

Left-sided IVC in left renal cell carcinoma



Ann. Ital. Chir., 2013 84: 107-110

pii: S0003469X1201799X

www.annitalchir.com

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AIM: Congenital anomalies of the inferior vena cava (IVC) are very rare and extremely diverse, reflecting the complexity of the embryological development of these structures. The variants must be differentiated from pathology, particularly lymphadenopathy, on imaging studies as their presence can affect surgical and interventional procedures in retroperitoneum. We describe two patients with renal cell carcinoma of left kidney and left IVC.

CASE REPORT: First patient was taken up for left radical nephroureterectomy. During surgery the existence of a transposed left IVC was demonstrated. The second case is a fifty-four-year-old man; abdomen and pelvic CT-scan with coronal maximum intensity projection reconstruction showed a 7 cm heterogeneously enhancing neoformation involving the left kidney with intraparenchymal hematoma and a transposed left IVC.

CONCLUSIONS: Preoperative detection of congenital IVC anomalies can prevent morbidity. Once diagnosed, appropriate care must be taken during the operation to expose and define the anatomic anomaly and protect it from injury.

KEY WORDS: Diagnostic Errors, Lymphatic Metastasis, Renal cell carcinoma, Retroperitoneal Neoplasms, Inferior Vena Cava

Introduction

Standard treatment of renal cell carcinoma (RCC) includes radical nephrectomy as well as removal of the paraaortic and paracaval lymph nodes. The potential benefits of a lymph node dissection (LND) include more

accurate staging, decreased local recurrence rates, and improved survival. Although little evidence supports the value of LND for RCC, occasionally, a patient may have very early metastasis confined to the area of the primary and secondary major lymphatic flow medially and perhaps benefit therapeutically. Patients with tumors on the right side undergo removal of all the tissue to the right and behind the inferior vena cava (IVC) as well as the interaortocaval lymph nodes. The superior extent is the renal hilum and the distal extent is the approximate level of the inferior epigastric artery. On the left side, the superior and inferior limits are the same. However, the LND is confined to the left paraaortic lymph nodes. This adds little time or risk to the surgery and provides important staging information. We reviews two RCC of left kidney associated with an unusual retroperitoneal lymph node metastasis.

Pervenuto in Redazione Luglio 2011. Accettato per la pubblicazione Ottobre 2011

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Left-sided IVC

During normal development, the infrarenal IVC develops from the right supracardinal vein during the seventh to eighth week of gestation. The posterior cardinal veins, which develop earlier during the fourth week, provide the outflow from the lower extremities by developing into the iliac veins. These veins form ventral and dorsal anastomoses in the midline, and then divide again as they run caudally. Once the right supracardinal vein develops, it descends and joins the iliac confluence in the dorsal location. The anterior anastomosis regresses; persistence of this anastomosis and joining of the right supracardinal vein at this point lead to preaortic iliac vein confluence. The left supracardinal vein generally regresses, as do the posterior cardinal veins above the iliac confluence. If the left supracardinal vein persists and the right vein regresses, this leads to a left-sided IVC. This is generally a mirror image transposition below the renal veins; the left gonadal vein inserts on the IVC and the right inserts on the right renal vein. The IVC crosses to the right side at approximately the level of the renal arteries, generally crossing in front of the aorta. Prevalence of this anomaly ranges from 0.04% to 1.5% in radiographic series, 0.1% to 0.3% in surgical series, and 0.1% to 0.6% in autopsy series. In general, a left-sided IVC is asymptomatic, as the caliber of the vessel is equivalent to that of the cava in the orthotopic position. Preoperatively, a cavagram should be obtained through a left femoral vein puncture to accurately roadmap the IVC anomaly, the renal veins, and the level of crossing of the IVC to the right. Exposure options for the aorta in this case include dissection and mobilization of the IVC, division of the right renal vein, and division of the IVC.

