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ORIGINAL ARTICLE

Computerized tomography and 3-D rendering help to select surgical strategy in leiomyosarcoma of the inferior vena cava

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Abstract Leiomyosarcoma of the inferior vena cava is a rare tumor that is usually fatal. The tumor may grow very slowly or occasionally very rapidly, shows extensive local invasion, and metastasizes more frequently than previously believed. Complete surgical resection remains the only potential curative therapeutic option. The aim of this study was to report the clinical experience in the management of a patient with leiomyosarcoma. A 65-year-old woman with a history of vague abdominal pain and leg swelling underwent computed tomography which demonstrated an occlusion of the inferior vena cava. The patient received a complete excision of the tumor without reconstruction and histological analysis confirmed the diagnosis of leiomyosarcoma type 1. At 3 years, the patient is still doing well with minimal leg edema and a contrast-enhanced CT demonstrates no evidence of recurrence locally or in distant sites. Leiomyosarcoma is a rare and aggressive tumor that presents with non-specific symptoms. Computerized tomography with 3-D reconstruction is a useful tool to define the presence and entity of the collateral circulation and therefore to decide on the surgical strategy. Resection probably offers the best opportunity for long-term survival.

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Introduction

Primary leiomyosarcomas of the inferior vena cava (LIVC) are rare tumors with only under 400 cases reported in the literature [1–4]. As reflected in case reports and small series, they are malignant with variable biological behavior. Some of them allow survival of several years and others cause demise within a short period with widespread metastasis. The mainstay of treatment is wide surgical excision of the cava and the associated lesion. The technical approach varies according to the location of the tumor. From this standpoint, LIVC may be classified into three distinct types: type 1 involves the infrarenal segment of the cava (35 %); type 2 are in the pararenal vein position (45 %); and type 3 are located in the retro-hepatic inferior vena cava up to—and occasionally entering—the right atrium (20 %) (Fig. 1) [5, 6].

In type 2 and 3 tumors, the vena cava must be reconstructed with a suitable conduit, and reimplantation of one or both renal veins or nephrectomy may be necessary. In type 3, extracorporeal circulation and circulatory arrest is often required. In type 1, the need for reconstruction after excision is debatable with an international registry reporting good results in 25 % of patients in whom no reconstruction was carried out [7]. The crucial issue, however, is to decide which patient benefits from reconstruction and which does not.

We report one case of LIVC treated with excision and no reconstruction, and describe how embryological considerations and anatomic information from computed tomography and 3-D rendering technique were helpful in deciding the surgical strategy.

