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Recurrent Hemangiopericytoma of the Liver: Report of a Case

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Abstract: We present herein the case of a 63-year-old man with a locally recurrent hemangiopericytoma in the hepatic hilum. The patient presented to our hospital for the investigation of elevated transaminases 6 years after undergoing resection of a hemangiopericytoma followed by chemotherapy at another hospital. Various imaging techniques demonstrated a well-circumscribed, hypervascular tumor with central necrosis in the hepatic hilum. Establishing a preoperative diagnosis would have been difficult without the information that he had previously had a hemangiopericytoma of the abdominal cavity. An extended right hepatic lobectomy was performed and histopathological examination confirmed the diagnosis of recurrent hemangiopericytoma. Although hemangiopericytoma is a rare tumor, the clinical presentation and radiologic features of this case were fairly typical.

Key Words: liver neoplasms, recurrence

Introduction

Hemangiopericytoma is an uncommon tumor of the soft tissue, first described in 1942 by Stout and Murray,[1] which originates from a pericyte, a spindle-shaped cell situated around the capillaries. Although the pathologic entity of hemangiopericytoma has been established, the diagnosis is still difficult pathologically and radiologically due to the existence of many hemangiopericytoma-like neoplasms. The treatment of choice is complete resection of the tumor, which is often complicated by severe hemorrhage, and even after an apparent complete resection, recurrence is encountered rather frequently. This case report presents the clinical characteristics of hemangiopericytoma and discusses the problems in diagnosing and treating patients with this tumor.

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Case Report

A 63-year-old man had a large abdominal mass resected at another hospital at the age of 57, the pathological diagnosis of which was confirmed to be hemangiopericytoma. He received a course of chemotherapy with mitomycin C, 5-fluorouracil (5-FU), cytosine arabinoside, and doxorubicin after the operation but then stopped visiting the hospital for follow-up. He presented to Nagoya Daini Red Cross Hospital for the evaluation of elevated transaminases which were noted on a routine annual examination. Ultrasonography (US) revealed a hepatic tumor and he was hospitalized. The laboratory data on admission showed moderately elevated serum levels of glutamic oxaloacetic transaminase, glutamic pyruvic transaminase, alkaline phosphatase, lactate dehydrogenase, γ-glutamyl transpeptidase, and leucine aminopeptidase. The tumor markers, including α-fetoprotein, carcinoembryonic antigen, protein-induced vitamin K antagonist-2, carbohydrate antigen 19-9 (CA 19-9), CA125, and other laboratory data were within normal limits.

Abdominal ultrasound revealed a well-circumscribed tumor with hypoechoic areas, compressing the anterior and posterior branches of the portal vein, but there were no signs of vascular invasion (Fig. 1a). Plain computed tomography (CT) showed a low-density tumor in the hepatic hilum measuring 8 cm in diameter with early enhancement after a bolus injection of contrast medium, although a central low-density area consistent with necrosis did not become enhanced (Fig. 1b). Magnetic resonance imaging (MRI) demonstrated a hepatic tumor less intense than the hepatic parenchyma on T1-weighted images and hyperintense on T2-weighted images (Fig. 1c), being partially enhanced

