Large retroperitoneal sarcoma invading the inferior vena cava successfully resected.

Technical notes of two cases

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Retroperitoneal sarcomas are rare neoplasms . They frequently reach a very large size and invade adjacent organs before they are detected. Involvent of the inferior vena cava is uncommon. Distant metastases are a late feature. The mainstay of treatment is compartmental resection and contiguous organ resection.
We report two cases of right-sided massive primary retroperitoneal leiomyosarcoma in pauci symptomatic women. In both cases treatment consisted of radical surgery. En bloc resection of the tumor and surrounding tissues and organs as well as part of the right wall of the subrenal IVC. To close the wall defect direct suture repair was used resulting in a reduced caliber but no hemodynamic sequelae or endoluminal thrombi. All the resection margins, including the inferior vena cava wall, were negative. The postoperative course was unremarkable and caval blood flow was optimal. The current gold standard treatment for retroperitoneal sarcoma is en bloc multivisceral resectionresection.

KEY WORDS: Peritoneal sarcoma, Surgery, Vena cava

Introduction

Retroperitonal sarcomas (RPS) are rare malignant tumors. The most common types of retroperitoneal sarcoma are leiomyosarcoma followed by liposarcoma. RPS make up 1-2% of all solid tumors [1,2]. These tumors are have hematogenous spread and are locally aggressive and local recurrence rates after resection are often high These tumors which arise from soft tissues in the retroperitoneal space often reach a large size without causing symptoms or causing non specific symptoms and therefore diagnosis is delayed. As they grow they press on/invade surrounding organs. On the right they displace/invade the kidney, adrenal gland, liver, gallbladder, duodenum, right colon and, rarely, vertebrae and blood vessels, as well as the corresponding organs and tissues on the left and the tail of the pancreas Diagnosis is usually made by computed tomography (CT) scan and magnetic resonance imaging (MRI) to determine the extension of the tumor and its characteristics, what organs are involved and whether there are distant metastases. Preoperative percutaneous core needle biopsy of the mass is a useful adjunct as is fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (18FDG-PET-CT) scan which helps determine area should be biopsied.
We report two cases of RPS. CT scan ad MRI showed right-sided RPS that originated from the subrenal portion of the IVC or from soft retroperitoneal tissue and invaded surrounding organs and tissues, Postoperative histology of the resected IVC w Case all confirmed the diagnosis.

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Case Reports

Case N. 1

A 51-year-old woman, presented complaining of epigastric discomfort for 2 months, refractory to medical treatment. She was otherwise fit and healthy,gravida 2 para 2,with no known comorbidities. She had undergone hysterectomy and salpingo-oophorectomy for uterine fibroids 5 years prior, and had no other significant medical/surgical history, and no history of trauma Total body CT scan revealed a large mass on the rightside of the abdomen This finding was confirmed by MRI.
CT scan with contrast and MRI were performed preoperatively (Fig. 1): There was a large retroperitoneal mass with sarcomatous features: areas of hemorrhage and necrosis and calcificatios. The heterogeneous mass was located on the right and extended from just below the diaphragm to the iliac crest, invading the inferior vena cava. The mass measured 25 × 12 × 15 cm. It was strongy adherent to the right crus of the diaphragm, the right renal vein, the liver hilum, and the right portal vein which appeared noticeably narrower than normal. The mass appeared to be inseparable from the parenchyma of the right lobe of the liver, especially at the fourth segment and encased the right adrenal gland. Morever, it displaced medially the gastric antrum, the duodenum and the head of the pancreas. There was posterior displacement of the kidney as well as caudal displacement to the level of the right iliac fossa and close to the mass there where newly formed veins. The spleen. pancreas and left adrenal gland were normal There was fluid in the pelvic cavity. The urinary bladder was normal,. There were left paraaortic lymphnodes 1 cm in diameter. There was a posterior subpleuric nodule 1 cm in diameter in the inferior lobe of the right lung
Results of full blood count and blood chemistry tests were normal with a hemoglobin level of 11,5 g/dl. The patient had no electrocardiogram abnormalities

