Rare case of a giant well-differentiated retroperitoneal liposarcoma

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KEY WORDS

retroperitoneum ◗ liposarcoma

ABSTRACT

Retroperitoneal liposarcoma is among the most common primary retroperitoneal tumors, along with malignant fibrous histiocytoma and leiomyosarcoma. It is slow-growing and has the propensity to displace rather than invade adjacent structures. Histologic subtypes include well-differentiated, myxoid (the most common form), and pleomorphic. Liposarcoma is most often seen in the fifth to seventh decades and is more common in men. We report the case of a patient who presented with a well-differentiated giant retroperitoneal liposarcoma.

INTRODUCTION

Sarcomas are rare tumors that annually represent less than 1% of all newly diagnosed malignancies in the United States [1]. These tumors may arise from mesenchymal tissue at any site in the body, including skeletal and extraskeletal connective tissues and the peripheral nervous system. Most of these tumors (approximately 75%) arise in soft tissue and the remainder in the bones. The majority of soft tissue sarcomas are found in the extremities, with only 13% occurring in the retroperitoneum [2]. The average annual incidence of retroperitoneal sarcomas was 2.7 cases per million population [3]. The majority of retroperitoneal tumors are liposarcomas and leiomyosarcomas. Most patients present with an asymptomatic abdominal mass, but symptoms may include early satiety, obstruction, and bleeding from pressure effects of the mass on neurovascular components.

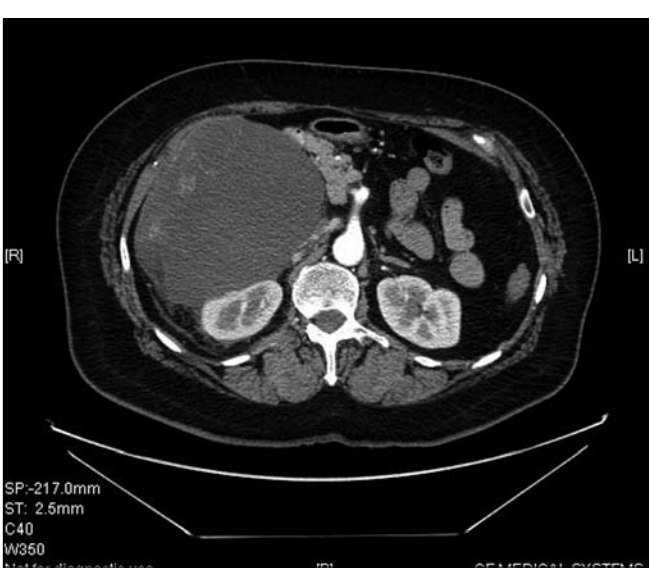

Fig. 1. Computed tomography of the abdomen.

We report the case of a patient who presented with diffuse abdominal pain and right-sided distention that had been increasing for approximately 10 weeks. Radiographic studies showed a large, heterogeneous, right-sided abdominal mass that extended across the midline and displaced various organs. Surgical excision yielded a well-differentiated retroperitoneal liposarcoma.

CASE REPORT

E.N., a 50 year old female patient, (Case History No. 865/2009) reported to a primary care physician (PCP) due to periodical pain in the right costovertebral angle region, which had been recurring for a few months. Laboratory tests revealed only an increase in the level of ESR to 80 mm/h. The PCP referred the patient for ultrasonography of the abdomen. The results revealed an encapsulated normoechogenic tumor below the right lobe of the liver 16 x 9.8 x 14.5 cm in size, which probably had come out from the dorsal surface of the right kidney and concurrently pressed on the kidney and translocated it. The patient was referred to the Outpatient Clinic of the Urology and Urological Oncology Clinic in the Medical University in Lublin.

Physical examination revealed the presence of pathologic resistance in the right costovertebral angle region, which reached the liver, laterally to the mid axillary line coming down to the right lower quadrant of the abdomen. On the 22nd of May, 2009, a computed tomography of the abdomen was conducted, which revealed a pathologic tissue structure 15.2 x 14.6 cm in size, extending from the hilus of the liver to the level of the ilium. The tumor pressed on the liver and gallbladder, distorted and flattened the right kidney from the back, and moved it inferiorly while embossing the abdominal integuments at the front. The tumor pressed on the inferior vena cava and widened the lower segment of the inferior vena cava and efferent vessels. There were hypodense areas in the area of the tumor with some necrotic traits. The described lesion did not seem to infiltrate the adjacent structures (Fig. 1).

