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Case Report

Undifferentiated Pleomorphic Sarcoma Originating in the Porta Hepatis: Report of a Rare Case

Sho Okimoto, Tsuyoshi Kobayashi, Shintaro Kuroda, Hirotaka Tashiro, Hideki Ohdan

Department of Gastroenterological and Transplant Surgery, Applied Life Sciences, Institute of Biomedical & Health Sciences, Hiroshima University, Hiroshima, Japan

Undifferentiated pleomorphic sarcoma (UPS) is a soft tissue sarcoma in adults. Although it can occur in any part of the body, its most common site is the extremities, followed by the trunk and retroperitoneum. Here, we describe a rare case of UPS originating in the porta hepatis of an 83-year-old man that was incidentally detected via contrast-enhanced computed tomography. Because malignancy was suspected, we performed a tumor resection with cholecystectomy. Consistent with a diagnosis of UPS, histopathology revealed highly cellular lesions with scattered pleomorphic spindle cell fascicles arranged in a storiform pattern; further, results of tests for all immunohistochemical markers were negative. One year after surgery, the patient is currently doing well without any evidence of tumor recurrence. To our knowledge, this is the first report of UPS originating in the porta hepatis.

Key words: Undifferentiated pleomorphic sarcoma – Hilar tumor – Malignant fibrous histiocytoma – Soft tissue sarcoma – Gallbladder

Undifferentiated pleomorphic sarcoma (UPS) is a soft tissue sarcoma formerly termed malignant fibrous histiocytoma (MFH). It has an aggressive behavior with a high potential for local recurrence and distant metastasis. The standard treatment for UPS is radical surgery.

The most frequent site of UPS is the extremities, followed by the trunk and retroperitoneum, and only a few cases of UPS at other sites have been reported.[1–8] UPS originating in the abdomen is rare, and primary UPS in the porta hepatis is extremely rare. Cases of primary UPS in the liver and gallbladder have been reported in the English literature[1–3,5] but not cases of UPS in the porta hepatis. In this report, we describe a case of UPS originating in the porta hepatis.

Corresponding author: Tsuyoshi Kobayashi, MD, Department of Gastroenterological and Transplant Surgery, Applied Life Sciences, Institute of Biomedical & Health Services, Hiroshima University, 734-8551, 1-2-3, Kasumi, Hiroshima, Japan.
Tel.: +81 82 257 5222; Fax: +81 82 257 5224; E-mail: tsukoba@hiroshima-u.ac.jp