Surgical Procedure

With the patient under general anesthesia, midline laparotomy extended to the right hypochondriac region with a transverse incision was performed.
There was a large retroperitoneal mass, 25 × 10 × 15 cm ,filling the right perirenal space, displacing the kidney caudally and the liver cranially. Inseparable from the visceral surface of segments V-VI (Fig. 2) The mass was carefully dissected free while preserving the kidney and its vessels The undersurfaces of segments V and VI was resected together with the gallbladder. Clamping of tthe liver peduncle and the subrenal IVC, apparently invaded laterally by the tumor, was followed by complete mobilization of the tumor mass. After longitudinal clamping, excision of the affected portion of the IVC wall, 5cm long and 1.5 cm wide, was performed (Fig.3) The IVC wall defect was repaired with continuous 5.0 prolene suture. The mass was removed en bloc togeher

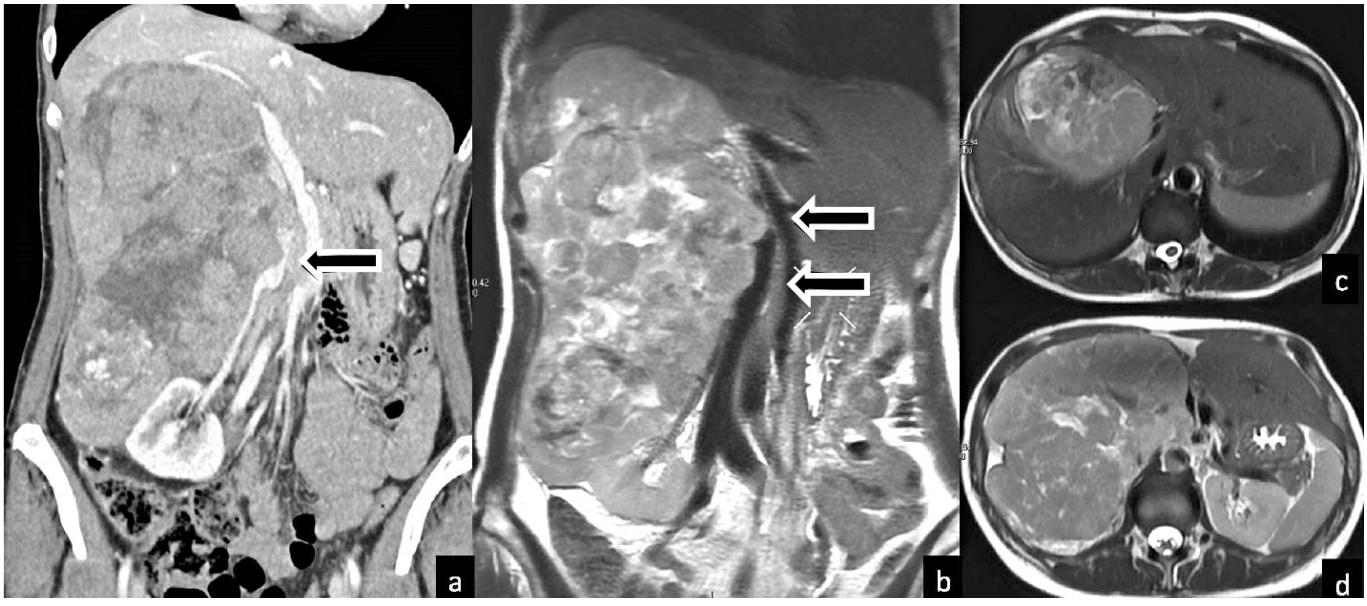

Fig. 1: Contrast-enhanced coronal CT image (a), and coronal (b) and axial (c and d) T2-weighted MRI images show a large well defined soft-tissue retroperitoneal mass on the right extending from immediately below the diaphragm to the level of the iliac crest,and infiltrating the IVC (arrows in a and b). The mass is composed of hemorrhagic, necrotic, and calcific areas without mascroscopic adipose tissue. It appears to be adherent to the right diaphragmatic crus, the right renal vein, and the right main branch of the portal vein, which is markedly thinned. The mass is indissociable from the hepatic parenchyma at the right lobe of the liver and engulfs the right adrenal gland. It displaces medially the stomach, the duodenum, and the head of the pancreas. The right kidney is displaced posteriorly and inferiorly.

