Myxoid Retroperitoneal Liposarcoma in a Patient with Personal and Family History of Different Cancers

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Abstract

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Introduction

Sarcomas of retroperitoneum contain heterogeneous groups of tumors with varying histology, probability of complete resection, and propensity to recur and difficulty in development of effective treatment and challenging. In this article we present a case of recurrent myxoid retroperitoneal liposarcoma in a patient with personal and family history of different cancers.

Case presentation

A- 66- year old man with history of low grade non- invasive bladder cancer (PTaG1), retroperitoneal sarcoma and family history of colon adenocarcinoma and malignant asterocytoma of brain in his daughter and nephew respectively was referred to our clinic due to recurrent retroperitoneal mass. Bladder tumor was managed with TURBT and single dose of intravesical mitomycin-c. Bladder tumor did not recur during 5 years of follow up. His first retroperitoneal surgery was performed 1.5 year before. The limited numbers of frozen sections from tumor bed were negative after tumor resection. Adjuvant radiotherapy was not included in patient's management. The patient was referred to our centre due to a recurrent retroperitoneal mass (Fig 1&2). Metastasis work- up was negative for metastatic disease. En-block resection of tumor which had grossly invaded psoas muscle was performed. The part of left psoas muscle lateral to vertebral body was removed consequently. Frozen sections of tumor bed were negative at the end of tumor resection. Adjuvant radiotherapy regimen was included in patient's management. The pathology report of specimen was myxoid liposarcoma. The patient was

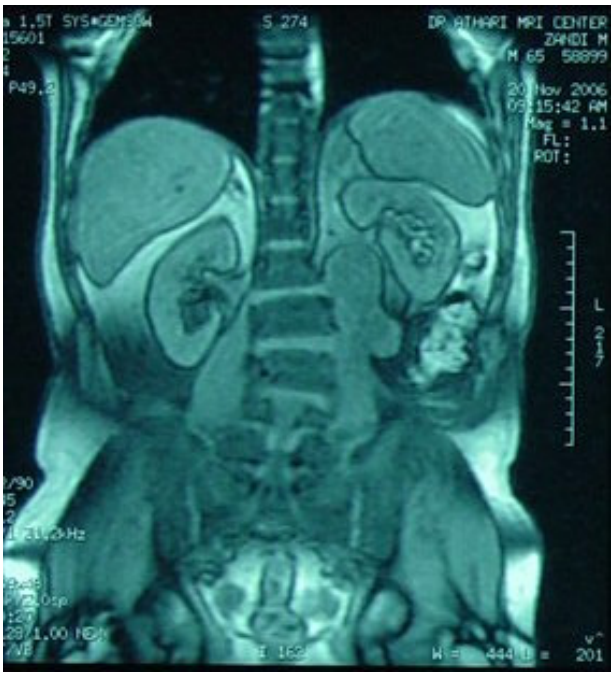

Figure 1. Retroperitoneal liposarcoma situated in lower part of left kidney.

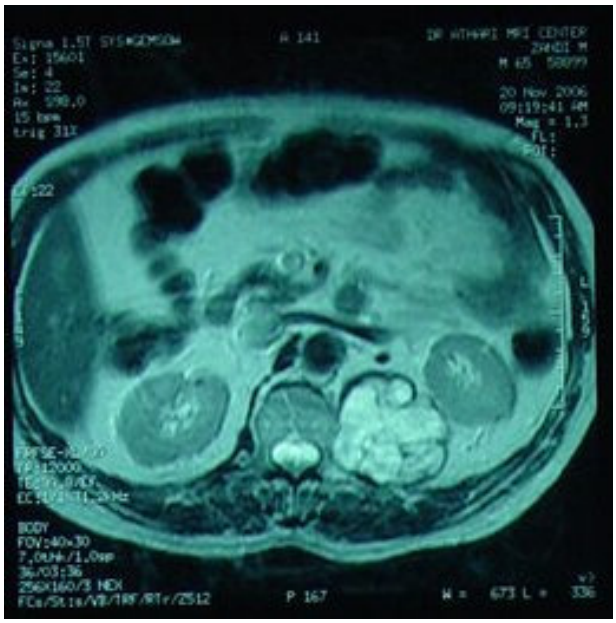

Figure 2. Retroperitoneal liposarcoma which has invaded left psoas muscle.

followed by medical history, physical examinations and blood chemistry every 3 months and also chest and abdominal CT scan every 6 months. There was not any local recurrence during 1 year of follow up. Unfortunately, the patient developed lung metastasis 1 year after second surgery (fig3).