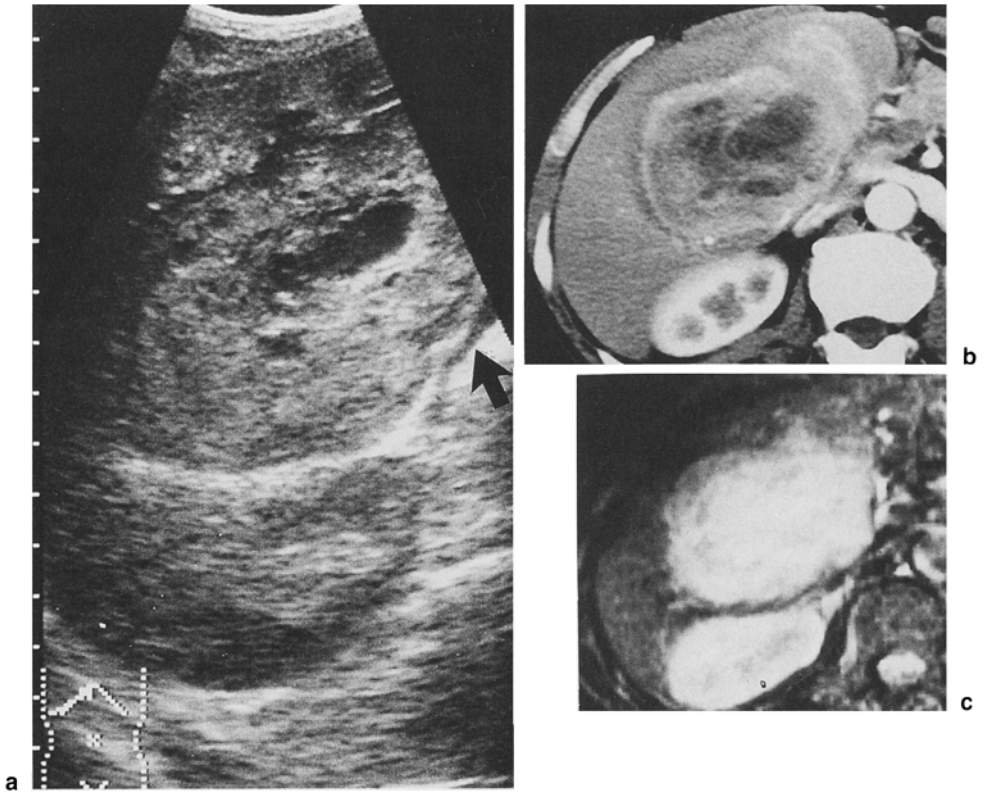

Fig. 1. **a** Ultrasonography reveals a well-circumscribed, heterogeneous tumor, 8 cm in diameter, in the hepatic hilum, compressing the posterior branch of the portal vein (*arrow*). **b** Dynamic computed tomography. The tumor was enhanced by contrast medium in the rapid phase, (35 s after contrast injection) except for a central area consistent with necrosis. **c** Magnetic resonance imaging of the T2-weighted image

by the administration of gadolinium-diethylenetriamine pentaacetic acid. An upper gastrointestinal series demonstrated an inferiorly displaced duodenum and a celiac angiogram revealed tumor vessels derived from the hepatic artery. In the capillary phase of the angiogram, an accumulation of contrast medium demonstrated a clearly delineated tumor. The anterior branch of the right hepatic artery was absent, but this was considered to be an artifact caused by the previous operation because that part of the artery was present on the angiogram taken before the first operation and the portal counterpart was patent (Fig. 2).

At laparotomy, a tumor was found in the hepatic hilum, part of which protruded over the surface of the liver and so an extended right hepatic lobectomy was performed. The cut surface of the tumor was grayish-yellow with liquid necrosis (Fig. 3a). Microscopically the tumor consisted of spindle-shaped cells tightly packed around staghorn-like capillaries. The nuclei of the tumor cells were slightly irregular in size and shape with scarce mitotic figures (Fig. 3b). In some peripheral parts of the tumor, an invasion of tumor cells into the capsule was noted. With silver staining, tumor cells were observed outside the vascular basement membrane surrounded by reticulin fibers (Fig. 3c), but staining for factor VIII was negative. These pathologic findings were compared with those of the previously resected tumor and the diagnosis of a recurrent hemangiopericytoma was confirmed.

The patient had an uneventful postoperative course and remains in good health with no evidence of recurrence 19 months after his second operation.

