A huge retroperitoneal liposarcoma: case report

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Summary

Background: Soft tissue sarcomas are rare and account for less than 1% of all newly diagnosed malignancies. One-third of malignant tumors arising in the retroperitoneum are sarcomas. Liposarcoma is the most common soft tissue sarcoma and retroperitoneal sarcoma. Liposarcoma accounts for at least 20% of all sarcomas in adults and up to 41% of all retroperitoneal sarcomas. Here we present the case of a huge retroperitoneal liposarcoma and a brief literature review. *Case report:* A 34-year-old woman was referred to our hospital from a local clinic, because of abdominal distention, pain, and palpable mass. On admission we found that her abdomen was markedly distended. Computed tomography showed a the huge left ovarian mass that occupied almost the entire abdominal cavity. The mass consisted mainly of fat, and calcified material. She was operated under the diagnosis of a huge teratoma. The tumor was located in the retroperitoneal cavity and it abutted the left adnexa. The retroperitoneal tumor, including the left adnexa was removed. The tumor measured 22 x 15 x 11 cm, and showed many histological and pathological findings. On the basis of the histopathological finding, the tumor was diagnosed as a dedifferentiated liposarcoma of the retroperitoneum. The patient is presently undergoing radiation therapy. *Conclusion:* In retroperitoneal liposarcoma, histological subtype, incomplete resection, contiguous organ resection, and older age are strongly associated with tumor-related mortality. For liposarcoma, it is necessary to customize the treatment strategy on a case-by-case basis.

Key words: Retroperitoneal liposarcoma; Histological subtype; Customized treatment.

Introduction

Soft tissue sarcomas are rare, accounting for less than 1% of all malignancies, and one-third of all malignant tumors arising in the retroperitoneum are sarcomas. Liposarcoma is the most common soft tissue sarcoma in adults, and is often located in the retroperitoneum. Liposarcomas originate from the mesoderm, and are derived from adipose tissue. The peak age of incidence ranges from 40 to 60 years. Retroperitoneal tumors grow slowly, but they may grow to a very large size [1, 2]. Here we report the case of a huge retroperitoneal liposarcoma, resembling a huge teratoma.

Case Report

A 34-year-old woman was referred to our hospital from a local clinic for a 2-week history of abdominal distention, pain and palpable mass. She denied experiencing any vesical irritation. She had no history of surgery or medication and her family history was unremarkable. Her abdomen was markedly swollen and firm, but tender. Laboratory findings including tumor marker results within normal limits. Computed tomography (CT) showed a huge, inhomogeneous mass arising from the left ovary, consisting mainly of fat and calcified material. It was considered to be a mature teratoma (Figure 1). During laparotomy, the mass was seen as a large lipomatous tumor occupying almost the entire abdominal cavity. It was located in the retroperitoneal space, and it abutted on the left ovary. Both the ovaries, however, were grossly free. The mass was completely excised, but it was difficult to obtain clear margins sparing the major vessels and adjacent organs. The retroperitoneal tumor, including the left adnexa was removed, and microscopic analysis of frozen tumor sections showed liposarcoma. We decided to remove the uterus. The mass measured 22 × 15 × 11 cm, grossly appeared multinodular, and contained many yellowish fatty areas (Figures 2 A, B, C). The histopathological diagnosis was of dedifferentiated liposarcoma (Figures 3). Positron emission tomography (PET) CT performed after surgery did not show metastasis or remnant tumor (Figure 4). The postoperative course was uneventful, and the patient was discharged nine days after the surgery. Presently she is undergoing radiation therapy.

Discussion

Liposarcomas originate from the mesoderm and are derived from adipose tissue. They account for 10~14% of all soft tissue sarcomas, and less than 1% of all malignancies. There are some types of sarcoma, liposarcoma (41%), leiomyosarcoma (28%), malignant fibrous histiocytoma (7%), fibrosarcoma (6%), etc. Liposarcoma is the most common tumor of the retroperitoneum accounting for 0.07-0.2% of all neoplasms [2]. The symptoms arise because of the compression of the organs, similar to that reported in our case. Patients mostly present with a mass only a few weeks or few months old, while metastasis at the first occurrence is not common. The histological type is the most important factor affecting survival rates for patients with liposarcoma [1].

