Neurilemmoma Mimicking a Multilocular Cystic Lesion of the Liver

Akihiro Yoshida, Kentaro Yamao, Mamoru Takenaka, Atsushi Nakai, Shunsuke Omoto, Ken Kamata, Kosuke Minaga, Takeshi Miyata, Hajime Imai, Ippei Matsumoto, Yoshihumi Takeyama, Takaaki Chikugo and Masatoshi Kudo

Abstract:
Neurilemmomas are benign tumors arising from the sheaths of peripheral nerves. They appear rarely in the abdominal cavity. We herein report an 80-year-old man with a multilocular cystic neurilemmoma mimicking a liver lesion. Preoperative images showed a lesion in the porta hepatis. Although a preoperative diagnosis was difficult, surgery was undertaken because of the possibility of malignancy. Histologically, the tumor consisted of spindle-shaped cells with positivity for S-100 protein. The final diagnosis was a neurilemmoma. Porta hepatic neurilemmomas are rare. When we encounter a multilocular cystic lesion of the liver, neurilemmoma should be considered in the differential diagnosis.

Key words: multiple liver cyst, neurilemmoma, porta hepatis, right hepatic artery

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Introduction

Neurilemmomas are benign neurogenic tumors that arise from the sheaths of peripheral nerves. The most common sites of neurilemmomas are in the head, neck, and extremities, although they rarely occur in the abdominal cavity, including the porta hepatis. The periportal region is anatomically complex because it includes the area between the superior aspect of the first portion of the duodenum and the porta hepatis, along with the hepatoduodenal ligament and extrahepatic bile duct, portal vein, hepatic artery, autonomic nerve fibers, and lymph nodes. Therefore, diagnosing and identifying the origin of a tumor that occurs in this area can be challenging, as it may originate from the liver, bile duct, pancreas, and retroperitoneum.

Neurilemmomas are mostly solid tumors, but rarely they appear as a solitary cystic tumor. These solitary cystic tumors of the liver are of various types (e.g., benign simple cysts and hamartomas, malignant cystadenocarcinomas, and cystic hepatocellular carcinomas). When diagnostic imaging studies are inconclusive, a fine-needle biopsy may be necessary. A biopsy for cystic lesions is not recommended, however, because of the risk of seeding. In such cases, surgical excision with a histopathological evaluation may be required.

Although it is rare to find neurilemmomas in the porta hepatis, we herein report a patient in whom a multilocular cystic neurilemmoma mimicked a hepatic lesion of the liver.

Case Report

An 80-year-old man was referred to our hospital because of what appeared to be an enlarging pancreatic cystic lesion that caused slight abdominal pain. His history included hypertension and reflux esophagitis, with no remarkable family history. The results of blood tests showed no abnormalities: total bilirubin 0.5 mg/dL, aspartate transaminase 26 IU/L, alanine transaminase 144 U/L, γ-glutamyltranspeptidase 23 IU/L, amylase 129 U/L, carcinoembryonic antigen 1.2 ng/mL, carbohydrate antigen 19-9 8 U/mL.
Figure 1. Contrast-enhanced computed tomography shows a well-defined, 7.0 × 7.0-cm cystic lesion. The lesion was located between the portal vein (arrow) and the hepatic artery (arrow head), seemingly sandwiched between the two vessels.

Abdominal ultrasonography revealed a multilocular cystic lesion with numerous septa in the caudate lobe of the liver with no pancreatic contact. The septum of the cyst was clearly revealed by contrast-enhanced ultrasonography. Abdominal contrast-enhanced computed tomography (CT) revealed a well-defined, 7.0 × 7.0-cm, cystic lesion between the portal vein and hepatic artery, seemingly sandwiched between the two vessels (Fig. 1). Abdominal magnetic resonance imaging revealed that the entire multilocular cystic lesion was hypointense on T1-weighted imaging (T1WI), slightly hyperintense on T2WI, and hypointense on diffusion-weighted imaging. Part of the cyst, however, showed a fluid-fluid level suggesting the presence of old bleeding (Fig. 2). Endoscopic ultrasonography performed to determine the origin of the lesion suggested that it was derived from the liver because the lesion’s site was distant from the pancreas. Contrast-enhanced endoscopic ultrasonography revealed a mural nodule but no wall thickness (Fig. 3). Based on these imaging results, we deemed the differential diagnosis to include cystadenoma, mesenchymal hamartoma, ciliated hepatic foregut cyst, abdominal lymphangioma, cystic hamangioma, hepatocellular carcinoma, and cholangiocarcinoma, but we were unable to confirm the exact diagnosis.

Despite the lack of a definitive diagnosis, we resected the lesion because of the patient’s symptoms and the possibility of malignancy. Intraoperatively, the lesion was seen to arise from the hepatoduodenal ligament, although there was no adherence to either the liver or the pancreatic parenchyma. The lesion was connected to the tissue around the right hepatic artery via a short stalk. The cyst’s content was absorbed, and the serous, pale blood was removed.