Discussion

Sarcomas represent 0.8% of new diagnosed malignancies yearly with liposarcoma accounting for 20% of all sarcomas. About 13% of sarcomas occur in the retroperitoneum [2,4]. The various types of retroperitioneal sarcomas originate from mesenchymal cells that present as different histology types depending on their cell of origin. These include liposarcoma, leiomyosarcoma, malignant fibrous histiocytomas, and fibrosarcomas[1,2,4]. Prognosis of liposarcoma depends on degree of differentiation, size, histological type and tumor staging. [5,6]. The liposarcoma is classified according to the histologial type, as well differentiated type , mixoid types and pleomorphic type. [2,4,7]. The mixoid types occur in 60% of situations , well differentiated in 25% and pleomorphic in 10% of cases[4]. The poleomorphic type is highly aggressive with high rate of metastasis. The retroperitoneal liposarcoma tends to be low or intermediate grade, while other sarcomas in this location tend to be high grade [5,6]. Most patients with retroperitoneal tumors have vague symptoms at the time of presentation. The symptoms such as constipation, urological problems, or flank pain may prompt the patient to seek medical care. The constitutional symptoms, such as fever, weight loss and night sweats can be indicative of lymphoma and therefore important to examine all nodal tissues in these patients [2, 4, 7]. If the tumor is retroperitoneal, the liver may be the first site of metastasis [5, 6]. After history and physical examination, imaging is warranted. A good quality CT scan of the abdomen and pelvis generally provides adequate anatomic details [1]. CT allows the surgeon to refine the preoperative differential diagnosis as well as to determine resectibility.[1] MRI may also be used, but probably does not provide additional information over CT for tumors in the abdomen and retroperitoneum [1,2]. Specific Tumor organ relationships must be determined preoperatively, as potentially curative surgery frequently mandates resection of involved organs, such as the kidney. As a rule, most tumors in the retroperitoneum are malignant, and will require removal. Retroperitoneal sarcoma (RPS) indicates of aggressive surgical management [2,3,4].There are several instances where biopsy of a suspected sarcoma may be recommended. If the diagnosis is under question, a neoadjuvant therapy protocol is designed, or for tissue diagnosis in the face of unrespectable metastatic disease biopsy will be needed. A percutaneous core needle biopsy is usually recommended in this situation [2,3,4]. Due to the difficulty in surgery and the imprecise determination of negative microscopic margins, there are chances that life can be saved at the cost of adjacent organs. Extensive resection may decrease the local recurrence which occurs in 39%-63% of cases undergoing complete resection. So, regular surveillance with abdomen pelvic CT and MRI is necessary [5,6]. Liposarcoma anatomical features and similarity between a well differentiated fatty tumor and normal fatty tissues make the complete removal of a whole liposarcoma to be difficult [5,6]. Complete excision has been achieved in less that 70% of cases and 5-year local recurrence rates ranged from 37 to 75%. In addition to technical difficulty of achieving negative surgical margins, the local recurrence rates can be affected by variations such as tumor grade and sarcoma histological sub type [5,6].

Because of a high rate of local recurrence there has been increased interest in identifying multimodality treatment regimens that may improve local relapse-free survival following resection of RPS. There is a good evidence that radiation therapy improves local control rates. Adjuvant radiation therapy has been found effective in improvement of post operative local control rate [5,6,8]. The role of

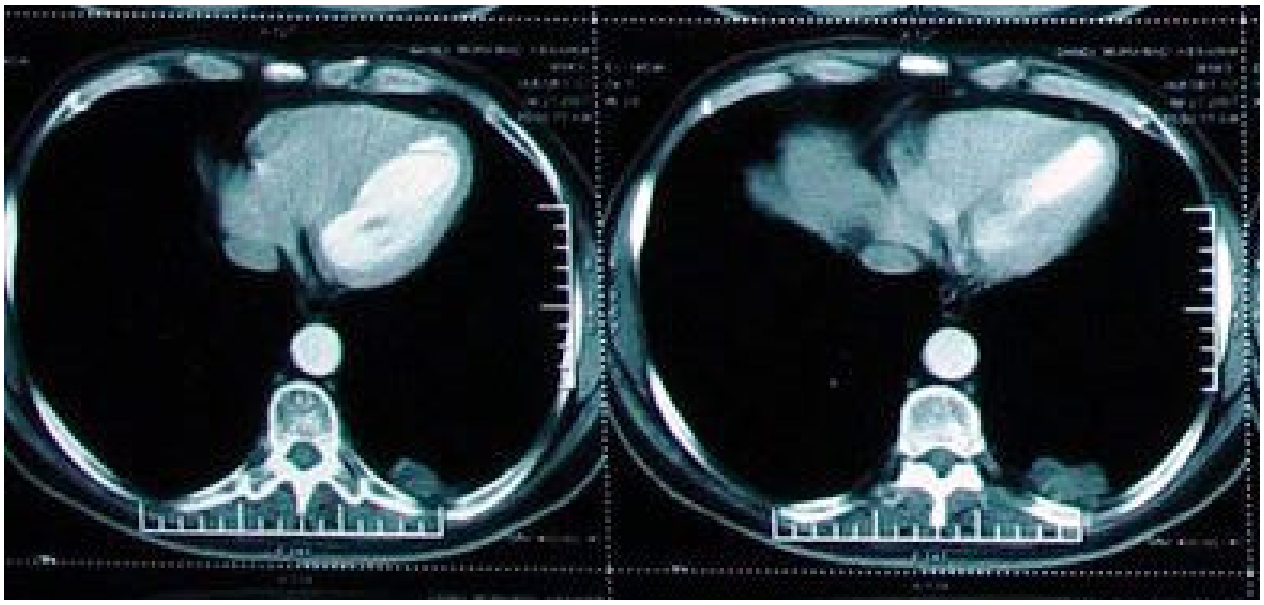

Figure 3. CT scan showing pulmonary metastasis

chemotherapy in handling RPS is under investigation [1,8]

Prompt reoperation for recurrent disease improves local control but long term improvement in survival has not been conclusively demonstrated

In our patient, it seems if adjuvant radiotherapy was added after primary resection, we would have had lower probability of retroperitoneal tumor recurrence. Meanwhile, It seems that micro metastasis was present at the time of second surgery ,so even with local eradication of tumor, the patient developed metastatic disease. The simultaneous bladder tumor and retroperitoneal sarcoma in him and death of 2 members of his family confirms that a full genetic work up including tumor suppressor gene assessment could be performed in his family.

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