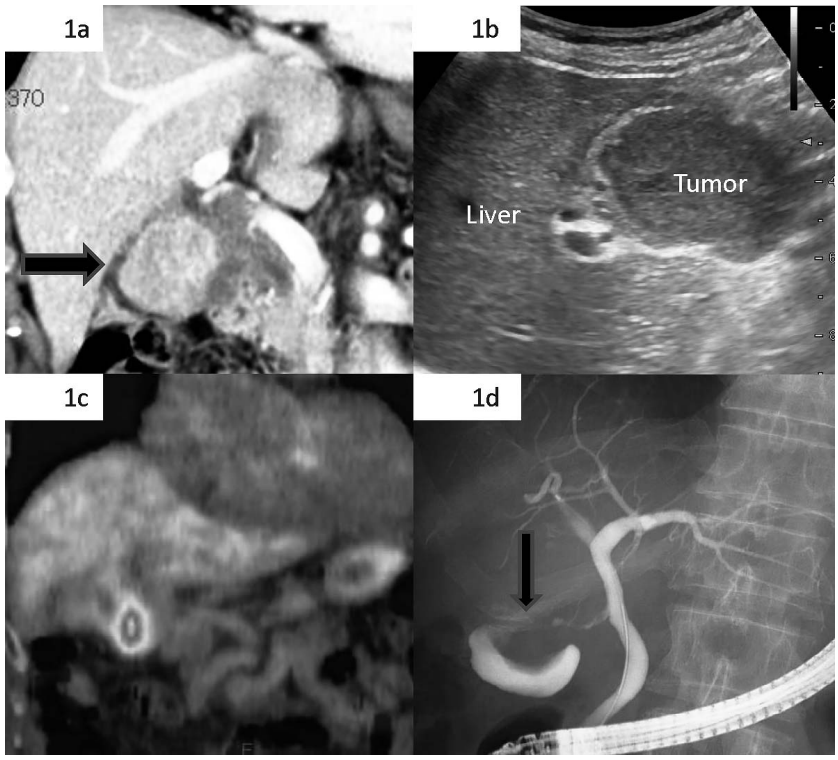

Fig. 1 (a) Contrast enhanced abdominal computed tomography shows a 50-mm enhanced tumor in the porta hepatis without dilatation of the intra- or extra-hepatic bile duct. (b) Abdominal ultrasound sonography revealed the presence of a solid tumor in the porta hepatis, and the border of the porta hepatis and the liver was described clearly. (c) Positron emission tomography-CT shows an accumulation of fluorodeoxyglucose (^{18}F) in the tumor; the standardized uptake value was 5.0. (d) Endoscopic retrograde cholangiography shows the tumor pressing the gallbladder without appreciably affecting the bile duct.

Case Report

An 83-year-old man underwent a lung lobectomy for a lung adenocarcinoma (stage 2B). Three years later, contrast-enhanced computed tomography (CT) performed at a follow-up visit revealed a 50-mm enhanced tumor in the porta hepatis without dilatation of the intra- or extra-hepatic bile ducts (Fig. 1a). He was referred to our hospital for further examination and surgical treatment.

A physical examination showed that the mass was not palpated, and there was no tenderness. Laboratory tests showed results that were almost within the normal range, and expression levels of tumor markers such as the interleukin-2 receptor, the carcinoembryonic antigen, and carbohydrate antigen 19–9 were normal. Abdominal ultrasound sonography revealed the presence of a solid tumor in the porta hepatis, and the border of the porta hepatis and the liver was described clearly (Fig. 1b). Positron emission tomography-CT showed an accumulation of fluorodeoxyglucose (^{18}F) in the tumor; the standardized uptake value was 5.0, and there was no accumulation at any other site (Fig. 1c). Endoscopic retrograde cholangiography and abdominal ultrasound sonography showed that the tumor pressed the gallbladder but did not appreciably affect the bile duct (Fig. 1d).

After the examinations, a tumor resection with cholecystectomy was performed. Laparotomy findings revealed that the tumor was located in the hepatoduodenal ligament at the back of the gallbladder (Fig. 2a) and had directly invaded the gallbladder.

The resected tumor was double mass-like and white and brown in color (Fig. 2b). The mucosal layer of the gallbladder had no remarkable changes. A histopathologic study revealed a highly cellular lesion with scattered pleomorphic spindle cell fascicles arranged in a storiform pattern and frequent mitoses (5 to 10 per high power field, Fig. 2c, 2d). The results of tests for immunohistochemical markers S-100, α-smooth muscle actin, CD99, BCL-2, H-caldesmon, synaptophysin, neuron-specific enolase, CD34, and c-kit were all negative (Fig. 3). The final pathologic diagnosis was undifferentiated pleomorphic sarcoma with invasion of the gallbladder. The postoperative course was uneventful, and recurrence was not detected for at least a year.

Discussion

UPS is the most common soft tissue sarcoma in adults. It has a slight male predominance. It can develop in any part of body but mainly occurs in the