Histologically the subtypes of liposarcomas are distinguished by the composition of lipoblasts. They are divided into four subtypes: well differentiated, dedifferentiated, myxoid/round cell, and pleomorphic. Dedifferentiated liposarcomas are characterized by the coexistence of well differentiated and poorly differentiated, non-lipogenic areas that are present either in a portion of the same tumor or in the primary and recurrent tumors. The dedifferentiated type is thought to be the transformation of well differentiated liposarcoma [3]. Conyers *et al*., reported that dedifferentiated liposarcoma is more aggres-

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Fig. 1

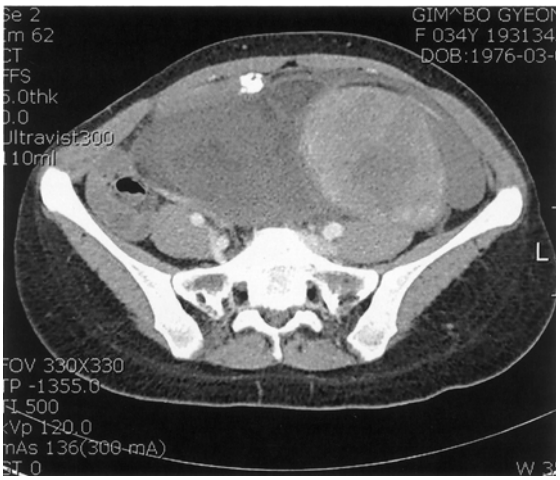

Fig. 2A

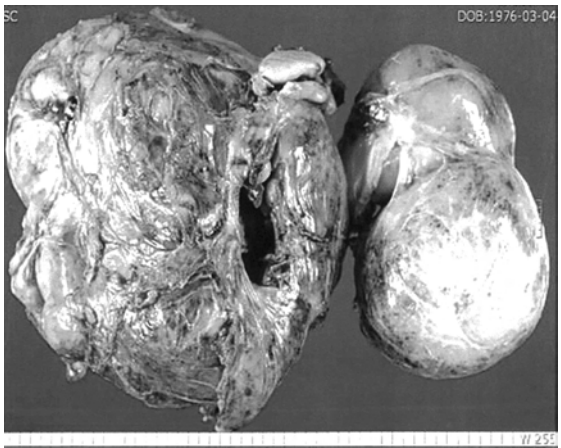

Fig. 2B

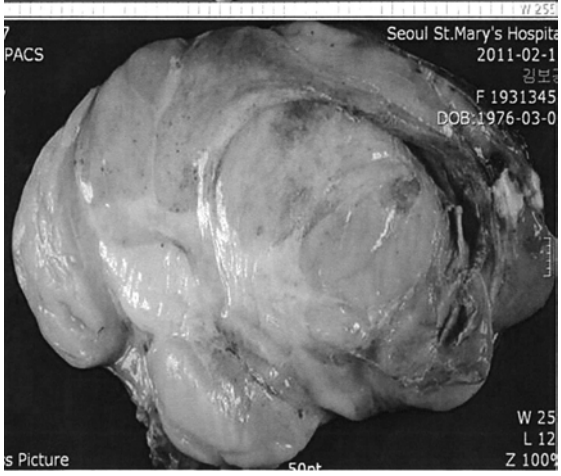

Fig. 3

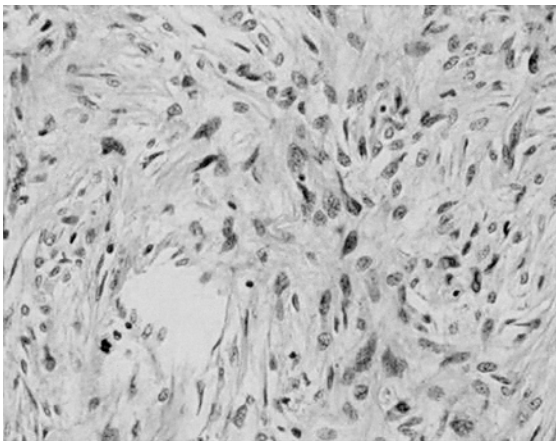

Fig. 2C

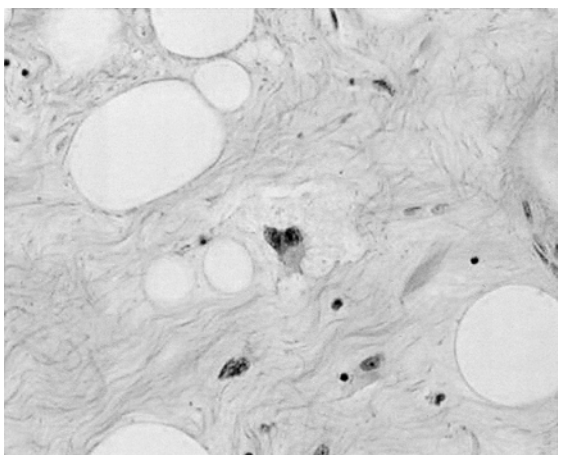

Fig. 4

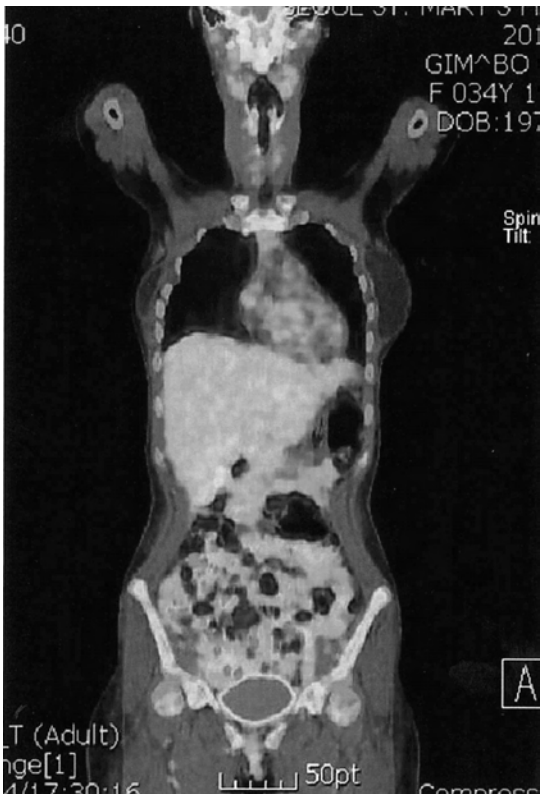

Figure 1. — Computed tomography (CT) scan obtained before the surgery shows a mass containing a large amount of fat and calcified material. It appears as a mature teratoma.

Figures 2) A, B — Photographs of the mass obtained after surgery. C) In cross section, large masses of fat can be seen. Shows a section of a well differentiated area of the dedifferentiated liposarcoma in which lipoblasts are seen in fibrotic background (× 200).

Figure 3. — The pathological findings of the tumor. Shows a section of dedifferentiated liposarcoma composed of spindle shaped cells (× 200).

Figure 4. — Positron emission tomography (PET) CT scan obtained after surgery did not show distant metastasis or remnant tumor.

sive and metastatic than its well differentiated counterpart [4]. McCallum *et al*. reported that for patients with liposarcoma, the 5-year survival rate of the patients with the well differentiated type is 90%, dedifferentiated type is 75%, myxoid/round cell type is 60~90%, and pleomorphic type is 30~50% [15]. With regard to tumor metastasis, the retroperitoneum is the location with the worse prognosis. Distant metastasis is more common in the dedifferentiated type than in the well differentiated type. In large tumors (> 20 cm), distant metastasis is more common [2].

