Leiomyosarcoma of the inferior vena cava

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Key words: *leiomyosarcoma; inferior vena cava; radiological and pathological examination; surgical treatment.*

Summary. *Leiomyosarcoma is a rare tumor of mesenchymal origin usually affecting the inferior vena cava. Early diagnosis is essential before surgical resection, which is the only therapeutic modality that prolongs patients' survival. Ultrasonography, computer tomography, and magnetic resonance imaging are the main imaging modalities in this case. Combined with guided biopsies, they form the mainstay of reliable diagnosis. We report a case with retroperitoneal tumor arising from the middle segment of the inferior vena cava. Radiological examination revealed retroperitoneal tumor and helped to choose surgical treatment. Histopathological examination confirmed the diagnosis of leiomyosarcoma of the inferior vena cava.*

Introduction

Leiomyosarcomas are tumors of mesenchymal origin usually afflicting the retroperitoneum or uterus. Leiomyosarcomas of primary vascular origin are rare tumors, which arise mainly from the midsegment of the inferior vena cava (IVC) and are often detected in middle-aged women (1).

We present a case of leiomyosarcoma of the IVC and steps of radiological diagnosis of the case, together with surgical and histopathological correlation.

Case report

A 42-year-old woman complained of chest pain, cough, and body tremor. Autoimmune thyroiditis with thyroid insufficiency had been diagnosed 13 years ago. The condition deteriorated, thus a more detailed examination was undertaken.

Abdominal ultrasound (US) examination revealed a mass of 4.8×3.5 cm, extending beneath the porta hepatis and above the right renal artery, surrounding the IVC from the posterior-lateral aspect and narrowing its lumen. Differential diagnosis included a tumor of the IVC and right adrenal gland, metastases, or some other retroperitoneal tumors (Figs. 1A and 1B).

Contrast-enhanced abdominal computed tomography (CT) revealed a heterogeneously enhancing tumor involving the lateral aspect of the IVC, located superior to the right renal artery and deforming the right adrenal gland. Differential diagnosis included a tumor of the right adrenal gland and a tumor of the IVC (Fig. 2).

Abdominal magnetic resonance imaging (MRI) revealed that the mass adjacent to and involving the IVC possessed signs of necrosis. Intraluminal caval growth according to MR images was rather limited. The lateral aspect of the right adrenal gland was identified, but the precise origin of the tumor could not be established (Fig. 3).

Thus, surgical resection was considered. An irregularly shaped tumor of 7×5×4 cm in the region of the right adrenal gland was fixed to the superior pole of the right kidney and involved the IVC. The tumor, the right adrenal gland, and 11-cm caval segment were resected (Fig. 4A).

The vein was substituted by prosthesis, inserting both renal veins posterior to the abdominal aorta (Fig. 4B).

The final pathological diagnosis was leiomyosarcoma. Histopathological examination revealed a relationship between the IVC and tumor (Fig. 5A).

Microscopically the tumor was composed of the areas of high cellularity with middle size cells possessing prolonged, cigar-shaped nuclei exchanging with no cellular areas (Fig. 5B). Immunohistochemical reactions were positive for actin, only 15% of all tumor cells were positive for desmin, while S-100, CD-34, CD-117 reaction was negative.

During follow-up two months after surgery, US examination revealed two intrahepatic hypoechogenic foci of about 1 cm in diameter. Abdominal CT detected liver metastases. Liver biopsy confirmed metastatic leiomyosarcoma. The patient was referred to chemotherapy.

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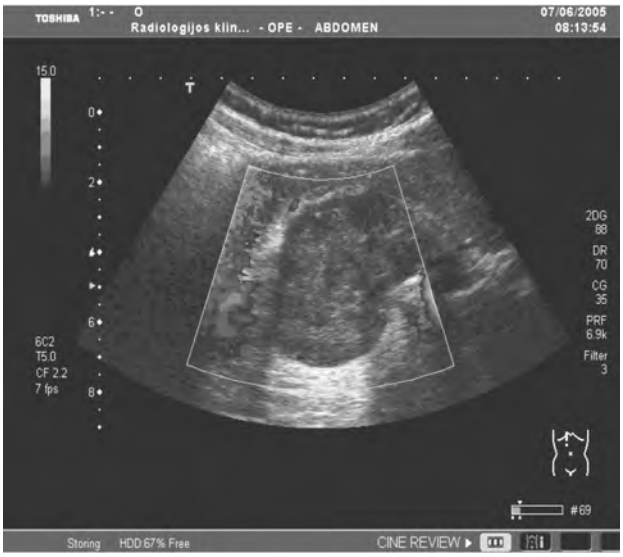

Fig. 1A. Abdominal ultrasound, including color Doppler technique
A heterogeneous vascular mass is evident.