Case report

The first patient is a sixty-six-year-old woman with myeloproliferative disease, who was admitted for occasional finding of a left renal mass during haematologic follow-up. Physical examination revealed no abnormalities. Hematological and biochemical parameters were within normal limits. CT scan of abdomen revealed a 4 cm oval shaped heterogeneously enhancing neoformation involving inferior iuxtapolar region of left kidney. There was a rounded structure at the left side of aorta and it was regarded to be a retroperitoneal lymphadenopathy. Patient was taken up for left radical nephroureterectomy. During surgery the existence of a transposed left IVC was demonstrated (Fig. 1a). The IVC moved to the right anterior of the aorta at the level of the kidney hilus. Hilar dissection showed absence of tumor thrombus in the left renal vein. Left renal vessels and ureter were ligated and the entire kidney mobilized outside Gerota's fascia, saving left adrenal gland. An extended

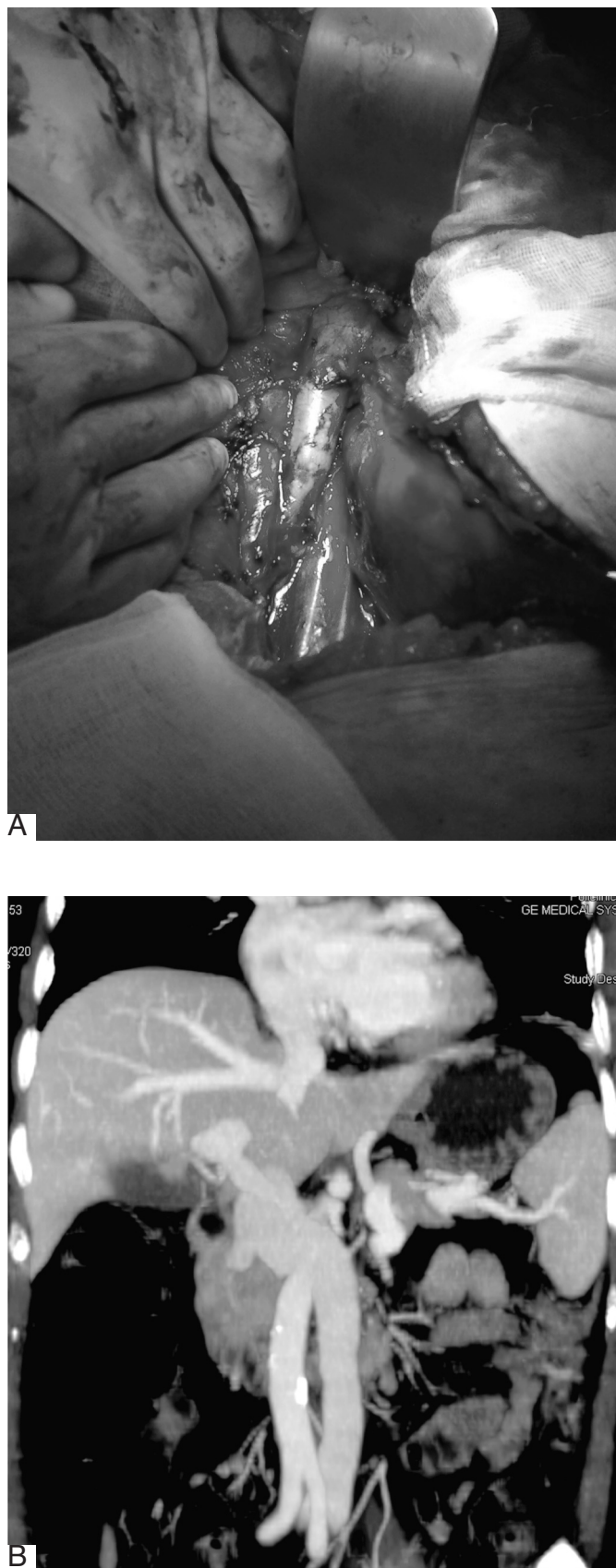


Fig. 1: A) Intraoperative image of infrarenal left vena cava. B) Postoperative chest and abdomen CT-scan with coronal maximum intensity projection reconstruction shows the total course of inferior vena cava.

