



Giant retroperitoneal dedifferentiated liposarcoma mimicking ovarian cancer: A case report

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ARTICLE INFO

Keywords

Retroperitoneal tumor
Dedifferentiated liposarcoma
Huge tumor

ABSTRACT

Retroperitoneal liposarcoma is a rare tumor, and its dedifferentiated subtype and a larger diameter are associated with a poor prognosis. However, there are few reports of retroperitoneal liposarcomas, both with a dedifferentiated subtype and a diameter of >30 cm. We report a case of a giant retroperitoneal liposarcoma with a dedifferentiated subtype. A 78-year-old woman presented to our hospital with abdominal distension and loss of appetite. Computed tomography and magnetic resonance imaging findings revealed a 35-cm-diameter solid tumor in the peritoneal cavity. CA125 (64.8 U/mL) and HE4 (229.0 pmol/L) were elevated preoperatively raising suspicion for ovarian malignancy. However, intraoperative findings revealed that the tumor originated in the retroperitoneal cavity. Reductive surgery for the tumor and partial resection of the sigmoid colon and left ureter were performed, and pathological examination confirmed a retroperitoneal dedifferentiated liposarcoma. Although her symptoms improved postoperatively, she died 11 months after surgery due to disease progression.

1. Introduction

Retroperitoneal sarcoma originates in the retroperitoneum and accounts for 0.15% of all malignancies (Zhuang et al., 2021). Liposarcoma (LPS) is the most common histological subtype of retroperitoneal sarcoma, accounting for 30% (Yokoyama et al., 2020). According to the World Health Organization classification, LPS is classified into five subtypes: 1) well-differentiated, 2) dedifferentiated, 3) myxoid, 4) pleomorphic, and 5) myxoid pleomorphic (Sbaraglia et al., 2021). The dedifferentiated and pleomorphic histological subtypes and a larger diameter are associated with a poor prognosis of retroperitoneal LPS (Xiao et al., 2021). Although complete resection is the mainstay of treatment, surgical resection for large retroperitoneal LPS entails a high risk due to the size and anatomical location of the tumor and difficulty in preoperative diagnosis (Schwarzbach et al., 2006; Kim et al., 2017). However, there are few reports of the dedifferentiated subtype of LPS that also has a diameter of >30 cm. We report a case of a giant retroperitoneal dedifferentiated LPS mimicking preoperative ovarian malignant tumor.

2. Case presentation

A 78-year-old woman presented to our hospital with abdominal distension and loss of appetite. She was G3P3 and had a history of total abdominal hysterectomy for uterine myoma at the age of 53, and dementia. Transabdominal ultrasonography and computed tomography (CT) revealed a solid tumor that occupied the pelvic cavity to the upper abdomen. The mass measured 35 × 20 × 13 cm and showed complex enhancement patterns and fatty components (Fig. 1A, B). In addition, the tumor suggested continuity with the arteriovenous vein of the left ovary. ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT showed abnormal accumulation of FDG in the tumor and no abnormal accumulation in other organs or lymph nodes (Fig. 1C). On magnetic resonance imaging (MRI), most of the tumor showed low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig. 1D). Blood analysis indicated renal dysfunction (Cre: 1.06 mg/dL; BUN: 12.6 mg/dL), and anemia (Hb: 8.7 g/dL). CA125 (64.8 U/mL) and HE4 (229.0 pmol/L) levels were elevated. CA19-9, CEA, SCC, and AFP levels were within normal ranges. Based on these

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<https://doi.org/10.1016/j.gore.2022.101088>

Received 31 August 2022; Received in revised form 11 October 2022; Accepted 17 October 2022

Available online 18 October 2022

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findings, we suspected a malignant tumor originating from the left ovary, such as malignant transformation of a mature teratoma. D-dimer level (11.6 $\mu\text{g}/\text{mL}$) was elevated, and ultrasonography revealed the peripheral deep venous thrombosis in the right lower limb. Although the general condition of the patient was unsuitable for surgical treatment with her performance status (PS) of 3, her family strongly desired surgery to alleviate her discomfort. Therefore, we performed transabdominal resection of the tumor with informed consent. Operative findings revealed that the tumor was located in the mesentery of the sigmoid and descending colon (Fig. 2A) and was continuous with the retroperitoneum. Bilateral ovaries were confirmed to be grossly unremarkable. Based on the operative findings, we considered that the tumor originated from the peritoneal cavity. The dorsal side of the tumor was widely adherent to the abdominal aorta. The left common iliac artery, inferior mesenteric artery, and left ureter were involved in the tumor. Reductive surgery for the tumor with partial resection of the sigmoid colon, ureterectomy, and anastomosis of the sigmoid colon and of left ureter were performed by gynecologic oncologists, a gastrointestinal surgeon, and a urologist. The resected tumor measured $32 \times 25 \times 20$ cm and weighed 7,800 g (Fig. 2B, C). The tumor was incompletely resected, and the residual part over a length of 6 cm remained on the surface of the left common iliac artery. The operative time and amount of blood loss were 427 min and 8,625 g, respectively. Grossly, the cut surface of the mass showed a multinodular appearance composed of yellow and glistening fat tissue-like areas and white-tan solid areas mixed with hemorrhage and necrosis (Fig. 3A). On histological examination, the fat tissue-like areas were composed of mature adipose tissue with occasional atypical cells with hyperchromatic and enlarged nuclei (Fig. 3B, right upper inset). The solid areas showed proliferation of short spindled and round cells with marked pleomorphism. Scattered lipoblasts were focally seen (Fig. 3B, left lower inset). These two areas were sharply demarcated with fibrous septa (Fig. 3B). Immunohistochemical stains showed the tumor cells in both two areas to be positive for CDK4 and MDM2. Based on the operative findings and the results of the histopathological examination, the tumor was diagnosed as a retroperitoneal