Discussion

Hemangiopericytoma was first described by Stout and Murray in 1942.[1] It is an uncommon tumor, constituting 5% of all soft tissue sarcomas and is considered to originate from the pericytes which are contractile cells surrounding the capillaries and postcapillary venules. The tumor can present in patients of any age and in any part of the body; however, the most common presentation is in the lower extremities and retroperitoneum of adults in the fifth to sixth decade of life.[2] The records of the first operation in our patient indicated that the tumor had a peduncle on the hepatoduodenal ligament with invasion to the liver and gallbladder. Therefore, the primary site was considered to be the hepatoduodenal ligament, with local recurrence at the site of hepatic invasion.

Hemangiopericytomas have some interesting clinical features. One of these features is the rate of recurrence which is as high as 52% of cases, mostly in the lungs,

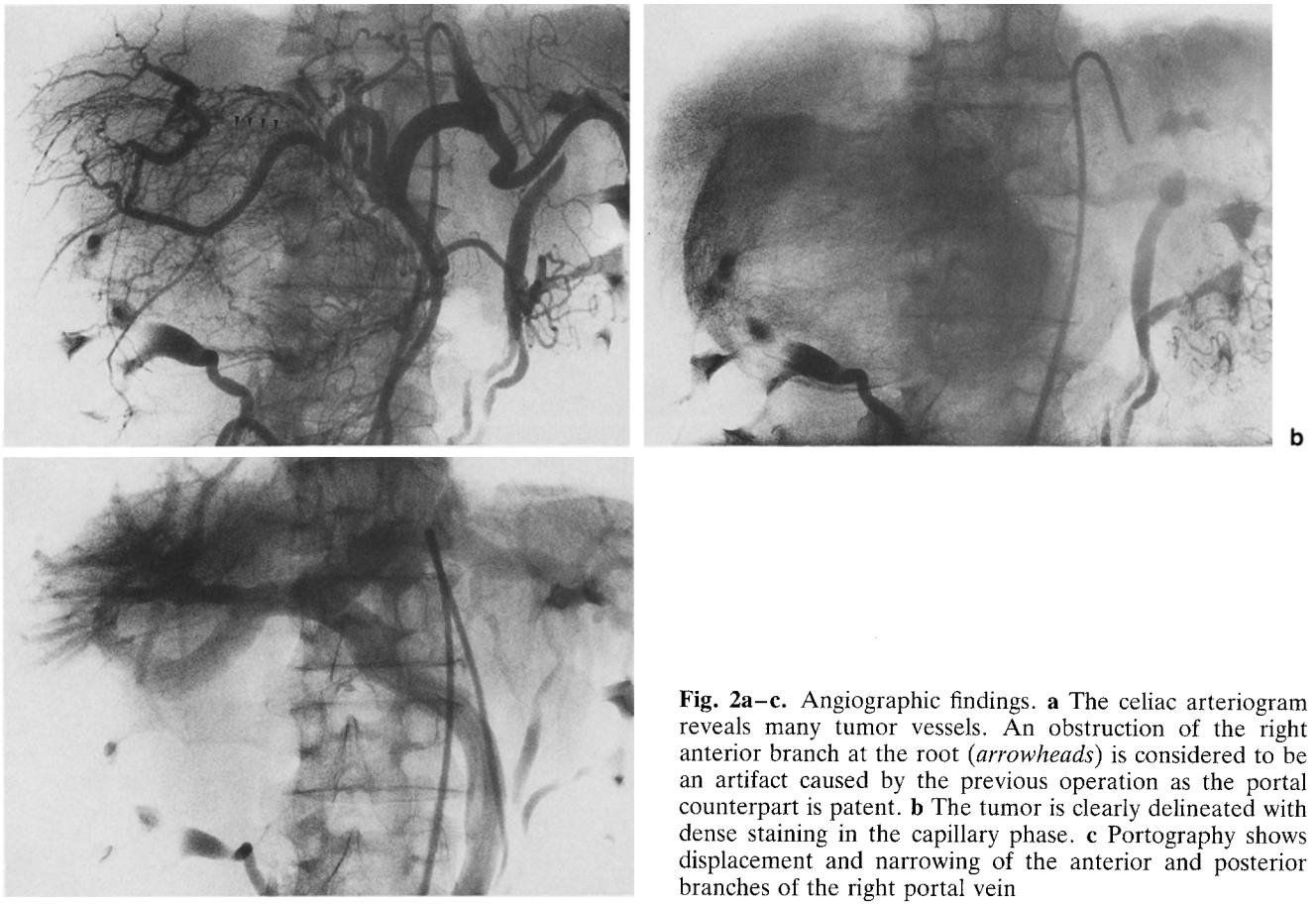

Fig. 2a–c. Angiographic findings. **a** The celiac arteriogram reveals many tumor vessels. An obstruction of the right anterior branch at the root (*arrowheads*) is considered to be an artifact caused by the previous operation as the portal counterpart is patent. **b** The tumor is clearly delineated with dense staining in the capillary phase. **c** Portography shows displacement and narrowing of the anterior and posterior branches of the right portal vein