On the 2nd of June, 2009, the patient was examined at the Urology and Urological Oncology Clinic of the Medical University in Lublin. Complete blood count revealed a decrease in the level of hemoglobin to 10.4 g/dl and hematocrit to 32.1% as well as an increase in the amount of blood platelets to 662 x10^3/ul. The remaining results were normal. The patient was prepared for surgical treatment.

On the 8th of June, 2009, the patient was operated on; the tumor located on the right side of the retroperitoneal space was removed transperitoneally (Fig. 2). Due to infiltration of the renal capsule and difficulties in removing the tumor from the right kidney, nephrectomy was carried out. The size of the removed tumor was 18 x 17 x 13 cm (Fig. 3). Within the first 24 hours the surgical treatment, due to a decrease in the level of hemoglobin and hematocrit to 8.66 g/dl and 22.9% respectively, 2 units of erythrocyte mass that was compatible with her blood type was transfused to

the patient normalizing the blood-count parameters. Further postoperative treatment was uncomplicated and 8 days after the operation the patient was discharged from the hospital. The sutures from the surgical wound were removed on the 10th day after the operation at the PCP's outpatient clinic.

The results of histopathological examination of the tumor segments (No. 24122/09) revealed areas with adipose tissue structure as well as areas of fibrous structure which contained collagen fibers with dispersed fusiform cells, as well as multilateral cells with hyperchromatic nuclei. Moreover, there were also numerous lipoblasts with irregular nuclei and a bright cytoplasm. Additionally, there were dispersed division figures (M1B1 – positive in about 15% of the tumor's cells). The structure of the tumor contained necrosis (about 10% of the tumor's mass). The lesion was encapsulated and there was an infiltration of the capsule without breaking its continuity. The immunohistochemical reactions in the tumor's cells: Vim (+), CK (-), Des (-), CD31 (-), CD 34 (focally+), S-100 (+), and SMA (-). The microscopic picture, including the immunoprofile, matched that of a well-differentiated sclerosing liposarcoma.

The patient was referred for further treatment at the Oncology Clinic. To this day, she has not reported for a check-up at the Outpatient Clinic of the Urology and Urological Oncology Clinic of the Medical University in Lublin.

DISCUSSION

Retroperitoneal liposarcomas are unusual tumors accounting for 0.15% of all malignancies and the majority of retroperitoneal sarcomas [4]. Four subtypes of liposarcoma are described: well differentiated, dedifferentiated, myxoid/round cell, and pleomorphic [6].