dedifferentiated LPS. After surgery, her general condition improved and her food intake gradually increased. Postoperative treatment, such as chemotherapy or radiotherapy, was not administered because of her age, PS, and lack of evidence of postoperative LPS treatment. Progression of the residual tumor was observed after 4 months, and the patient died 11 months after surgery due to disease progression.

3. Discussion

LPS is a mesenchymal cell-derived tumor that differentiates into lipocytes. The dedifferentiated subtype arises from well-differentiated LPS, and histopathologically dedifferentiated LPS shows highly atypical spindle-shaped cells with pleomorphic and multinucleated features, resembling undifferentiated pleomorphic sarcoma. Rarely, a lipogenic differentiation with accompanied lipoblasts can be seen in the dedifferentiated component as in the present case (Sbaraglia et al., 2021; Henricks et al., 1997). Although there have been several reports on retroperitoneal dedifferentiated LPS (Yokoyama et al., 2020; Xiao et al., 2021; Kim et al., 2017), only a few cases of retroperitoneal dedifferentiated LPS with a diameter of >30 cm have been reported. Therefore, few surgeons have sufficient experience with giant retroperitoneal dedifferentiated LPS, contributing to inaccurate diagnosis and incomplete surgery. The present case of giant retroperitoneal dedifferentiated LPS would be informative for management of the disease.

Preoperative diagnosis is important for optimal treatment of retroperitoneal LPS. CT is the most useful imaging method for evaluating tumor location, size, origin, and relationship to adjacent tissues and organs (Messiou et al., 2017). Displacement of the retroperitoneal organs indicates that the tumor is retroperitoneal in origin. The presence of abnormal fatty tissues is useful in the diagnosis of well-differentiated LPS. The well-differentiated subtype showed $>75\%$ adipose tissue and internal nodular areas on CT. In contrast, the dedifferentiated subtype typically shows non-lipomatous components and nodular areas with dense and heterogeneous signals on contrast-enhanced CT. In the case of dedifferentiated LPS with well-differentiated components, such as in this