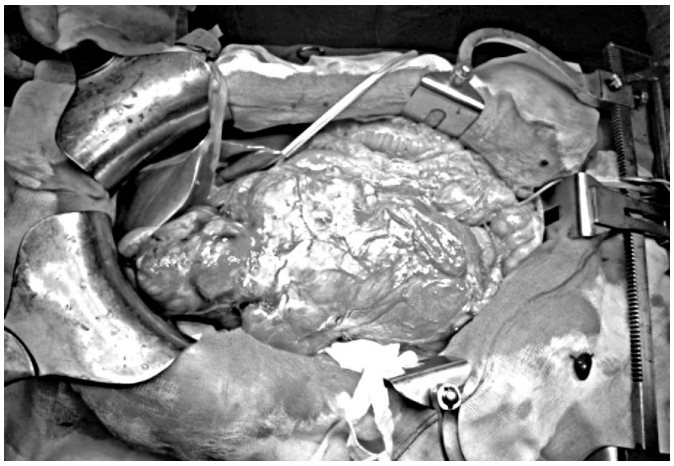

Fig. 2: Midline laparotomy extended to the right hypochondriac region with a transverse incision showing the voluminous retroperitoneal mass.

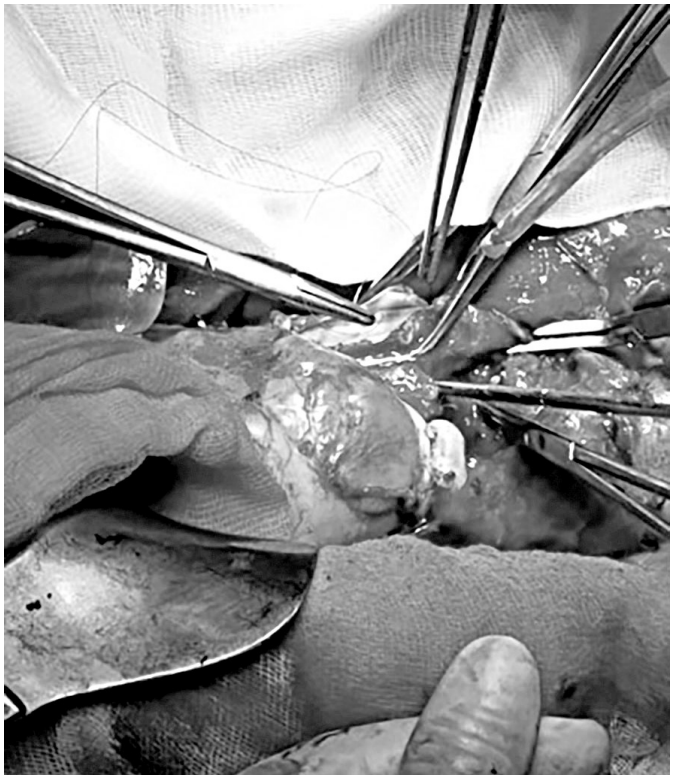

Fig. 3: Longitudinal clamping and resection of the IVC wall.

with the 2 subsegments of the liver, the gallbladder, the IVC wall., the right adrenal gland, and the perirenal fat. Metal clips were left to mark the resection margins,, especially along the IVC.
Hemostasis was obtained in part with suture transfixation. A drain was placed in the abdominal cavity .The abdomen was closed in layers. The patient had received a blood transfusion 3 units of packed red blood cels, 250ml each. The weight of the surgical specimen was ca. 1.5 Kg.

Intraoperative frozen section analysis of several fragments of the mass revealed mixed sarcoma with fibroadipose components
Postoperative course: was unremarkable. Low molecular weight heparin was administered for 30 days in scaled doses and followed by color Doppler of the IVC. On postoperative day 5 the drain was removed, bowel function returned and the patient started oral feeding. She was discharged on postoperative day 7.
Histopathology report stated: mesenchymal neoplasia with spindle cells, partly contained by a thin fibrous capsule formed of spindle cells in a fascicular pattern, which sometimes fuse with the vessel walls . The cells are characterized by fusiform nuclei with blunted ends and varying degrees of atypia, and eosinophilic cytoplasm with clear perinuclear vacuoles and poorly defined margins. There are areas of necrosis and hemorrhage. Mitotic activity is high (>20 mitoses in 10 HPF) with some atypical mitoses. The cells are strongly positive for vimentin, smooth muscle actin, and desmin, and weakly positive for CD 34 and negative for CD 117, citocheratin AE1/AE3 and S 100. The neoplasia is adherent to Glisson's capsule without invading it. And without extending into the liver parenchyma which is characterized by diffuse steatosis. There is chronic inflammation of the gallbladder with adenomyomatsis of the fundus. There is no neoplastic invasion of the adrenal gland. There is neoplastic invasion in the fragment of the IVC wall. Diagnosis: High-grade retroperitoneal leiomyosarcoma

