Primary leiomyosarcoma of the inferior vena cava: a case report

Leiomyosarcome primaire de la veine cave inférieure : à propos d’un cas

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Abstract. Primary leiomyosarcoma of the inferior vena cava (IVC) is a rare malignant tumor originating from the vein smooth muscle. We present one case of primary leiomyosarcoma of the IVC. The patient benefited of surgical exploration at seventh day after admission. Tumor located in the junction of the anterior wall of the IVC and the left and right renal vein. We carried out the tumor resection, vena cava artificial vascular patch prosthetics. The patient did not take anticoagulant drugs after surgery and was discharged at 12 days after surgery. Currently, the patient had survived for nearly six months, repeated abdominal computed tomography examinations showed no clear recurrence.

Key words: leiomyosarcoma, inferior vena cava, treatment

Résumé. Le leiomyosarcome primaire de la veine cave inférieure est une tumeur maligne du tissu musculaire lisse veineux. Nous présentons ici un cas clinique de cette pathologie rare. Après le diagnostic, le patient a été traité chirurgicalement de sa tumeur localisée à la jonction de la veine cave inférieure et de l’artère rénale. Le patient est sorti 12 jours après l’intervention, sans traitement anticoagulant ; les contrôles par imagerie (scanner) n’ont pas montré de récidive sur une période de 6 mois après le diagnostic initial.

Mots clés : leiomyosarcome, veine cave inférieure, traitement

Primary leiomyosarcoma of the inferior vena cava (IVC) is a rare tumor that originates from the smooth muscle tissue. Although many tumors of the IVC develop secondarily to a primary tumor elsewhere, few cases of primary tumor of the IVC have been reported, indicating that this condition has a very low incidence. We present one case of primary leiomyosarcoma of the IVC.

Case report

The patient was a 42-year-old man that presented with more than two month’s history of an intermittent dull pain, located in the right upper abdominal quadrant. The type-B ultrasonic of out-patient showed the substantive holder of the IVC. General physical examinations on admission were no clear positive signs; there was no particularly relevant past medical history. Preoperative abdominal enhanced computed tomography (CT) and chest and abdominal angiography examinations showed an oval low-density block shadow of hilum. The shadow of hilum was $2.3 \times 2.9$ cm, uneven density, sustained slight enhancement. The boundaries between the shadow of hilum and the portal vein, the IVC were unclear. A part of the lesion invaded the IVC and the IVC filled with defects (figure 1). The hepatic artery and portal vein were not obvious abnormalities. CT diagnosis considered intravascular (outer) skin tumors. Our hospital positron emission tomography (PET)/CT showed that irregular low density shadow located at the area of in front of the IVC, near the head of pancreas and slightly increased uptake of radioactivity. The result of PET/CT showed that the lesion originated from mesenchymal tissue. After improving preoperative examination, we basically excluded the secondary tumors. The patient was performed with surgical exploration at seventh day after admission, with the anti-“L” type incision in the abdomen. After routinely exploring the abdominal cavity, we excluded the secondary tumors. Hilum and mesenteric root lymph nodes were not swollen. We fully freed hepatic inferior vena cava. Tumor located in the junction of...
the anterior wall of the IVC and the left and right renal vein. The diameter of tumor was about 2.5 cm. The tumor was hard, hoar, enveloped. The tumor extruded the blood vessels to the area between the lower section trunk of the portal vein and the vena cava, grew to the vena cava. The tumor did not exhibit any adhesion with surrounding tissues and not violated posterior vena cava. Then, we carried out tumor resection, vena cava artificial vascular patch prosthetics. We used the Brandt clamp blocking the left and right renal vein, and vena cava clamp blocking the superior and inferior vena cava. We cut the IVC and removed the tumor completely with the wall of the IVC. The anterior wall of the IVC formed about 2 × 3 cm oval defect. We took a period of artificial blood vessel graft, trimmed into an oval patch with a considerable defect. We continuous sutured the patch and the IVC with a 4-0 prolene line. After anastomosis, we opened up the blocked blood vessel. Repairing section of the IVC filled good. No oozing in repairing section was observed when we appropriate pressured. The patient recovered well after surgery. Serum creatinine level was slightly increased in second day after surgery, then decreased gradually, and returned to the normal level in the fifth day after surgery. The patient did not take anticoagulant drugs after surgery and was discharged 12 days after surgery. Postoperative pathology showed the IVC leiomyosarcoma (figure 2). Currently, the patient had survived for nearly six months; repeated abdominal CT showed no clear recurrence.

Discussion

Primary leiomyosarcoma of the IVC is a rare malignant tumor originating from the vein smooth muscle. Primary leiomyosarcoma of the IVC often occurs in the middle and inferior segment of the IVC. Primary leiomyosarcoma of the IVC may grow to the extravascular and blood vessel, the former is more common. The distant metastasis organ of primary leiomyosarcoma of the IVC can be liver, lung and bone, but the transfer rate is usually less than 50%. Primary leiomyosarcoma of the IVC are growing slowly and, because of their retroperitoneal location, often present later and insidiously with nonspecific symptoms depending on the location, size, and growth rate of the tumor, such as abdominal pain, lower extremity edema, liver and kidney dysfunction, and weight loss. Primary leiomyosarcoma of the IVC is difficult to detect early, and could not undergo surgery because of advanced stage at the time of diagnosis [1]. The prognosis of patient with primary leiomyosarcoma of the IVC is poor [2, 3]. Surgical resection rate was 40%-60%. Recurrence rate was 36%. The 5-year survival rate was only 30% [4]. The patient had no specific symptoms, the placeholder was only found in physical examination, tumor markers AFP, CEA, CA19-9, CA125 were negative. The occurrence of early distant metastasis is low; therefore aggressive surgical treatment is necessary and radical resection is the preferred surgical approach [5-8]. For this
Current practice

Purpose, surgeries often require the removal of a part of the IVC and violated adjacent tissues, then reconstruct, repair or anastomosis the vascular. The IVC reconstruction often used artificial vascular graft. The tumor of this patient located in the junction of the anterior wall of the IVC and the left and right renal vein. The location is very special. The tumor was only violated the anterior wall of the IVC and the left and right renal vein was not violated. So the IVC artificial vascular patch prosthetics was carried out. During the operation, we used the Brandt clamp blocking the left and right renal vein about 30 minutes. Creatinine was slightly increased in second day after surgery, then decreased gradually, and returned to the normal level in fifth day after surgery. The result of our surgery suggests that the occlusion of renal vein in a short time (≤ 30 minutes) is safe. If surgery requires the reconstruction of renal vein, it is necessary to systematically assess preoperative renal function of patients.

Because of the special location of primary leiomyosarcoma of the IVC, primary leiomyosarcoma of the IVC were previously thought to be not sensitive to radiotherapy and chemotherapy. Furthermore, postoperative radiotherapy and chemotherapy has been reported that easily lead to graft thrombosis. So it is usually surgical resection alone. With the improvement of the artificial vascular graft technology, the incidence of artificial vascular graft thrombosis is significantly decreased. Some authors have reported that preoperative chemotherapy (the combination application of doxorubicin and ifosfamide) can help to reduce the size of tumor and ease removal of tumor; postoperative radiotherapy can consolidate the results of operations, reduce local recurrence and prolong the median survival time [9]. Therefore, for patients with leiomyosarcoma of the IVC, aggressive surgical resection, postoperative radiotherapy and chemotherapy are important to treat this disease and improve prognosis of patients. The patient was treated by the above combination chemotherapy. We need further follow-up to determine the prognosis of our treatment.

Conflicts of interests: none.

References