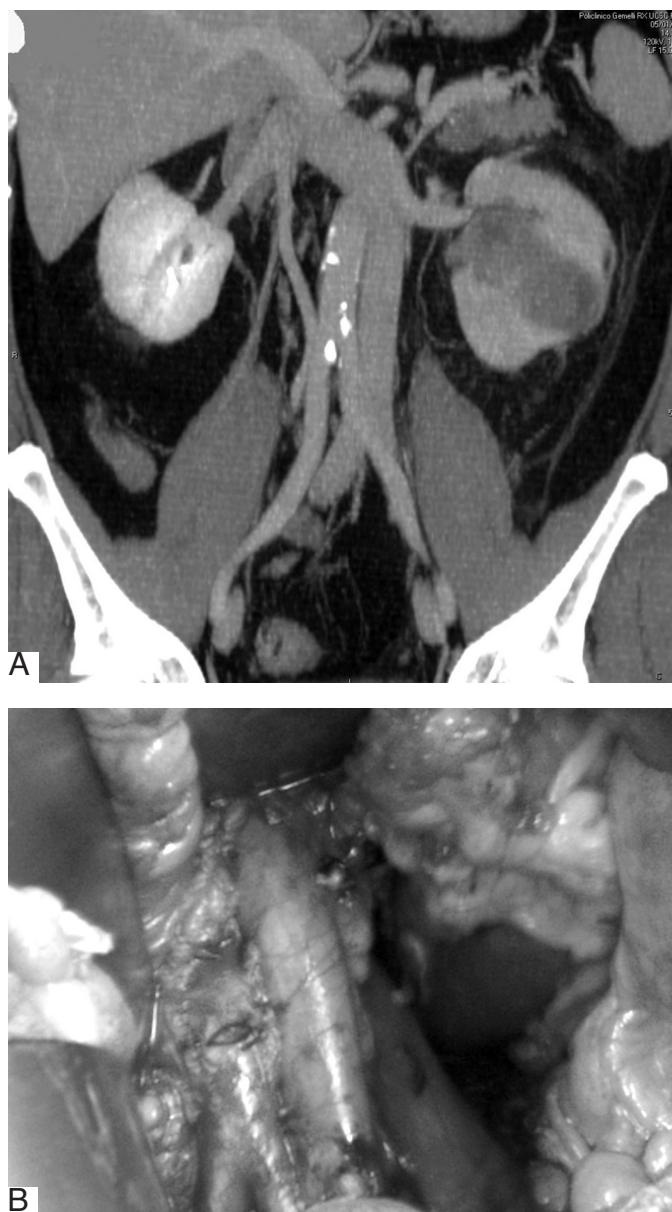


Fig. 2: A) Preoperative abdomen and pelvic CT-scan with coronal maximum intensity projection reconstruction shows the neoformation involving the left kidney and a transposed left IVC. B) Intraoperative image of left inferior vena cava.

lymphadenectomy was performed. There were no postoperative complications, except vestibular vertigo.

Histopathological examination showed RCC, without lymph node metastases.

As the vena cava abnormalities represent a possible risk factor for deep vein thrombosis and they may suppose a more difficult approach to future radiological, interventional, or surgical procedures involving these structures, in the postoperative period we performed a chest and abdomen CT-scan to study the total vena cava course and other possible cardiovascular defects (Fig. 1b).