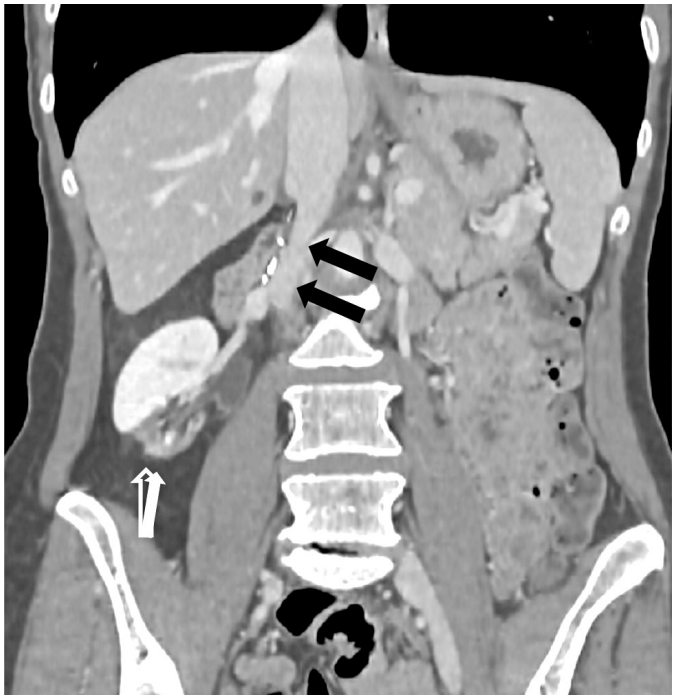

Fig. 4: Contrast-enhanced coronal CT image, CT scan performed 1 year after surgery, shows IVC clips (arrows). There is no recurrence. Areas of infarction are present at the level of the lower pole of the right kidney (white arrow).

(Federation Nationale des Centres de Lutte Contre le Cancer [FNCLCC] system), invading the IVC and compressing the liver parenchyma. Union for International Cancer Control (UICC) classification: pT 4.
Postoperative check-up (Fig. 4): One year after surgery the patient was in excellent health. CT scan did not show any signs of local recurrence or distant metastases. There were areas of infarction in the lower pole of the right kidney.

Case N. 2

A 69-year-old woman, otherwise fit and healthy, with no past medical or surgical history of note, and no history of trauma, presented complainig of the sudden onset of severe pain in the left flank and left hypochondriac region. She was admitted to hospital as an emergency because of suspected gallbladder disease.
Results of full blood count blood chemistry tests were normal, except for a low level of hemobglobin 10.4 g/dl The chest xray a—nd electrocardiogram were normal.
On physical examination the patient had a nondistended abdomen; normal movement with respiration. There was tenderness on palpation of the central and right-hand regions of the abdomen and an elastic mass was felt in the right hypochondriac and epigastric region.
Ultrasound, CT scan with contrast and MRI were performed (Fig. 5) and revealed a large solid abdominal mass that was contrast-enchanced peripherally with some peripheral areas of hypervascularization The mass measured 14 × 10 × 13 cm and extended from the second segment of the duodenum to the right hypochondriac region and right flank,and had a close relationship with the right colon laterally, the transverse colon anteriorly, both the renal vessels and the middle third of the ureter posteriorly, and medially with the IVC ,compressing it significantly from right to left. There was concomitant ectasia of the pelvic venous plexuses, the ovarian veins and the splenic veins, apparently because of the compression of the IVC. There was a moderate amount of hemorrhagic ascites. The patient had uterine fibroids.. The working diagnosis was duodenal gastrointestinal tumor (GIST).

Surgical Procedure

The procedure was performed with the patient under general anesthesia. A moderate amount of bloody fluid was found in the abdominal recesses; (300ml). There was a retroperitoneal mass fixed on the deepest plane, invading the right mesocolon and encasing the right colon and duodenum, with signs of bleeding. The mass filed the right half of the abdomen . Its cranio-caudal diameter was ca. 20 cm. The liver and gallbadder were not involved. The right paracolic gutter was opened. The anterior portion of the renal capsule was removed and the kidney was dissected bare up to the renal hilum. This was followed by mobilization of the right colic flex-