A pathological examination of the specimen revealed a multilocular cystic lesion with central necrosis and hemorrhaging. Macroscopically, the tumor was clearly demarcated, white-gray, with focal cystic changes. Histologically, the tumor consisted of spindle-shaped cells and showed positivity for S-100 protein (Fig. 4). The final diagnosis was neurilemmoma.

The lesion included Antoni-A and Antoni-B areas. The Antoni-A area was composed of spindle-shaped cells in a palisading pattern. The Antoni-B area was composed of a myxoid component in which the tumor cellular density was relatively low. The nuclear palisading formation of the tumor exhibited Verocay bodies.

The patient’s postoperative course was uneventful. At six months postoperatively, he is well and has shown no signs of recurrence.

Discussion

A multilocular cystic neurilemmoma arising from the right hepatic artery is rare, and it was difficult in this case to make a preoperative diagnosis. Most neurilemmomas are derived from Schwann cells and are benign. They are commonly located in the head and neck (44.8%), upper extremities (19.1%), and lower extremities (13.5%) (1), although they can arise from any nerve trunk or organ, except for the olfactory and optic nerves, which do not contain Schwann cells (5). The literature on neurilemmomas occurring in the abdominal cavity is sparse, and that on those originating in the porta hepatis is even rarer.

In a review of 15 patients, the lesions were in the porta hepatis in 9 patients (60%), the hepatoduodenal ligament in 5 patients (33%), and the proper hepatic artery in 1 patient (7%) (6). The most commonly reported sites are the proper hepatic artery (7), hepatoduodenal ligament (8-12), and porta hepatitis (details unknown) (5, 6, 13-19). Many of the reported cases were difficult to diagnose preoperatively, with the (incorrect) preoperative diagnoses being focal nodular hyperplasia (17), Klatskin’s tumor (16), giant lymph node hyperplasia (12), cystadenoma (4), and abdominal lymphangioma (4). In almost all cases, a malignant tumor was suspected, and surgery was performed. As most neurilemmomas are benign, however, surgery is unnecessary if they are properly diagnosed. A fine-needle biopsy may be sufficient to gain a preoperative diagnosis, whereas a biopsy for cystic lesions is not recommended because of the risk of seeding. In some cases, surgical excision with a histopathological evaluation is required.

In the present case, a biopsy was not performed preoperatively and the imaging results were unclear. CT findings showed that part of the lesion was in contact with the caudate lobe and was located between the portal vein and hepatic artery, seemingly sandwiched between the two vessels. If the lesion had originated from the caudate lobe, the portal vein and hepatic artery would be compressed on one side with its growth. However, the lesion originated from the plexus around the right hepatic artery and, as it grew, became wedged between the portal vein and hepatic artery. Therefore, when considering the organ from which a lesion originates, we must not only look for characteristic findings, such as the bird’s beak sign, but also evaluate the lesion’s
Figure 2. A: Magnetic resonance imaging revealed that the lesion was entirely visualized as hypointense on T1-weighted imaging (T1WI). B: The lesion was slightly hyperintense on T2WI. The fluid-fluid level seen in some of these cysts suggests the presence of old bleeding.

Figure 3. Contrast-enhanced endoscopic ultrasonography shows the lesion with some cysts containing liquid and others with mucinous material, debris, and residue of old bleeding.

Figure 4. A: Hematoxylin and Eosin staining shows the tumor present at the margins, with cystic degeneration and old bleeding at the center. There was a mixture of densely and sparsely aligned spindle-shaped cells with strong expression. No ovarian stroma characteristic of a mucinous cystic neoplasm was observed. B: Immunostaining reveals that the tumor was S-100-positive.

relationship with the peripheral anatomy, including vessels.

Neurilemmomas appear as two morphological types: 90% are solitary, and 10% are cystic. A review of 15 neurilemmomas that originated from the porta hepatitis reported that 7 of the 15 lesions (47%) had cystic changes (6). Although neurilemmomas are generally classified as solid tumors, when the size of the lesion increases rapidly, degenerative necrosis or cystic degeneration often occurs within the tu-
The authors state that they have no Conflict of Interest (COI).

Microscopically, the hallmark of neurilemmoma is a pattern of alternating Antoni A and Antoni B areas, with varying relative amounts (22). Antoni A areas are hypercellular and characterized by closely packed spindle cells with occasional nuclear palisading and Verocay bodies, whereas Antoni B areas are hypocellular and occupied by loosely arranged tumor cells (23). Immunohistochemical staining of a schwannoma is strongly and diffusely positive for S-100 protein, consistent with the findings of a nerve sheath tumor (24).

In conclusion, porta hepatis neurilemmomas are rare. Making a correct diagnosis preoperatively is challenging because of the tumor’s rarity and its nonspecific imaging and clinical manifestations. When confronted with a multilocular cystic lesion with numerous septa arising in the porta hepatis, neurilemmoma should be considered in the differential diagnosis.

Informed consent was obtained from the patient to participate in this study. All procedures were performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

The authors state that they have no Conflict of Interest (COI).

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