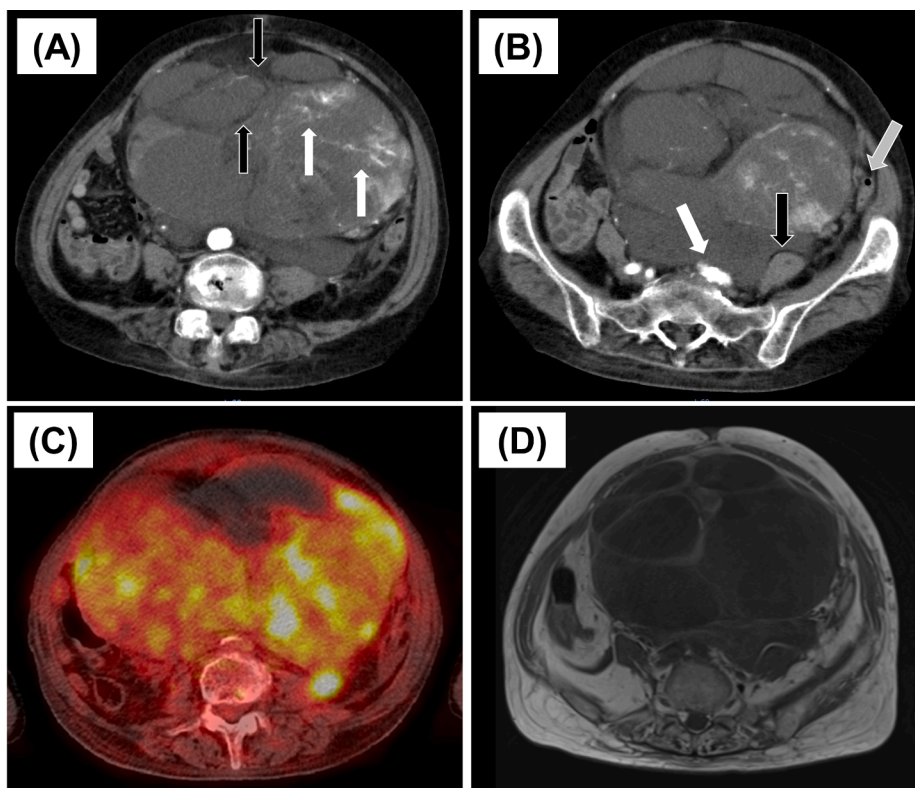


Fig. 1. Computed tomography (CT), ^{18}F -fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT, and magnetic resonance imaging, axial view. (A) CT shows a 35-cm-diameter solid tumor occupying the area from the pelvic cavity to the upper abdomen. White and black arrows show complex enhancement patterns and fatty components, respectively. (B) The mass is in contact with the left common iliac artery (white arrow), descending colon (gray arrow), and left iliopsoas muscle (black arrow). (C) PET-CT shows abnormal accumulation of FDG inside the tumor. (D) T2-weighted image shows low signal in a large part of the tumor.

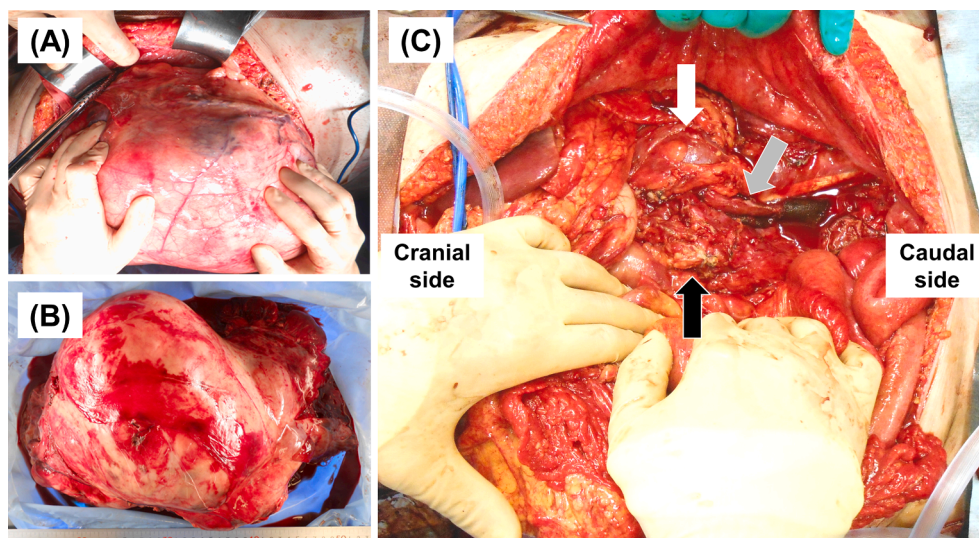


Fig. 2. Operative findings. (A) The tumor was located in the mesentery between the sigmoid colon and the descending colon. (B) Resected tumor measured 32 × 25 × 20 cm. (C) White, gray, and black arrows show the left kidney, repaired left ureter, and residual tumor adherent to the abdominal aorta, respectively.

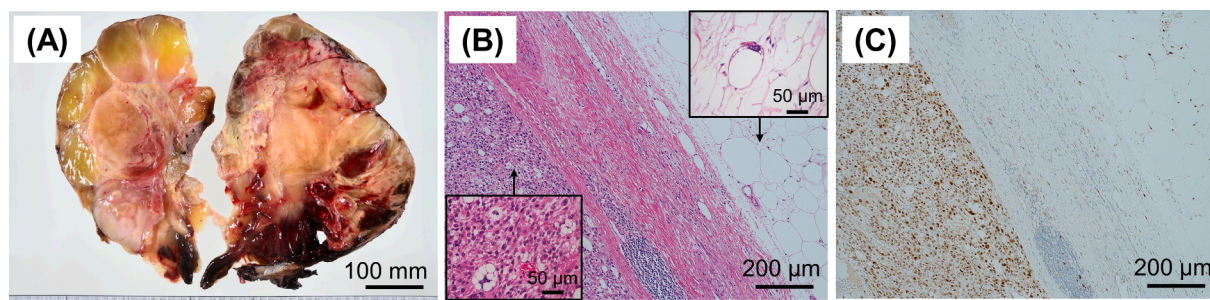


Fig. 3. Histopathological findings. (A) Cut surface of the resected mass. The heterogeneous appearance composed of fat tissue-like areas and solid areas mixed with necrosis and hemorrhage. (B) Well-differentiated and dedifferentiated components are present with abrupt transition. In the well-differentiated area, atypical cells are scattered (right upper inset). In the dedifferentiated area, short spindle-shaped cells with atypical nuclei are present at high cell density accompanied by lipoblasts (left lower inset). (C) Both well-differentiated and dedifferentiated components showed nuclear positivity for MDM2 on immunohistochemical staining.