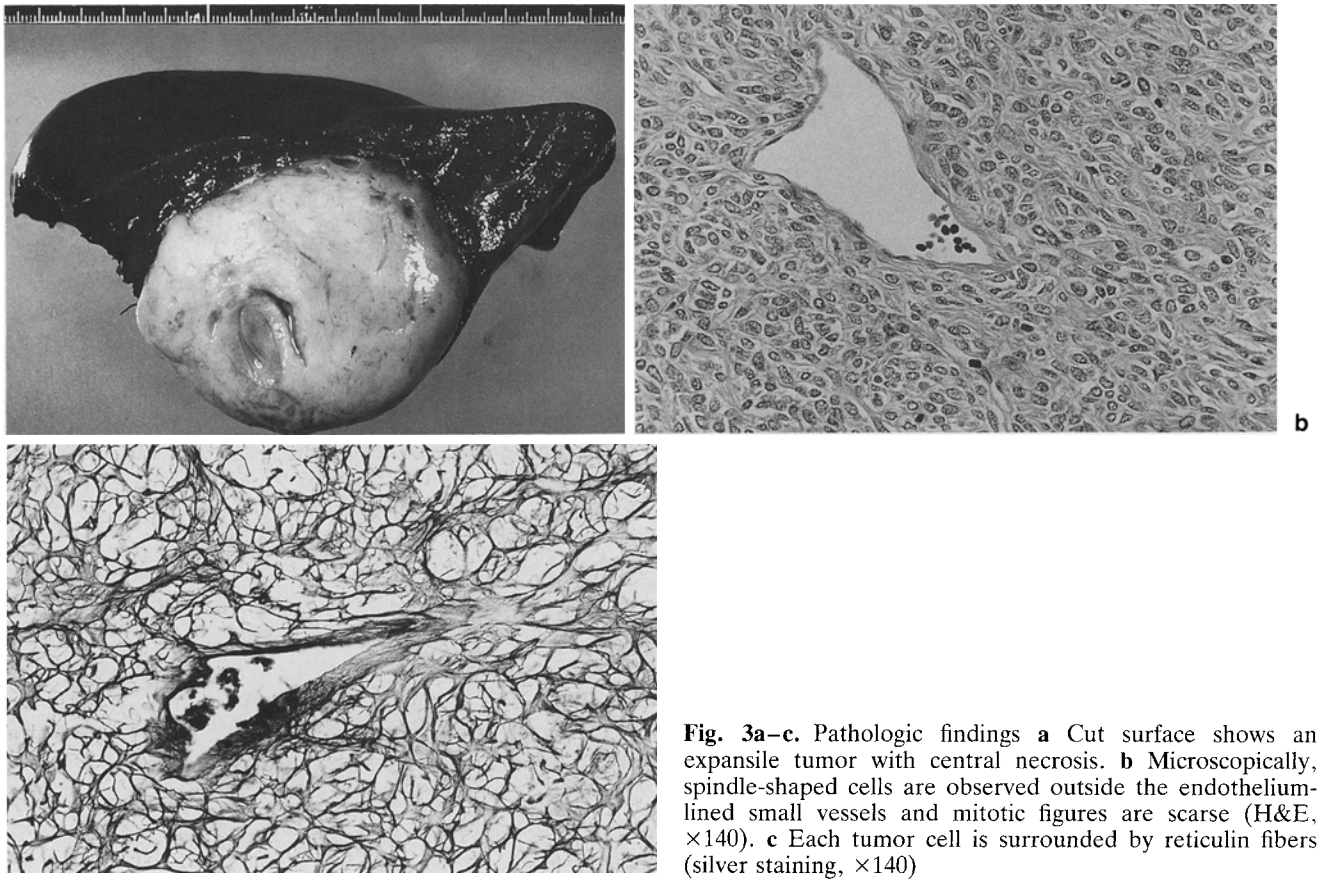

Fig. 3a–c. Pathologic findings **a** Cut surface shows an expansile tumor with central necrosis. **b** Microscopically, spindle-shaped cells are observed outside the endothelium-lined small vessels and mitotic figures are scarse (H&E, ×140). **c** Each tumor cell is surrounded by reticulin fibers (silver staining, ×140)

liver, and regional lymph nodes, necessitating long-term follow-up after resection of the primary tumor. Another interesting feature is the various paraneoplastic symptoms accompanying this neoplasm, including hypoglycemia[4–7] and hypertension.[8] Our patient originally presented with hypoglycemic symptoms including a blood glucose level of 23 mg/dl on his previous admission, which normalized after resection of the tumor. Several mechanisms, such as the secretion of insulin-like substances and the hyperutilization of glucose, which were suspected in our patient, have been proposed as possible causes of tumor-related hypoglycemia. A review of the literature revealed that the size of a tumor causing hypoglycemic symptom ranges from 12 cm to 27 cm. In our patient, the size of the primary tumor was 22 × 18 × 10 cm and that of the recurrent tumor was 8 × 7 cm. During his second hospitalization, the glucose tolerance test showed no abnormalities, probably because the recurrent tumor was not large enough to induce this symptom.

There are three important issues concerning the histologic diagnosis of this neoplasm. The first relates to the pathologic entity of hemangiopericytoma. As opposed to Stout and Murray's view of the pericyte as a distinct cell, several ultrastructural and immunohistochemical studies support the concept that the pericyte is differentiated from primitive mesenchymal cells, and endothelial, myoid, and fibrohistocytic directions may be other lines of potential differentiation.