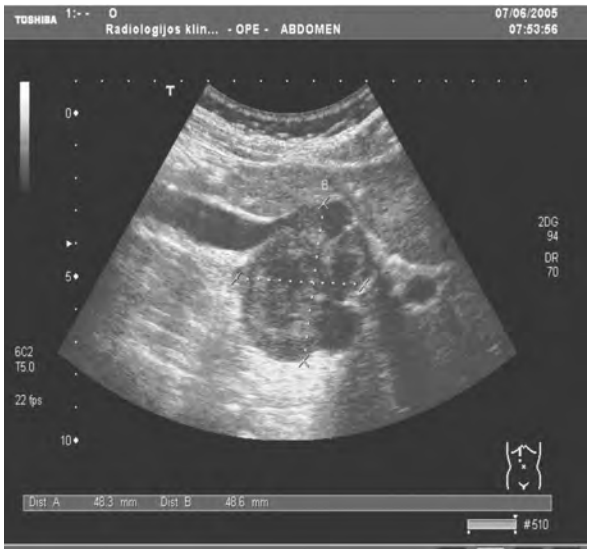

Fig. 1B. The tumor mass measuring 4.8×3.5 cm is evident
It is located along the posterior-lateral aspect of the inferior vena cava. The caval lumen is narrowed by the tumor.

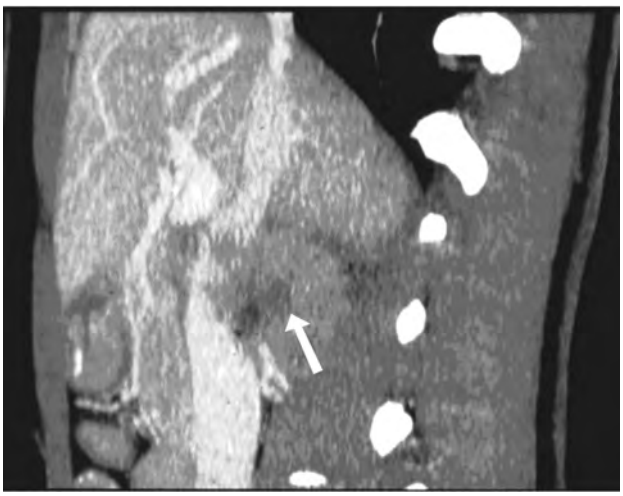

Fig. 2. Contrast-enhanced abdominal computed tomography, maximum intensity projection, MIP
Heterogeneous tumor is situated lateral-posterior to the inferior vena cava.

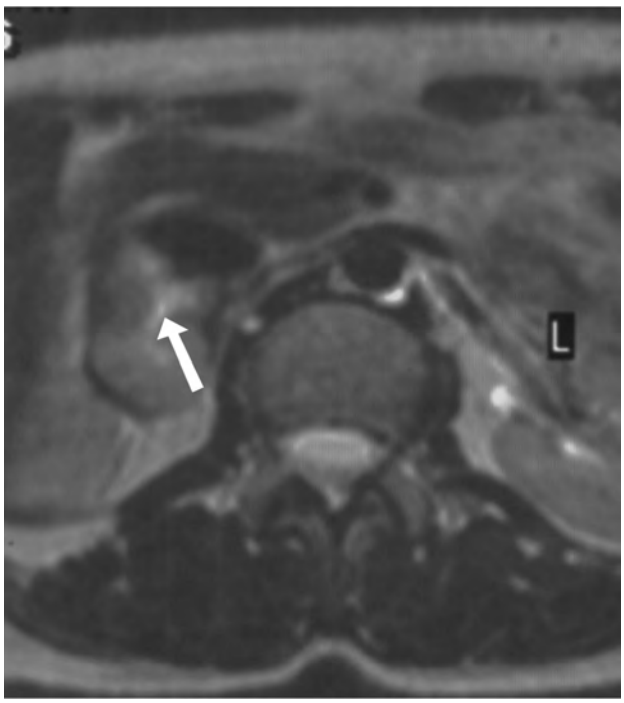

Fig. 3. Abdominal magnetic resonance imaging
Noncontrast T1W revealed a tumor with avascular regions (arrow) suggesting necrosis.

Discussion

Leiomyosarcomas occur mostly in adults aged 50 years and more. The mean age of patients with leiomyosarcomas is 55 years (range, 24–83 years) (2). Patients with certain inherited diseases, including neurofibromatosis (von Recklinghausen's disease) and Li-Fraumeni syndrome, are at increased risk of developing soft tissue sarcoma. Leiomyosarcoma is a tumor of mesenchymal origin usually arising in the retroperitoneum or uterus. It is the second most common primary retroperitoneal tumor after liposarcoma, accounting for 11% of all retroperitoneal malignant tumors (3).

