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Case Report

A Case of Cystic Retroperitoneal Dedifferentiated Liposarcoma Diagnosed by Percutaneous Image-Guided Biopsy

Kaede Morihara[a] Takahito Negishi[a] Nobuki Furubayashi[a] Hiroki Kobayashi[a] Mikako Jinnouchi[b] Kenji Shinozaki[b] Yutaka Koga[c] Kenichi Taguchi[c] Motonobu Nakamura[a]

[a]Department of Urology, National Hospital Organization Kyushu Cancer Center, Fukuoka, Japan; [b]Department of Diagnostic Imaging and Nuclear Medicine, National Hospital Organization Kyushu Cancer Center, Fukuoka, Japan; [c]Department of Pathology, National Hospital Organization Kyushu Cancer Center, Fukuoka, Japan

Keywords

Cystic liposarcoma · Retroperitoneum · Biopsy-proven dedifferentiated liposarcoma

Abstract

Introduction: Dedifferentiated liposarcoma (DDLP) was initially defined as a tumor containing differentiated liposarcoma and distinct regions of nonlipogenic spindle cell or pleomorphic sarcoma. Retroperitoneal liposarcomas feature a characteristic appearance with a predominantly fatty component, and cystic liposarcomas are rare. We describe a case of retroperitoneal DDLP predominantly consisting of multilocular cysts. ***Case Presentation:*** A 77-year-old man previously visited a doctor because an echo scan unexpectedly revealed an abdominal tumor. Contrast computed tomography (CT) disclosed a large multilocular cystic tumor spanning from the left upper abdomen to the retroperitoneum, and poorly marginated soft tissue structures were present around the abdominal aorta, inferior vena cava, pancreas, mesentery, and left kidney. CT also revealed a right lung mass. The soft tissue structures in the retroperitoneal cystic tumor and right lung mass were strongly enhanced on 2-deoxy-2-[fluorine-18] fluoro-D-glucose positron emission tomography, suggesting a malignant retroperitoneal tumor and lung metastasis. CT-guided percutaneous biopsy targeting the left perirenal soft tissue structure was performed, and the tumor was diagnosed as DDLP. Lung metastasis was present, and the retroperitoneal tumor surrounded multiple organs. Therefore, the tumor was not suitable for surgical resection

Correspondence to:
Takahito Negishi, n2takajin@gmail.com

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but it was indicated for chemotherapy based on multidisciplinary discussion. ***Conclusion:*** We experienced a case of retroperitoneal cystic DDLP diagnosed by percutaneous image-guided biopsy and treated appropriately based on the pathological diagnosis.

Introduction

Dedifferentiated liposarcoma (DDLP) was initially defined by Evans [1] in 1979 as a tumor containing differentiated liposarcoma and distinct regions of nonlipogenic spindle cell or pleomorphic sarcoma. DDLP is a high-grade and aggressive disease that most commonly arises in the retroperitoneum, followed by the limbs, trunk, and scrotum/vas deferens [2]. Retroperitoneal liposarcomas have a characteristic appearance with a predominantly fatty component, and cystic liposarcomas are rare [3]. In this study, we described a case of retroperitoneal DDLP predominantly consisting of multilocular cysts. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000535072).

Case Report

A 77-year-old man previously visited a doctor because an echo scan performed by a local physician unexpectedly revealed an abdominal tumor. Contrast computed tomography (CT) uncovered a large multilocular cystic retroperitoneal tumor that extended under the diaphragm to the lower pole of the left kidney and surrounded the stomach, spleen, pancreas, left kidney, and abdominal aorta (Fig. 1a–c). The liver was widely bordered, and poorly marginated soft tissue structures were present around the abdominal aorta, inferior vena cava, pancreas, mesentery, and left kidney (Fig. 1b, c). CT also revealed a right lung mass (Fig. 1a). The soft tissue structures in the retroperitoneal cystic tumor and right lung mass were strongly enhanced on 2-deoxy-2-[fluorine-18] fluoro-D-glucose positron emission tomography, suggesting a malignant retroperitoneal tumor and lung metastasis (Fig. 1d, e). The patient underwent endoscopic ultrasound-guided fine needle aspiration through the stomach and duodenum, targeting the soft tissue component at the hepatic helium, but it failed to provide a definitive diagnosis. The patient was subsequently referred to our hospital for further examination. Contrast CT performed 1.5 months after the previous examination illustrated that the soft tissue structures were generally progressing. CT-guided percutaneous biopsy targeting the left perirenal soft tissue structure, which was surrounded by cystic masses, was conducted (Fig. 1f). Histopathological finding revealed the proliferation of spindle- to polygonal-shaped tumor cells with enlarged hyperchromatic nuclei arranged in a haphazard pattern (Fig. 2a). Immunohistochemically, the tumor cells were positive for MDM2 (Fig. 2b) and CDK4 (Fig. 2c). We diagnosed the tumor as compatible with DDLP. Chemotherapy was initiated as described in a clinical trial.

