig. 1 Axial T2-W MR image shows a double right-sided inferior vena cava (arrows). The two inferior venae cavae ascend on the right side of the abdominal aorta and join at the renal level.

abnormalities. In view of these abnormal features, MR imaging confirmed that the left iliac vein had crossed the midline behind the aorta and ascended as double IVC (Fig. 1). The two inferior venae cavae then joined at the renal level. The patient subsequently underwent endoscopic retrograde cholangiopancreatography with sphincterotomy performed, but no common bile duct stone was found. Nonetheless, the liver function tests gradually improved afterwards. The patient preferred conservative treatment for the gallstones in view of her advanced age.

Case 2

A 51-year-old chronic, alcoholic man presented with recurrent epigastric pain associated with an elevated serum amylase level. CT of the abdomen revealed a pancreatic pseudocyst associated with pancreatic ductal dilatation and features suggestive of gastric outlet obstruction. There was an incidental finding of double IVC on CT. In addition to the usual right-sided IVC, another IVC was seen on the left side of the abdominal aorta (Fig. 2), which drained into the left renal vein (Fig. 3). The patient subsequently underwent internal drainage of the pancreatic pseudocyst. The postoperative course was uneventful.

Case 3

A 45-year-old man with good past health presented with a few months' history of perrectal bleeding. Digital rectal examination showed a circumferential rectal tumour

Department of Surgery,
The Chinese University of Hong Kong,
Prince of Wales Hospital,
Shatin,
New Territories,
Hong Kong SAR

Ng WT, MBChB,
MRCSE
Resident

Ng SSM, MBChB,
FRCSE, FHKAM
Associate Professor

Correspondence to:
Dr Simon SM Ng
Tel.: (852) 2632 1495
Fax: (852) 2637 7974
Email: simonng@
surgery.cuhk.edu.hk

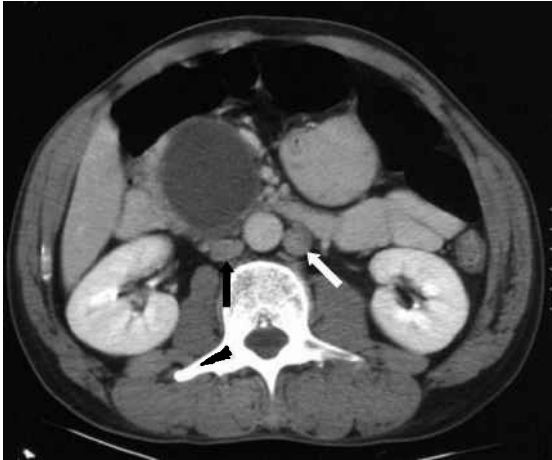


Fig. 2 Axial CT of the abdomen shows a double inferior vena cava (IVC). In addition to the usual right-sided IVC (black arrow), another IVC is seen on the left side of the abdominal aorta (white arrow).

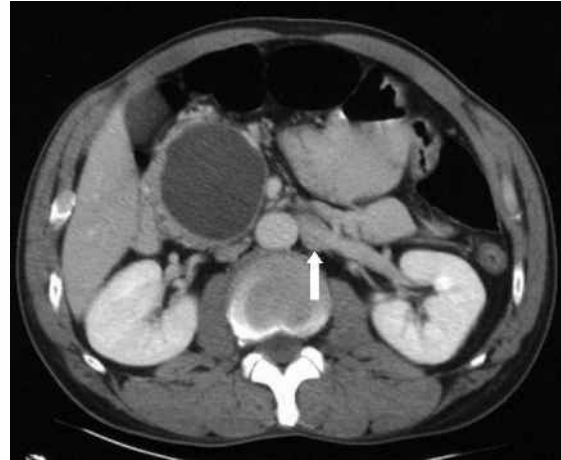


Fig. 3 Axial CT of the abdomen shows a double inferior vena cava (IVC). The left-sided IVC drains into the left renal vein (arrow).

located 7 cm from the anal verge. CT of the abdomen and pelvis revealed a rectosigmoid tumour with liver metastasis. Double IVC was incidentally noted on CT. In addition to the usual right-sided IVC, there was a left-sided IVC that drained into the left renal vein. The patient underwent palliative Hartmann's procedure with residual disease left in the pelvic side wall. Extra care was taken during mobilisation of the left-sided colon to avoid injury of the left-sided IVC. The patient received palliative chemotherapy after surgery, but he finally died of disease progression.

DISCUSSION

Embryogenesis of IVC is a complex process involving the development, regression, anastomosis and replacement of three pairs of embryonic veins (posterior cardinal, subcardinal and supracardinal veins). This process begins at the sixth week of gestation and is completed by the tenth week. The posterior cardinal veins appear first on the posterior aspect of the embryo. These veins regress, except for the distal aspects which become the iliac bifurcation. The subcardinal veins then appear anterior and medial to the posterior cardinal veins. The right subcardinal vein remains to form the suprarenal IVC, while the left subcardinal vein completely regresses. Subsequently, the supracardinal veins appear dorsally to the subcardinal veins. The left supracardinal vein then regresses, and the right supracardinal vein forms the infrarenal IVC.^(1,6)

The anomalies of IVC arise from the failure of normal embryogenesis. The most commonly-described anomalies of IVC include circumaortic left renal vein (1.5%–8.7%), azygous or hemiazygous continuation of IVC (0.6%), retroaortic left renal vein (2.1%), double IVC

(0.2%–3%) and isolated left-sided IVC (0.2%–0.5%).⁽¹⁾ Double IVC results from the failure of regression in embryogenesis.^(1,7) The most common pathogenesis is the failure of regression of the left supracardinal vein. The duplicated left IVC usually drains into the left renal vein, which then crosses anterior to the aorta and joins the right IVC in a normal fashion. In our series, two cases demonstrated such an anomaly. The remaining case showed two right-sided inferior venae cavae. It is speculated that the two inferior venae cavae are derived from the right supracardinal and subcardinal veins.⁽⁶⁾

The majority of cases of double IVC are diagnosed incidentally by imaging for other reasons, but these anomalies can have significant clinical implications. Radiologically, the presence of double IVC can be mistaken as a pathological lesion such as lymphadenopathy,^(8,9) or left pyeloureteric dilatation.⁽¹⁰⁾ There are case reports describing patients who underwent exploration for presumed metastatic testicular carcinoma based on the CT appearance of the anomaly.^(8,11)

The presence of double IVC may also complicate retroperitoneal surgery.^(2,3) The double IVC can be inadvertently injured or ligated during retroperitoneal surgery. Therefore, it is important to identify such anomalies preoperatively, as illustrated in Case 3. Moreover, it has been suggested that the transperitoneal approach rather than the retroperitoneal approach should be adopted for patients with an abdominal aortic aneurysm and concomitant double IVC.⁽²⁾

There are several case reports of thromboembolic events occurring in patients with double IVC.^(4,5) There appears to be an increased incidence of thrombosis formation in double IVC, but the exact cause is unknown. Some authors suggested that this may be related to

the degree of narrowing of the vessel as it crosses the aorta.⁽¹²⁾ In patients who require IVC filter placement, separate filters are needed for both IVCs.

In conclusion, double IVC is a rare congenital anomaly. The majority of cases are diagnosed incidentally on imaging for other reasons. However, these venous anomalies should be recognised, as they can have significant clinical implications, especially during retroperitoneal surgery or in the treatment of thromboembolic diseases.

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