Complaints are nonspecific (malaise, weight loss, abdominal or back pain, etc.) and related to tumor site and extent, as well as associated with vascular thrombosis. Symptoms in patients with involvement of the suprahepatic caval segment include nausea, weight loss, Budd-Chiari syndrome, and lower extremity edema (4). Cardiac abnormalities, including arrhythmias, are encountered in patients with tumors extending to the right atrium. Patients with tumors involving the middle segment present with epigastric and right upper quadrant pain. Some of

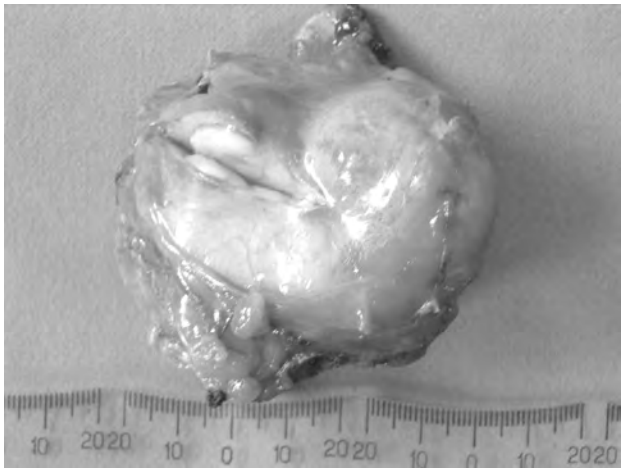

Fig. 4A. Findings at surgery:
a posterior view of the tumor with the removed inferior vena cava segment

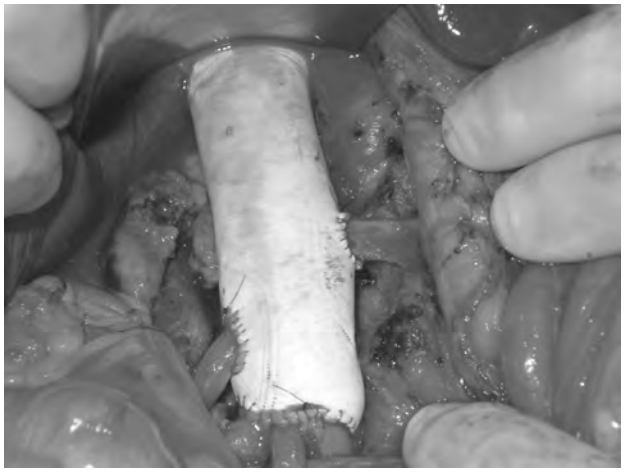

Fig. 4B. Findings at surgery: prosthesis was used for reconstruction of the removed inferior vena cava segment
Left and right renal accessory veins are seen as connected to the prosthesis.

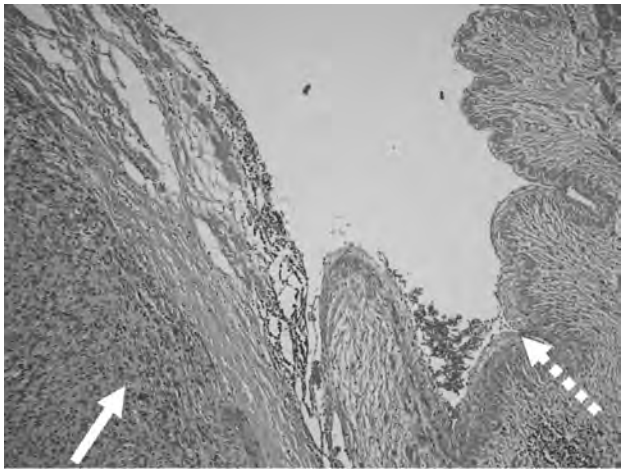

Fig. 5A. Histopathological examination
Wall of the inferior vena cava (dashed arrow) and the tumor (solid arrow) (hematoxylin-eosin, ×100).