With regard to treatment modalities for liposarcoma, there are several opinions. Lahat *et al*. reported that clinicians generally treat dedifferentiated liposarcoma patients with systemic chemotherapy followed by an aggressive surgical approach, whereas those with well differentiated type are treated only with less aggressive surgery. However less aggressive treatment does not improve the long-term outcome in patients with well differentiated liposarcoma [6]. Herrera-Gomez *et al*., stated that for patients with well differentiated liposarcoma, adjuvant radiation therapy was not considered, because of its side-effects such as GI problems which override its therapeutic effect. However they recommend that clinicians should consider radiation therapy in incomplete resection or inoperable cases. Further they also mentioned that radiation therapy may increase overall and disease-free survival [2]. However regarding liposarcoma treatment, there are also contradictory reports. Singer *et al*. stated that the histological subtype of retroperitoneal liposarcoma, incomplete resection, contiguous organ resection, and older age are strongly associated with tumor-associated mortality. An aggressive surgical approach should be adopted to achieve complete resection. If necessary en bloc resection of adjacent organs should be performed to achieve complete resection. Despite aggressive treatment, 80% of dedifferentiated liposarcomas will recur locally and 30% will metastasize to distant sites within three years of diagnosis. A combination of surgical therapy, and systemic and locoregional therapies is needed to improve outcomes in patients with retroperitoneal liposarcoma [7].

The present case was the dedifferentiated subtype of liposarcoma in a relatively young patient. To lower the possibilities of local recurrence and distant metastasis, we decided to give the patient radiation therapy, and she has been tolerant to the therapy.

Conclusion

Retroperitoneal liposarcomas are rare; however, they require an aggressive surgical approach, including multi-organ resection, if necessary, or multiple resections in the case of recurrence. In retroperitoneal liposarcoma, histological subtype, incomplete resection, contiguous organ resection, and older age are strongly associated with tumor-related mortality [5]. Especially in the dedifferentiated type local recurrence and distant metastasis rates are high. For liposarcoma, it is necessary to customize the treatment strategy on a case-by-case basis.

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