[9,10] This concept, although still controversial, can explain the morphologic variety of this tumor. The second issue addresses the differential diagnosis of hemangiopericytoma. There are many types of tumors with hemangiopericytoma-like vascular patterns, such as fibrous histiocytoma, synovial sarcoma, chondrosarcoma, liposarcoma, and leiomyosarcoma,[2] which can all be potentially misdiagnosed as hemangiopericytoma. The third issue concerns the malignant nature of this tumor. Many authors have tried to predict the malignant behavior of individual tumors by histologic and radiologic criteria,[11] including the presence of increased cellularity, prominent mitotic activity, and foci of necrosis or hemorrhage.[2] However, there appears to be a high rate of recurrence, regardless of the histologic appearance.

Various imaging techniques reveal hemangiopericytomas to be hypervascular tumors with central necrosis.[12–14] Angiographically, hemangiopericytomas are reported to have feeder arteries in a pedicle, with radial branching wrapping around a well demarcated, longstanding tumor containing fine unrecognizable capillaries.[15] However, these features are not specific enough to make a definite diagnosis of hemangiopericytoma and therefore, it is impossible to distinguish hemangiopericytoma from other hypervascular tumors by imaging modalities.

The treatment of choice is surgical resection, while chemotherapy and radiotherapy[16,17] are also considered useful for residual or metastatic tumors, although the paucity of cases makes it difficult to evaluate the effectiveness of these modalities.

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