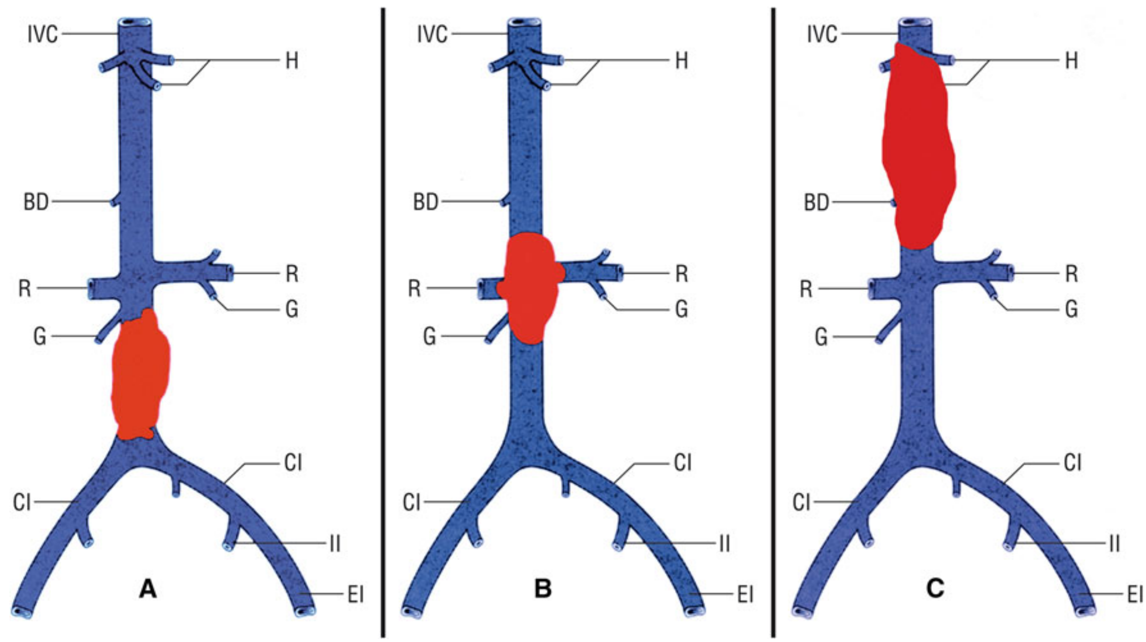

Fig. 1 Topographic classification of the leiomyosarcomas of the inferior vena cava. Type 1 (**a**); Type 2 (**b**); Type 3 (**c**). *IVC* inferior vena cava, *H* hepatic veins, *BD* body wall veins, *R* renal vein, *G* gonadal vein, *CI* common iliac vein, *II* internal iliac vein, *EI* external iliac vein

Methods

A 65-year-old woman presented to our observation with 1-year history of vague abdominal pain and leg swelling of 6 months' duration. Her past medical history was negative; she had no previous surgery and was not a smoker. One year earlier, she had an abdominal ultrasound to rule out colelithiasis, which was reported as negative. Non-invasive evaluation of the lower extremities with duplex scanning showed normal flow in the venous system without evidence of deep venous thrombosis. Computerized tomography (64-MSCT, Somatom Sensation, Siemens Medical Solutions) with contrast enhancement demonstrated a tumor of the inferior vena cava causing total obstruction of the lumen with large collateral veins through the gonadal system (Fig. 2). Multi-planar reconstruction (MPR) along the coronal plane defined the upper edge of the tumor, which lay below the renal veins (Fig. 3). Preoperative risk assessment included an ECG, which was normal, and an echocardiogram, which demonstrated a normal ejection fraction with normal chamber dimensions and no valvular disease. Based on CT findings, surgery was planned and carried out with simple excision of the cava below the renal and cranial to the common iliac veins. A midline incision was used for laparotomy. The postoperative course was uneventful and the patient was discharged home 8 days after surgery.

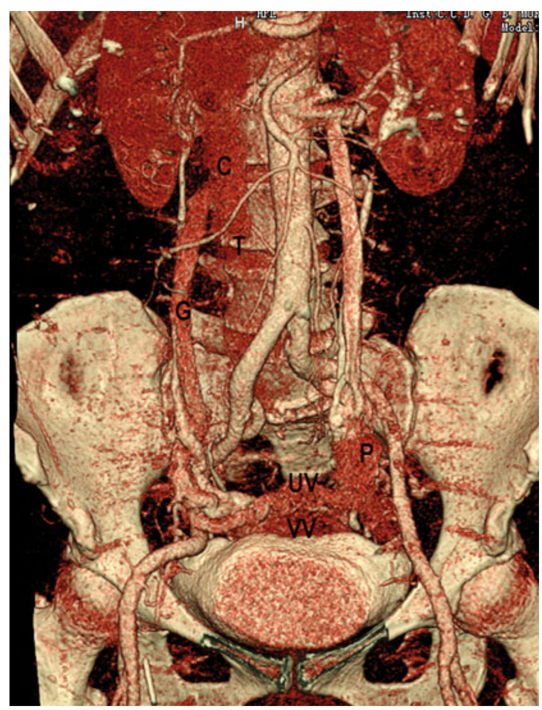

Fig. 2 Computed tomography and 3-D rendering image. Computed tomography and 3-D rendering image demonstrate large collateral veins stemming from the pelvic and gonadal system (*G*) and draining through the gonadal veins in the infrarenal vena cava (*C*) on the *right* and in the renal vein on the *left*. Leading edge of the tumor (*T*); pampiniform plexus (*P*); utero-vaginal (*UV*) and vesico-vaginal (*VV*) plexus

Fig. 3 MPR reconstruction image. MPR reconstruction along the coronal plane demonstrating the tumor within the inferior vena cava and caudal to the renal veins. The *arrow* points to the proximal leading edge of the tumor

Fig. 4 Computed tomography at 3 years. The *arrows* point to a large gonadal vein which runs parallel to the aorta near the native cava which has been removed

Results

Histological analysis demonstrated interlacing fascicles and whorls of cellular smooth muscle fibers with some large, bizarre, hyperchromatic nuclei. Mitoses were not common and ranged from 2 to 3 per high power field. The involved segment of the vein wall was diffusely replaced by malignant tumor. The diagnosis was leiomyosarcomas. At 3 years, the patient is still doing well with minimal leg edema and a contrast-enhanced CT demonstrates no evidence of recurrence locally or in distant sites. Furthermore, the large gonadal system seems to have replaced the function of the inferior vena cava (Fig. 4).