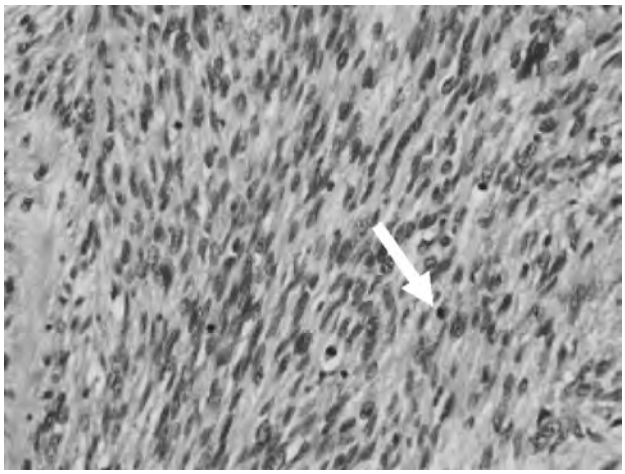

Fig. 5B. Histopathological examination of the tumor revealed numerous mitoses and cigar-shaped nuclei (arrow) (hematoxylin-eosin, ×400)

the tumors involve kidney, resulting in renal vein thrombosis and nephrotic syndrome. Though uncommon, arterial hypertension is a complication. Patients with involvement of the infrarenal caval segment tend to develop right lower quadrant pain, back/flank pain, and leg edema (5). Infrarenal leiomyosarcomas are often dormant for protracted periods thus causing venous obstruction at later stages.

US reveals these tumors to be predominantly mixed in echogenicity, with necrotic echo-poor areas. Hemodynamics of caval obstruction and renal vein thrombosis can be accurately depicted in Doppler studies. Bland thrombus may be differentiated from tumor by the presence of vascularity in the latter (5).

CT shows a lobulated well-defined heterogeneous tumor with signs of hemorrhage and necrosis (6).

MRI reveals a tumor of isointense signal on T1-weighted and iso- to hyperintense signal on T2-weighted images. Regions of hemorrhage result in a hyperintense T1-weighted signal. These tumors usually enhance markedly. With a complex appearance, both CT and MRI are useful in differentiating between neoplasm and a simple thrombus. Leiomyosarcomas with an extensive extravascular component are much more difficult to differentiate from retroperitoneal tumors compressing or invading IVC (6). At times, a tumor is so extensive that identification of the site of origin is not possible radiologically, surgically, or even pathologically.

Differential diagnosis of an intraluminal caval tumor includes leiomyosarcoma, angiosarcoma, tumor thrombus, and bland thrombus (4). Diagnosis can be suggested by imaging-guided biopsy, although

the specific type of sarcoma may be difficult to establish.

The prognosis is poor, with a mean survival time of one month in inoperable patients. The mean 5-year postresection survival rate is 28%. The incidence of metastases and local recurrence rate after surgical resection are reported to be approximately 36%.

Surgical resectability is highly dependent on tumor extent and location. Complete resection is often possible in the infrarenal segment. In the middle segment, a more complicated en bloc resection, at times including right nephrectomy, is necessary if the renal vein is involved. If a tumor involves the suprahepatic caval segment, complete resection is usually not possible due to frequent extension into hepatic veins and the right atrium (7).

Leiomyosarcomas of the IVC respond only minimally to chemotherapy or radiotherapy, thus surgical resection appears to be the only potentially curative treatment. Aggressive surgical management combined with adjuvant therapy offers the best treatment for these patients; however, the International Registry of Inferior Vena Cava Leiomyosarcomas reported that more than half of the patients developed tumor recurrence after radical resection, with recurrence consisting of local spread, distant metastases, or both (8).

In conclusion, leiomyosarcoma of the IVC is a rare tumor, with no specific clinical symptoms. This rare entity must be considered in differential diagnostics of mass lesions adjacent to the middle segment of the IVC. Renal cell carcinoma or adrenal carcinoma with extension into the IVC, angiosarcoma of the IVC, thrombus, and retroperitoneal sarcoma invading the IVC are other lesions occurring at this site. Accurate interpretation of clinical, imaging findings and histological examination usually allows accurate diagnosis.

Apatinės tuščiosios venos lejomiosarkoma

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Raktažodžiai: lejomiosarkoma, apatinė tuščioji vena, radiologinis ir patologinis tyrimas, chirurginis gydymas.

Santrauka. Lejomiosarkoma yra retas mezenchiminio audinio navikas, dažnai pažeidžiantis apatinę tuščiąją veną. Ankstyva šių navikų diagnostika prieš planuojamą chirurginį gydymą yra labai svarbi. Pilvo organų echoskopija, kompiuterinė tomografija bei magnetinio rezonanso tomografija – tai pagrindiniai tyrimo metodai šiuo atveju. Šių navikų diagnostikai svarbi kartu atliekama pritaikomoji biopsija. Mes pateikiame retroperitoninio tarpo naviko, išaugusio iš vidurinio apatinės tuščiosios venos segmento, atvejo aprašymą. Radiologiniais tyrimais nustatytas retroperitoninio tarpo navikas, skirtas operacinis gydymas. Histopatologinio tyrimo rodmenys patvirtino apatinės tuščiosios venos lejomiosarkomos diagnozę.

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