Discussion

Retroperitoneal sarcomas comprise a distinct subgroup, accounting for <10% of all sarcomatous tumors [4]. They include liposarcoma, leiomyosarcoma, undifferentiated sarcoma, solitary fibrous tumor, malignant peripheral nerve sheath tumor, and fibrosarcoma [4].

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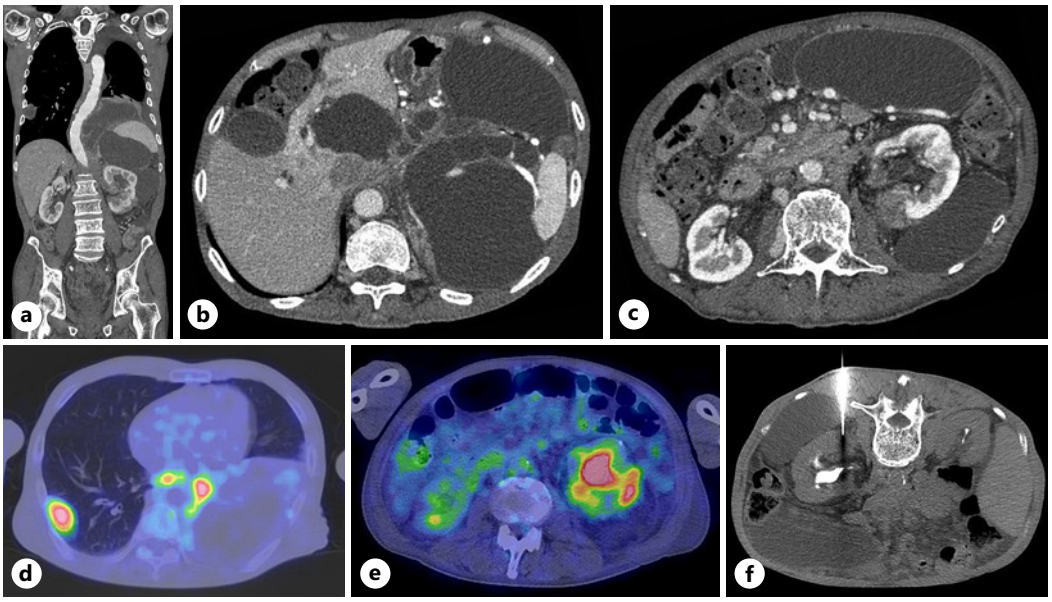

Fig. 1. Coronal image (**a**) and axial images (**b**, **c**) showed a large multilocular cystic tumor which extended under the diaphragm into the pelvis and surrounded the left kidney and spleen. The right lung mass (**d**) and the soft tissue structures in the retroperitoneal cystic tumor (**e**) were strongly enhanced by FDG-PET. CT-guided percutaneous biopsy-targeted left perirenal soft tissue structure was underwent in prone position (**f**).

Among them, liposarcoma is the most common primary retroperitoneal sarcoma, accounting for 40% of cases [5]. Retroperitoneal liposarcoma is typically a large encapsulated mass containing variable amounts of fatty and soft tissue components [6]. Well-differentiated liposarcoma is a low-grade tumor, the characteristic CT findings of which include macroscopic fat comprising >75% of the tumor [5]. Conversely, DDLP has areas of high-grade poorly differentiated sarcoma [3], which exhibits increased density and contrast enhancement on CT [7], within, adjacent to, or surrounding a fatty mass [6, 8]. In the present case, the tumor mainly consisted of a cystic mass without a fatty component and a small solid component. To our knowledge, four other cases of cystic retroperitoneal liposarcoma have been reported [9–12].