Discussion

Several studies show that multi-slice computed tomography (MSCT) is reliable and accurate in defining arterial and venous anatomy and pathology. The 3-D information of the cardiovascular system allows identification of even the most unusual pathology in the coronary circulation [8–10], which is traditionally difficult to study. We have already reported on a rare anomalous communication between the left coronary artery and the pulmonary artery diagnosed with MSCT [11].

Diagnosis and treatment of sarcomas of the vena cava have relied until now on angiography as the gold standard [4–6]. In our patient, we used computerized tomography as the only diagnostic tool. It was performed with 64 × 0.625 mm detector collimation, 120 kVp and 180 mAs, and data were reconstructed using 0.75-mm slice thickness and 0.5-mm interval. Visualization of the infrarenal vena cava with contrast-enhanced CT may be challenging, since contrast is diluted by the venous return from the pelvis and lower extremities. We used 150 ml of Ultravist 370 (Bayer) injected via an antecubital vein at a rate of 4.0 mL/s and obtained delayed scans 45 s after the dye injection. Data were then sent to a freestanding workstation (Wizard, Siemens Medical Solutions) where multiplanar and 3D images were generated. This technique allowed us to obtain a thorough visualization of the inferior vena cava and pelvic circulation.

In all inferior vena cava tumors, a reconstruction of the vena cava seems a logical course of surgical treatment. While this is true for type 2 and 3 LIVC [7], we feel that in those localized to the infrarenal position a reconstruction is not always necessary or desirable. The reasons are that the synthetic prosthesis is often occluded when used to replace the inferior vena cava; surgical intervention is prolonged when the cava is removed and reconstructed, increasing the risks of surgery; and a rich collateral circulation around the inferior vena cava already exists when the tumor occludes it. This collateral circulation can be carefully evaluated with computerized tomography, which helps in selecting those patients who benefit from reconstruction from those who do not.

In this work we present a type 1 LIVC treated with simple excision and no reconstruction of the inferior vena cava. This surgical strategy was chosen because of the large collaterals, which were demonstrated with computed tomography, and because of anatomic and embryological considerations related to this organ. Furthermore, reconstruction of the infrarenal vena cava adds complexity to the surgery and is associated with a patency rate of 50–65 % at 5 years [12–15].

Embryological considerations

At 4 weeks, the embryo has three main venous systems: the omphalomesenteric veins (or vitelline veins), which carry blood from the yolk sac to the sinus venosus; the umbilical veins, which originate in the chorionic villi and carry oxygenated blood to the embryo; and the cardinal veins, which drain the body of the embryo proper. The cardinal system consists of the anterior (or superior) cardinal veins, which drain the cephalic part of the embryo and the posterior (or inferior) cardinal veins, which drain the rest of the embryo. The anterior and posterior veins join to form the short common cardinal veins and enter the sinus horn.

During the fifth week, another two venous systems appear: the subcardinal veins, which drain the mesonephros and the supracardinal veins, which drain the body wall. They run parallel to and posterior to the posterior cardinal veins. The subcardinal veins connect with the posterior cardinal veins through multiple anastomoses. The sacrocardinal veins, which drain the lower extremities, are also formed during this week.

The formation of the IVC occurs between the sixth and eighth weeks of gestation. The anastomosis between the subcardinal veins in part atrophy and the one remaining forms the left renal vein. When this communication has been established, the left subcardinal vein disappears, and only its distal portion remains as the left gonadal vein. The right subcardinal vein becomes the main drainage channel and develops into the renal segment of the IVC. The infrarenal segment of the cava is formed by the supracardinal veins. The anastomosis between the sacrocardinal veins forms the left common iliac vein. The right sacrocardinal vein becomes the sacrocardinal segment of the IVC. The right vitelline vein forms the hepatic segment of the IVC; the renal segment of the IVC connects with the hepatic segment, and the IVC is complete. The IVC therefore consists of multiple embryological anlagen including hepatic, renal, and sacrocardinal veins (Fig. 5).