The second case is a fifty-four-year-old man admitted

for left inferior quadrant pain, fever, leucocytosis and increased creatinina levels, non responsive to ciprofloxacin therapy. Physical examination revealed no abnormalities. Abdomen and pelvic CT-scan with coronal maximum intensity projection reconstruction showed a 7 cm heterogeneously enhancing neoformation involving the left kidney with intraparenchymal hematoma and a transposed left IVC (Fig. 2a). The IVC moved to the right anterior of the aorta at the level of the kidney hilus. Patient underwent a left radical nephroureterectomy and extended lymphadenectomy (Fig. 2b). The surgical exploration confirmed the presence of tumor involving the left kidney and the IVC malformation. There were no postoperative complications. Histopathological examination showed RCC, without lymph node metastases. There was no abnormality and no recurrence reported in both cases in the late follow-up.

Discussion

IVC anomalies are caused by aberrant development during the seventh to eighth week of gestation^{1,2}. The embryology of IVC is complex; it involves the formation, regression, and fusion among three longitudinal pairs of veins. The normal IVC is composed of four main segments in a caudal direction: hepatic, suprarenal, renal, and infrarenal segments. The hepatic segment is derived from vitelline vein. The right subcardinal vein develops into the suprarenal segment. The renal segment develops from right suprasubcardinal and postsubcardinal anastomoses. The infrarenal segment derives from right supracardinal vein¹⁻⁷. Because of the many transformations that occur during the IVC formation, anomalies in its final form may occur. Left IVC, embryologically, is formed by the regression of the right supracardinal vein and persistent left supracardinal vein. Such anomalies occur in 0.3% of otherwise healthy individuals and in 0.6–2% of patients with other cardiovascular defects⁸⁻¹⁰. Lately, IVC anomalies have been recognized as a possible risk factor for deep vein thrombosis, particularly in young adults. Gayer and co-workers described patients with an inferior vena cava anomaly who presented with widespread deep thrombosis of the pelvic and femoral veins, suggesting an interaction between the vascular anomaly and thrombophilia in the pathogenesis of deep thrombosis⁸. The study retain that an IVC anomaly should be considered in young patients who present with deep vein thrombosis of the femoral and iliac veins. Chuang and co-workers classified the anomalies of the postrenal segment of IVC into four types: type A is persistent right posterior cardinal vein (retrocaval ureter), type B is persistent right supracardinal vein (normal IVC), type C is persistent left supracardinal vein (left IVC), and type BC is persistent right and left supracardinal veins (double IVC)⁶. Many examples are reported in literature of different and variously associat-

ed congenital IVC anomalies. Kumar et al. presented a patient with renal cell carcinoma of left kidney with double IVC (DIVC) and tumor thrombus involving left renal vein and both IVCs; in this case a vascular abnormality was suspected after preoperative CT-scan and confirmed by MRI¹⁰. Guray et al. described a case of a left-sided IVC communicating with the hemiazygos vein which is draining into the dilated coronary sinus via persistent left superior vena cava pointing out the importance of identifying these anomalies to prevent difficulties in abdominal surgery, placement of caval filters, placement of temporary or permanent pacemaker to the right atrium and heart catheterization⁹. The left IVC typically ends at the left renal vein, which crosses anterior (or, more rarely, posterior) to the aorta in the normal fashion to join the right IVC. Most cases of left IVC are asymptomatic and diagnosed on cross-sectional imaging. Moreover, the anomaly mimics left-sided paraaortic lymphadenopathy, and may result in an inaccurate diagnosis, as in our case.

Riassunto

Descriviamo due pazienti con carcinoma a cellule renali del rene sinistro e con IVC a sinistra. Le anomalie congenite della vena cava inferiore (IVC) sono molto rare, estremamente variabili e rispecchiano la complessità dello sviluppo embrionale di questa struttura. Le varianti anatomiche devono essere differenziate da possibili patologie, in particolare "linfadenopatie periaortiche", in quanto la loro presenza può influenzare le procedure chirurgiche e interventistiche nello spazio retroperitoneale. L'individuazione preoperatoria di anomalie congenite IVC può ridurre la morbilità di interventi sullo spazio retroperitoneale. Una volta diagnosticata la malformazione, ogni cura deve essere presa durante l'intervento per evitare lesioni accidentali.

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