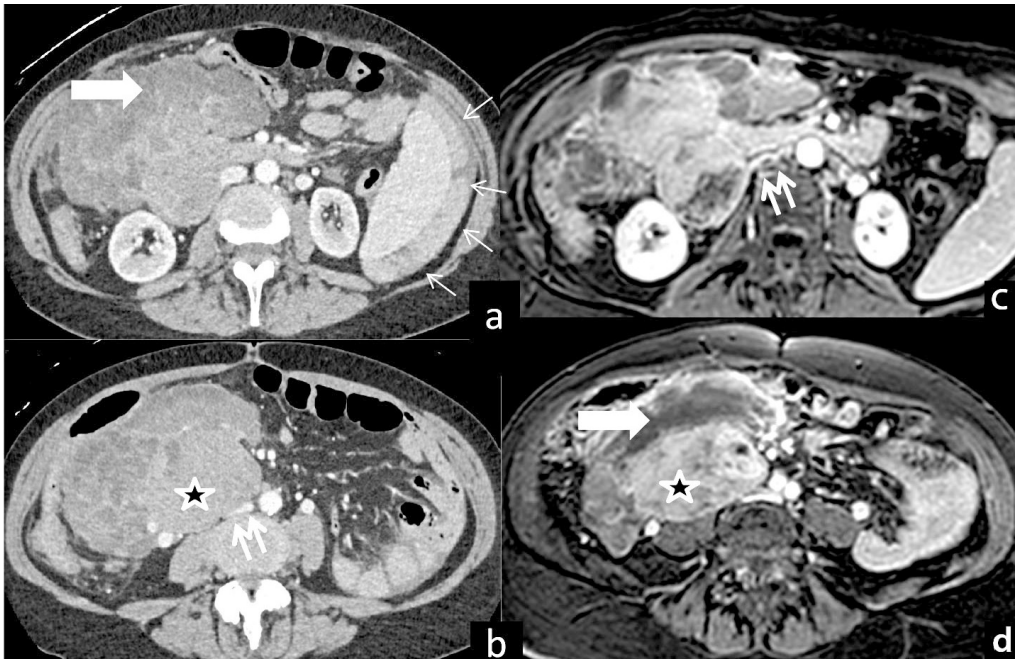

Fig. 5: Contrast-enhanced transverse CT image ((a) and (b)) and contrast enhanced fat saturation T1 MRI images (c and d) show a large mass at the level of the right hypochondriac region and right flank strongly adherent to the second portion of the duodenum and IVC, which appears to infiltrate the IVC (arrows) .In the mass there are solid components (asterisk in (b) and (d)) and areas of necrosis (arrow in (a) and (c)). The arrows in a point to hemorrhagic perisplenic fluid.

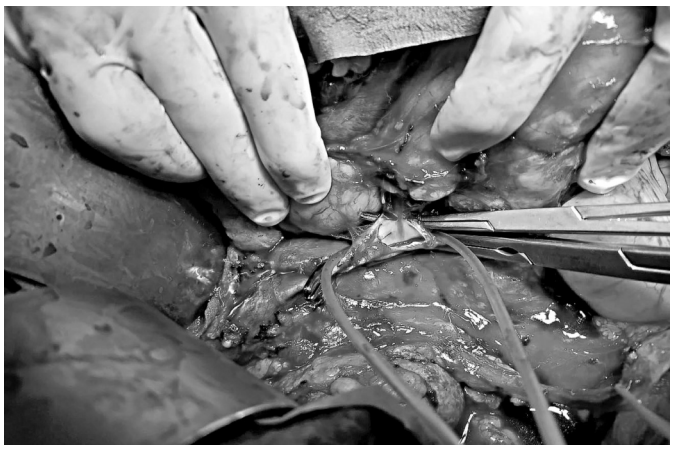

Fig. 6: Lateral clamping of the subrenal IVC and excision of a small portion of the vessel wall.