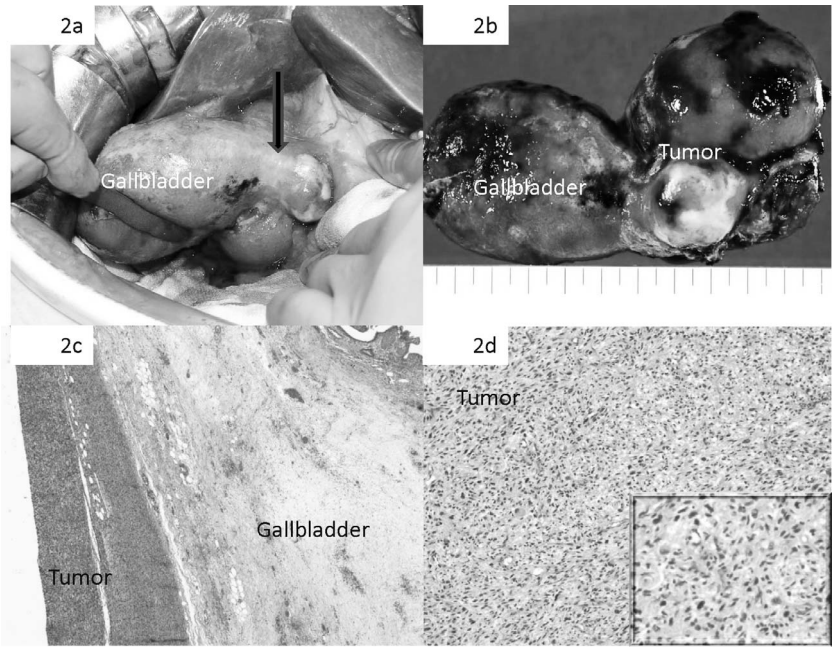

Fig. 2 Laparotomy findings. (a) The tumor was located in the hepatoduodenal ligament on the back of the gallbladder. (b) The resected specimen is double mass-like and white and brown in color. The mucosal layer of the gallbladder had no remarkable changes. (c, d) The histopathologic study reveals a highly cellular lesion with pleomorphic spindle cells fascicles arranged in a storiform pattern in places and frequent mitoses (5 to 10 per high power field).

extremities, followed by the trunk and retroperitoneum.[9] Although a few cases of UPS originating in the liver and gallbladder have been reported,[1–3,5] this is the first report of a UPS originating in the porta hepatis in the English literature.

Because UPS has no specific features, either clinical or radiologic, and because there are no typical symptoms, its diagnosis is not easy.[10] When UPS develops in any part of the body other than the extremities and trunk, its diagnosis is even more difficult. Therefore, UPS is often found incidentally, and tumor resection is commonly undertaken despite suspicion of a malignant tumor. In our case, the patient had no symptoms, and the UPS was detected incidentally as a mass on a contrast-enhanced CT scan at a follow-up visit after surgery for a lung adenocarcinoma. Before the operation, we thought it might be a metastasis of the lung cancer, a gallbladder cancer, a malignant lymphoma, a schwannoma, or other type of sarcoma.

UPS was previously termed MFH. The number of diagnoses of UPS has decreased in the last decade because advances in diagnostic implements provide more precise classifications. Cases previously diagnosed as MFH are now classified as sarcoma subtypes such as liposarcoma and fibrosarcoma.[9] UPS is a type of sarcoma with an uncertain origin and is considered a diagnosis of exclusion for sarcomas that cannot be precisely categorized. An accurate diagnosis of UPS depends on an accurate pathologic diagnosis based in part on the results of immunohistochemistry. UPS has no specific markers and therefore is negative for most antibody panels used for tumor classification. In our case, the UPS consisted of undifferentiated pleomorphic cells and neoplastic spindle cells arranged in a storiform pattern. It was negative for S-100, α-smooth muscle actin, CD99, BCL-2, H-caldesmon, synaptophysin, neuron-specific enolase, CD34, and c-kit.

Because UPS has an aggressive behavior with a high potential for local recurrence and distant metastasis, its prognosis is poor. The standard treatment for UPS is radical tumor resection, whose goal is eradication of all disease from the affected area.[9] The effects of chemotherapy and radiotherapy on UPS tumors are unclear. The chemotherapy drug, doxorubicin, may improve prognosis, and the radiation therapy may prevent local tumor recurrence.[9]

In surgical therapy, removing the tumor and maintaining function are both important. For UPS in the extremities, there are 2 surgical options: amputation and limb-sparing. From an oncologic standpoint, UPS tumors should be removed via amputation; however, if the tumor can be completely excised, limb-sparing may be a more compassionate choice. For UPS in the abdomen, there are multiple surgical procedures depending on the location of the tumor, and a standard procedure has not established. Use of wide resection for abdominal UPS tumors is still debatable. In our case, we selected tumor resection