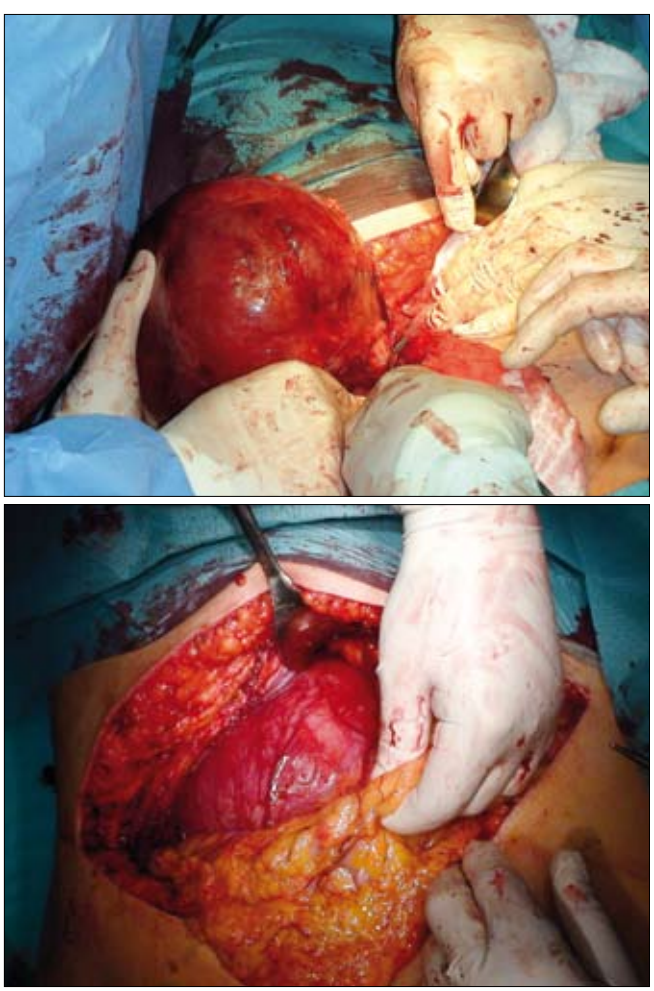

Fig. 2. Intraoperative transperitoneal view of the tumor.

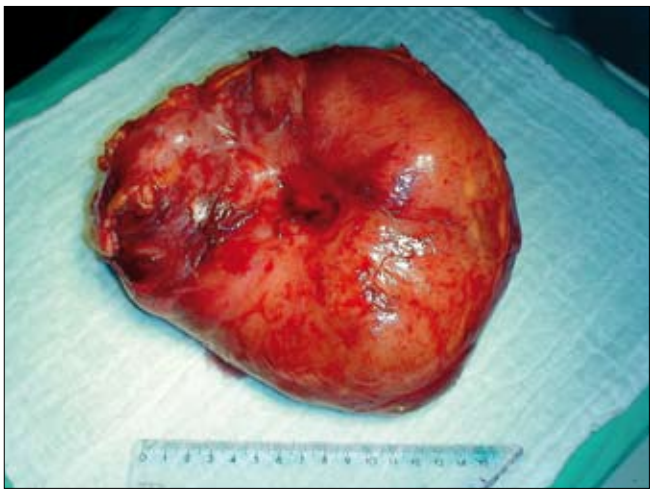

Fig. 3. Removed tumor – a well-differentiated liposarcoma.

The overall prognosis is most dependent on the completeness of resection (margin status) and the histologic grade of the malignancy (high grade versus low grade), with the most favorable prognosis found in complete resection of a low-grade malignancy [5, 7, 9]. Dedifferentiated liposarcoma is associated with a fourfold increase in risk of local recurrence compared with well-differentiated histology and will recur locally in nearly 80% of cases despite aggressive surgical therapy. They metastasize in approximately 18% of cases with the liver and lung being the most common sites.

Treatment of retroperitoneal liposarcoma is primarily surgical. The ability to completely resect the tumor is the most important predictor of local recurrence and overall survival [8]. Unfortunately, retroperitoneal liposarcoma is almost always large at the time of diagnosis owing to its slow growth and vague symptoms [15]. Preoperative evaluation is best accomplished with abdominal CT scanning; however, MRI may be useful in select circumstances where involvement of unresectable structures is suggested by the CT. In select cases, angiography may be helpful. Metastatic evaluation with CT of the chest is also advised when approaching these patients with curative intent.

Vague abdominal discomfort and abdominal mass are the presenting complaint in 60-80% of cases with only about 6% of patients presenting without symptoms. Other less common presenting symptoms are weight loss, fever, anorexia, genitourinary complaints, and bowel obstruction [7].

The paucity of symptoms allows these malignancies to attain considerable size before presentation, making adjuvant treatment with tumoricidal doses of radiation hazardous and likely to cause significant morbidity to adjacent vital structures [10, 11]. In one large series, the presenting size was over 10 cm in 71% of the cases. Attempts at intraoperative radiotherapy combined with external beam radiotherapy have demonstrated no survival benefit when compared to standard external radiotherapy alone. Chemotherapy in tumors with a significant mitotic rate also fails to show any improvement in overall survival [12, 13]. Overall survival rates of 43-55% at 5 years are typically reported [14].

Complete resection is possible in 60% of cases and often involves removal of adjacent organs such as kidney, ureter, and large bowel. The most frequent organ resected is the kidney. More complex cases may involve en-block resection of gallbladder, psoas muscle, small bowel, spleen, pancreas, and major vascular structures. In selected cases of unresectable tumors, incomplete resection can increase survival and provide palliation of symptoms compared to biopsy alone.

CONCLUSION

Retroperitoneal liposarcoma is among the most common primary retroperitoneal tumors. Its appearance shall always be taken into consideration especially when a tumor is found in this area.

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