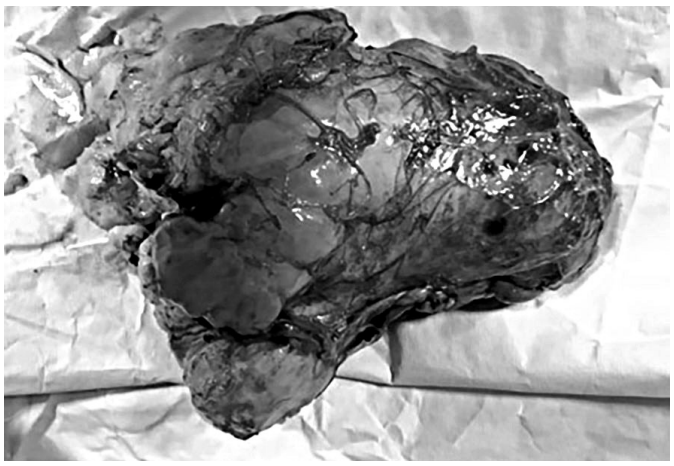

Fig. 7: The surgical specimen, which includes,the tumor in the right colon,a segment of the transverse colon, the last loop of the ileum, a piece of the duodenal wall and of the IVC. Total weight: 2.55 Kg.

ure and Kocherization of the duodenum, the inferior flexure of which semed inseparable from the solid most cranial part of the mass. The ovarian peduncle was encased in the mass. The ureter was isolated and completely freed. Dissection continued towards the ligament of Treitz and the duodenum compressed by the hemorrhagic mass. An en bloc right hemicoectomy, with the mass, was required: therefore the omental bursa was opened to prepare the transverse mesocolon, and the middle third was diveded, sparimg the middle colic pedicle. This manouever allows control of the middle colic vessels, the trunk of Henle, and the ileocolic pedicle which are divided between two ligatures. There is invasion of the duodenal flexure and tangential resection with TLC is required. The right colon and the last loop of the ileum are divided with TLC, completing the intestinal phase and right IVC plane is exposed. The distal ovarian pedical is divided between two ligatures and the IVC is prepared. The ovarian vein ostium is isolated and cut where it enters the IVC, and closed with prolene.. The mass is inseparable from the IVC for a from where the ovarian vein drains into the IVC to 2 cm distally. After lateral clamping of the subrenal IVC, a partial resection was performed, removing a piece of the vessel wall 4cm in diameter (Fig. 6). Then the mass was removed the IVC defect was repaired with prolene sutures. Hemostasis was achieved. Peritoneal washing was performed. Total blood loss was ca.1000ml and the patient received 2 blood trasfusions A mechanical ileocolic anastomosis was fashioned. The mesenteric defect was closed. After placement of two drains, subhepatic and retroanastomotic, the abdomen was closed in layers. The histopathology report stated: surgical specimen comprising the tumor, the right colon, a segment of the transverse colon, the last ileal loop, a fragment of the duodenal wall and one of the IVC wall. Total weight: 2,550 Kg (Fig. 7). Areas of low-moderate differentiation, with spindle cells, rare nuclear atypia and cystic degeneration with myxoid appearance. Typical of classical leiomyosarcoma, and areas of high-grade differentiation pleiomorphic, with spindle cells, poligonal cells and numerous giant multinucleate osteoclast-like cells. The neoplasia invades the tunica muscularis propria of a fragment of duodenal wall, the tunica serosa and subserosa of the small intestine, the tunica serosa and subseosa of the large intestine and the lateral wall of the IVC. Diagnosis: differentiated pleiomorphic leiomyosarcoma which invades the duodenum,ileum, right colon, transverse colon, and IVC.

The postoperative course was unremarkable.The patient was afebrile. Her bowel function returned on postoperative day 3 and she was discharged on postoperative day 8. Postoperative check-up: The patient was asymptomatic. CT scan and color Doppler ultrasound of the IVC showed no changes in blood flow or signs of local recurrence or distant metastasis

Discussion

Invasion of the IVC by a retroperitoneal tumor is rare and is not mentioned by many authors. Even the monograph by the Italian Society of Surgery (Società Italiana di Chirurgia) published in 1994 [4] which describes the invasive charateristics of these tumors and the structures affected, does not list the IVC.

Before the year 2000, other authors reported rare cases of RPS. Later surgery a new impulse was given to surgery of the IVC thanks the skills accquired by liver transplant surgeons. The group of Ghose and colleagues in India [5] reported 6 out of 37 cases of sarcoma operated on in 2015-2016, 4 of which were primary tumors of the IVC while 2 were cases of invasion of the IVC by primary retroperitoneal soft tissue tumors. In 3 of these 6 cases caval reconstruction with a vascular prosthesis was required, in 1 reconstruction was not required