patient, features of both well- and dedifferentiated LPS were found. Furthermore, a recent study revealed that the maximum standardized uptake value measured using PET-CT was associated with the prognosis and pathological grade of retroperitoneal LPS (Subramaniam et al., 2021). MRI is a useful option when precise anatomical information is required (Messiou et al., 2017). In addition, selective angiography has been reported to be useful in identifying the blood vessels supplying the tumor and in reducing blood loss during surgery, and venous pyelography is also useful for deciding whether to perform a total nephrectomy (Zeng et al., 2017).

Surgical resection is the mainstay of treatment for retroperitoneal LPS. Complete resection of the tumor has been reported as the most consistent prognostic factor (Zhuang et al., 2021; Xiao et al., 2021; Schwarzbach et al., 2006). A report of 500 retroperitoneal sarcoma cases indicated that the median survival for patients who underwent complete resection was 103 months compared to 18 months in those with incomplete resection (Lewis et al., 1998). However, resection of other organs such as the intestines, kidneys, and blood vessels is often required for complete surgery (Xiao et al., 2021). Previous reports on retroperitoneal sarcoma indicated that the concurrent resection rates of colon, pancreas, unilateral kidney, or vascular resection were 57%, 9.0–12.3%, 4.0–55.4%, and 17.7%, respectively (Zhuang et al., 2021; Xiao et al., 2021; Schwarzbach et al., 2006). The median operative time is 4 (range, 1–12) hours, blood loss is 500 (range, 20–13,000) g, and the post-operative intensive care unit admission rate is 69% (Zhuang et al.,

2021). Therefore, surgical resection entails a high risk, and preoperative evaluation is crucial for surgical treatment. The beneficial evidence for pre- and postoperative radiotherapy or chemotherapy is limited. The only randomized trial (STRASS EORTC 62092) comparing preoperative radiotherapy plus surgery with curative surgery alone in non-metastatic retroperitoneal sarcoma demonstrated the beneficial effects of preoperative radiotherapy (Grabenbauer, 2021). In contrast, a recent study that analyzed 607 patients with retroperitoneal LPS reported that perioperative radiotherapy was not associated with prognosis (Bachmann et al., 2020). The response rate to doxorubicin, ifosfamide, trabectedin, eribulin, pazopanib, and other antitumor agents is approximately 20% for soft tissue sarcoma (Yokoyama et al., 2020), and some case reports have shown significant efficacy of several chemotherapy regimens for retroperitoneal dedifferentiated LPS, such as the combination of doxorubicin and ifosfamide, cisplatin and ifosfamide, and eribulin alone (Yokoyama et al., 2020; Horowitz et al., 2020; Yokoi et al., 2009). However, no randomized controlled trial has investigated the efficacy of perioperative chemotherapy.

In the present case, we first suspected malignant transformation from a mature ovarian teratoma based on the suspicion of continuity between the tumor and the left ovarian vessels, the fatty component in the tumor, and elevated levels of CA125 and HE4. However, on reviewing the preoperative CT and MRI findings retrospectively, the tumor seemed to be in contact with the abdominal aorta, left common iliac artery, descending colon, and left iliopsoas muscle (Fig. 1B), suggesting a

retroperitoneal tumor. In addition, the fatty component and heterogeneous enhancement pattern in the tumor indicated a dedifferentiated LPS that underwent dedifferentiation from the well-differentiated component. Because of the discrepancy between the preoperative and postoperative diagnosis, incomplete resection of the tumor was performed, and partial resection of the sigmoid colon and left ureter was needed. Although adjuvant treatment was considered, it was not administered because of her age and PS.

In conclusion, retroperitoneal LPS should be considered in the case of a giant intra-abdominal tumor with a fatty component. Tumor location, size, origin, and the relationship between the tumor and adjacent tissues and organs should be carefully evaluated using imaging modalities.

Informed consent

Informed consent was obtained from the patient described herein.

CRedit authorship contribution statement

Akihiro Tani: Conceptualization, Methodology, Writing – original draft. **Yosuke Tarumi:** Conceptualization, Writing – review & editing. **Akiyo Kakibuchi:** Investigation. **Kohei Aoyama:** Investigation. **Tetsuya Kokabu:** Conceptualization, Investigation. **Hisashi Kataoka:** Conceptualization, Investigation. **Kaori Yoriki:** Conceptualization, Investigation. **Michiko Nagamine:** Supervision. **Taisuke Mori:** Supervision.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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