From a biologic standpoint, it is conceivable that in a system with a complex embryological development,

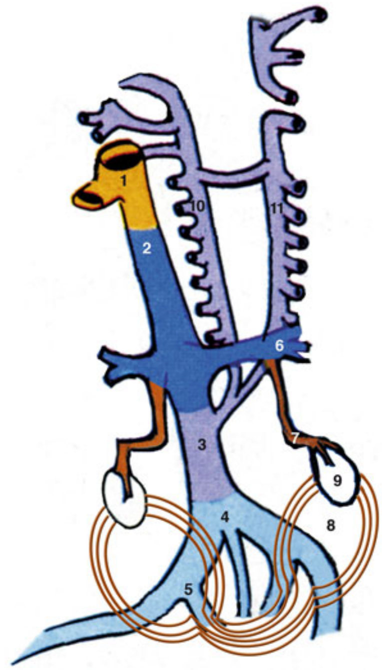

Fig. 5 Schematic representation of the different anlagens forming the inferior vena cava. *1* vitelline component, *2* subcardinal segment, *3* supracardinal segment, *4* posterior cardinal segment, *5* internal iliac vein, *6* renal vein, *7* gonadal vein, *8* utero-vaginal and vesico-vaginal (VV) plexus, *9* ovary, *10* azygos vein, *11* hemiazygos vein

multipotent mesenchymal cells may be present or trapped and develop malignant potential later on in life [16–20].

Anatomic and hemodynamic considerations

Vascular conduits are living tissues and are known to respond to hemodynamic stimuli in a compensatory fashion. For example, a stenosis in an arterial system is accompanied by overall enlargement of the artery so that the stenosis may persist without perceivable reduction in overall lumen and therefore volume flow. These adaptations are mediated through known (locally produced epithelial growth factor) and unknown metabolic pathways. Likewise, in the venous system, the occlusion of a major vein may be compensated by the hypertrophy of collateral systems, which may carry a blood glow similar to the one originally occluded. This seems to have occurred in our patient in whom the gonadal system has developed to a point where no significant symptoms of leg swelling have occurred in spite of the occlusion and then the removal of the inferior vena cava.

Biological considerations

The biological behavior of the tumor in our patient was locally malignant with infiltration of the wall of the cava, but histologically the relatively low number of mitoses per HPF suggested a slow growing malignant tumor. This characteristic was probably associated with a very slow occlusion of the vena cava, giving the time for the development of a significant collateral system and therefore causing only minimal leg swelling. The relatively benign course was confirmed also by the natural history of the disease after the resection, which after 3 years is not associated with recurrence.

Turning back the clock of organogenesis

The tumor in this patient had occluded the entire lumen of the inferior vena cava in its segment below the renal and above the common iliac veins. This corresponds to the segment of the cava embryologically derived from the right subcardinal vein. There are two considerations borne out from this. The first, as we mentioned earlier, pertains to a possible histopathologic explanation of the reason why the LIVC are more common in this venous district: it is most likely that in an area with a complex organogenesis, embryonic cells remain and develop oncogenetic potential later on in life. The second allows explaining the mechanics of development of the collateral circulation. The abdominal venous system is predisposed to compensate for occlusions of one or more segments. If the occlusion is above the renal veins, the azygos system may hypertrophy and shunt blood in the superior vena cava. If the obstruction of the cava is below the renal veins, the gonadal system represents the main collateral pathway. This is clearly illustrated in our patient (Fig. 2). The pampiniform gonadal plexus, which drains through the gonadal vein on the right in the cava, and on the left in the left renal vein, represents the collateral pathway in obstructions of the inferior vena cava below the renal veins. In this circumstance, the flow of blood in the pampiniform plexus reverses its direction and drains in the internal iliac veins through the utero-vaginal and vesico-vaginal plexus. The right and left gonadal veins therefore assume the function of the inferior vena cava. Teleologically, we may say that an occlusion of the infrarenal vena cava turns back the clock of organogenesis and the gonadal veins resume the role of main venous drainage of the lower body as the posterior cardinal veins (from which they originate) did at 4 weeks of embryonic life.

Conclusions

In conclusion, primitive leiomyosarcomas of the vasculature are rare. The most common location is the inferior vena cava and we hypothesize that this is related to the complex origin of the adult vena cava and to the possibility that in this environment there is a larger number of unstable cells. In type 1 LIVC, vascular reconstruction is not always necessary, particularly if both gonadal veins are patent. Finally, computerized tomography with 3-D reconstruction is a useful tool to define the presence and entity of the collateral circulation and therefore to decide on the surgical strategy.

Conflict of interest The authors declare that they have no conflict of interest.

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