because sufficient collateral circulation had developed, and in the otther 2 longitudinal vessel wall resection was performed with primary reconstruction.
In 2003 Grotemeyer and colleagues [6] described 6 cases of neoplastic invasion of the IVC treated with resection and caval reconstruction with a vascular prosthesis, In 2012 Fiore and colleagues at the The National Cancer Institute of Milan [7] reported 13 cases 9 of which required reconstruction with a prosthesis and 1 direct suture repair,. In 2020 Ruiz and colleagues published a large series of 52 cases of caval resection [8] in 17 of which direct repair of the caval defect was performed .According to Quinones-Baldrich and colleagues [9] primary reconstruction of the vena cava is well tollerated if the marrowing of the lumen does not exceed 50%, and moreover, narrowing of the lumen causes and increase in blood flow velocity and thus reduces the risk of thrombosis . Out of 37 cases of IVC surgery reported, 36 were for invasion by retroperitoneal sarcoma. Eleven of these 36 patients underwent primary reconstruction which was very well tollerated hemodynamically. After partial resection of the IVC wall the defect can be filled using pieces of autologous vein or synthetic material or autogenous peritoneo-fascia graft material as proposed by Chin and colleagues [10]. Surgical mortality essentially concerns segmental circular resections of the IVC, especially if associated with multivisceral excision and reimplantation of the renal vessels. In these extreme conditions mortality rates can reach 15% [11].
The completenes of caval resection, confirned by negative margins, is the most important prognostic factor [12]. Similarly, the entire surface of the mass removed must have negative margins and can lead to compartmental excision to reduce the risk of recurrence [13]. Simply detatching the tumor from the surrounding tissues and organs does not constitute a radical oncologic resection[14]. From the foregoing it follows that, for RPS, given the current ineffectiveness of other therapy, surgery is the best treatment and should tend towards complete radicality even when that means compartmental excision with resection of the IVC if there is tumor invasion.
To achieve radicality in our first case the large tumor was removed along with the gallbladder, part of segments V and VI of the liver to which the tumor was fixed, as well as the homolateral adrenal gland. In our second case radicality required resection of a small piece of the second part of the duodenum as well as the right colon. In both cases partial longitudinal resection of the subrenal inferior caval vein was necessary since the tumor mass was tightly adherent to the vessel wall, which histologic examination showed was due to neoplastic invasion.
At 1. year follow-up CT scan showed that the first patient, young, slim, of medium height, had no locoregional recurrence or distant metastases, and although the caliber of the IVC was small there were no thrombi or other obstruction. However invasion of the IVC is a negative prognostic factor as demonstrated by the studies by Mingoli and colleagues in Italy in 1992 [15] and Dangoor ad colleagues in the United Kingdom in 2016 [16]. In the second patient follow-up at the time of writing was only 3 months and therefore too short for an oncologic evaluation but long enough to evaluate the patency of the IVC and note the absence of thrombi or hemodynamic disturbances distally.
Overall, in patients with retroperitoneal sarcoma treated with radical surgery, the survival rate at 5 years is ca. 50% and at 10 years 35% [17]. Careful postoperativr follow-up makes possible timely reintervention to remove local recurrence and provides good chances of long-term survival [18]. Pre-, intra- and postoperatve radiotherapy does not modify the prognosis [19].

Riassunto

I Sarcomi retroperitoneali sono tumori rari. La loro caratteristica è quella del grande sviluppo locale con invasione degli organi adiacenti e solo tardivamente la metastatizzazione a distanza. Eccezionalmente infiltrano la vena cava inferiore. La terapia è quasi esclusivamente chirurgica compartimentale con interventi estesi agli organi circostanti.
Vengono presentati due casi di due donne affette da voluminosi leiomiosarcomi primitivi retroperitoneali destri paucisintomatici nei quali l'exeresi della neoplasia, allo scopo di ottenere una radicalità oncologica, comportò la resezione in blocco del tumore con tessuti ed organi circostanti e di una parte della parete destra della vena cava inferiore sottorenale. La perdita di sostanza della vena cava fu riparata con sutura diretta dei margini e conseguente riduzione del calibro, ma senza conseguenze emodinamiche nè trombi endoluminali. I margini di resezione delle neoplasie compresa la porzione cavale risultarono istologicamente esenti da invasione neoplastica e le pazienti ebbero un decorso post operatorio senza complicanze e con un ottimo flusso ematico cavale .Le resezioni multiviscerali comprese quelle della vena cava, in blocco con l'asportazione del tumore primitivo, costituiscono oggi il gold standard nella terapia dei sarcomi retroperitoneali.

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