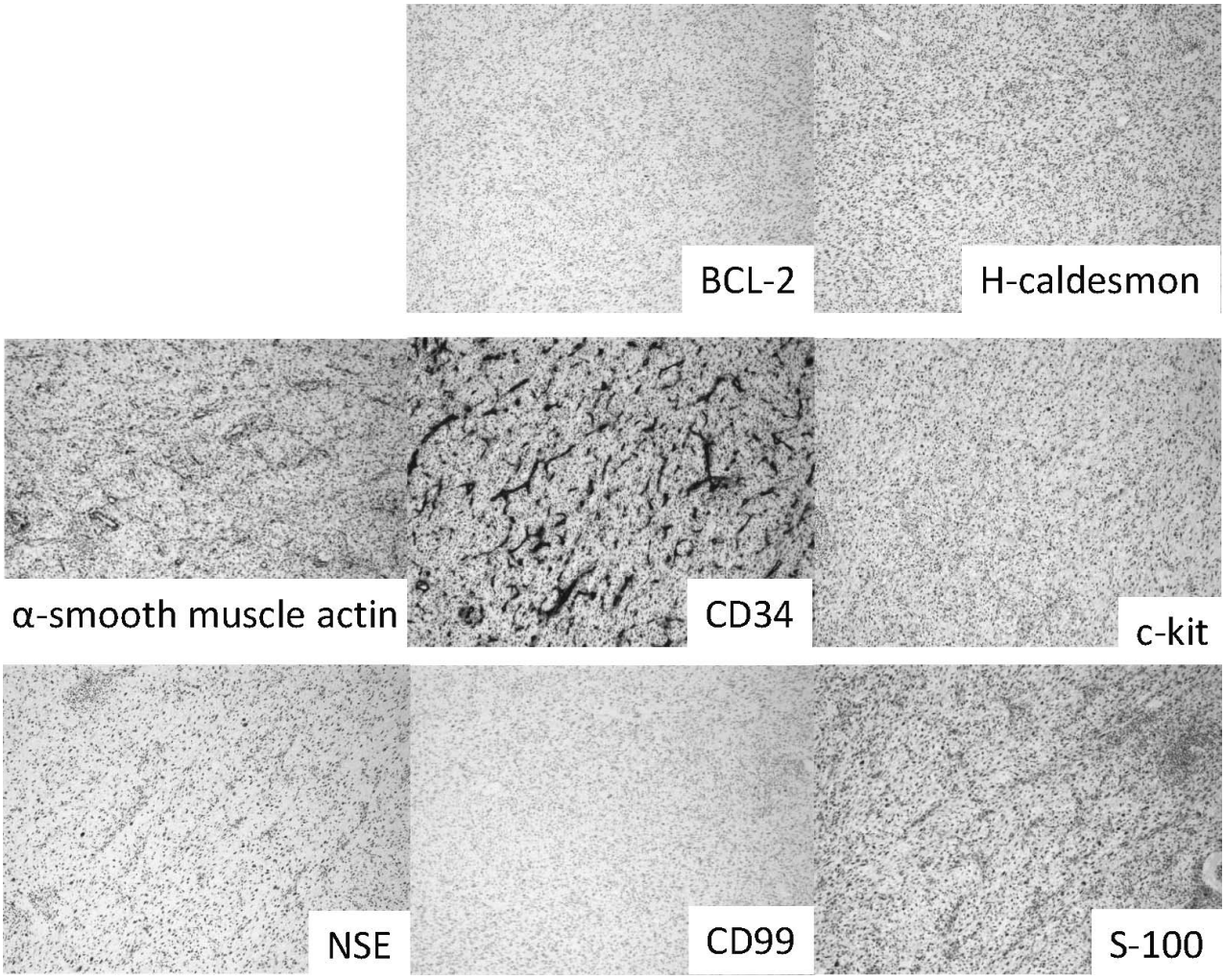

Fig. 3 Immunohistochemical markers were all negative.

as the sole treatment. Complicated ablations of bile ducts and vessels of the hepatoduodenal ligament and lymph node dissection were not done. When removal of the other organs with tumor resection is necessary, procedures become more invasive. Therefore, with respect to UPS tumors, the surgical method should be decided carefully with discussion about the advantages and drawbacks of all options.

In conclusion, we describe the first case of a UPS originating in the porta hepatis. The tumor directly invaded the gallbladder and was adjacent to the liver. On the basis of an intraoperative examination and the pathology of an excised specimen, our diagnosis was a UPS tumor originating in the porta hepatis and hepatoduodenal ligament with invasion of the gallbladder.

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