To obtain a pathological diagnosis of retroperitoneal tumor, image-guided tumor biopsy is routinely recommended [4, 13]. Among the four previously reported cases of retroperitoneal cystic liposarcoma, 2 patients underwent biopsy, and they received a provided histological diagnosis of high-grade sarcoma before surgery [10, 11]. All 4 patients underwent tumor resection surgery [9–12]. Despite the guideline recommendation, percutaneous biopsy of cystic tumors can be difficult because cystic tumors often have few solid components or solid components are sometimes located in an area that is anatomically challenging for biopsy. All four tumors were histologically diagnosed as well-differentiated/DDLP based on the features of surgically resected specimens. In the present case, CT-guided percutaneous biopsy could access the perirenal solid component in a prone position and successfully provide a histological diagnosis of DDLP without tumor resection. MDM2 and CDK4 expressions in tumor cells are almost invariably observed and also allow separation of DDLP from pleomorphic liposarcoma [14]. The diagnosis was made as DDLP was based on the results of MDM2 and CDK4 immunohistochemistry. This was the first case of cystic retroperitoneal DDLP that could be diagnosed without tumor resection.

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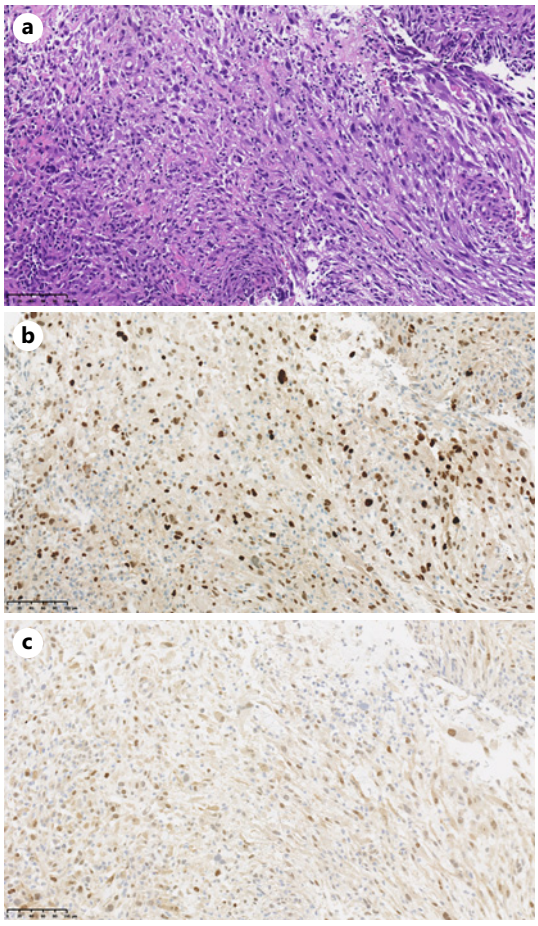

Fig. 2. Histopathology revealed the proliferation of spindle- to polygonal-shaped tumor cells with enlarged hyperchromatic nuclei arranged in a haphazard pattern (**a**). Immunohistochemically, the tumor cells were positive for MDM2 (**b**) and CDK4 (**c**).

The differential diagnoses of cystic retroperitoneal tumors include neoplastic and non-neoplastic tumors, such as mucinous cystadenoma and cystadenocarcinoma, cystic mesothelioma, lymphangioma, Müllerian cysts, epidermoid cysts, and pancreatic pseudocysts [5]. Most of these tumors are benign. In the present case, the cystic retroperitoneal tumor had a possibility of malignancy because it had solid components, which grew over time, and enhancement was present on 2-deoxy-2-[fluorine-18] fluoro-D-glucose positron emission tomography. Therefore, CT-guided percutaneous biopsy was performed, and the histological diagnosis was successfully obtained.

A chemotherapy protocol currently being investigated in a clinical study was administered to the patient without surgery. Histological cystic change of liposarcoma was presumably attributable to necrosis, lysis, hemorrhage, or ischemia of the tumor, according to previous case reports [9–12]. Further histological investigation of cystic lesion was not completed because surgery was not performed. In conclusion, we experienced a case of retroperitoneal cystic DDLP diagnosed by percutaneous image-guided biopsy and treated appropriately based on the pathological diagnosis.

Acknowledgment

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Statement of Ethics

The present study was approved by the Institutional Review Board of National Hospital Organization Kyushu Cancer Center (2014-99). Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

K.M. and T.N. drafted the manuscript. N.F., H.K., M.J., K.S., Y.K., K.T., and M.N. revised the manuscript critically for important intellectual content. All authors acquired the data and read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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