A Case Report of Benign Schwannoma of the Liver

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Abstract
Benign schwannoma is a very rare confronted entity in the liver. Only a very few cases have been reported in the medical literature. A 56-year-old woman was admitted to our hospital with epigastric pain. In the computed tomography scan a cystic mass was observed in the liver. The mass was resected with a prediagnosis of hydatid cyst; intraoperatively a 15×10×10 cm mass filled with hemorrhagic fluid was found. The histological examination confirmed the diagnosis of a benign schwannoma, proven by verocay bodies and a positive immunoreaction with the neurogenic marker S-100 protein.

Key words: benign schwannoma, liver

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Introduction
A schwannoma, also called neurilemoma, is a benign tumor that originates from the Schwann cells of the nerve sheath. Schwannoma occurs at all ages but it is most common in persons who are >20 or <50 years old. The tumors have a predilection for the head, neck and flexor surfaces of the upper extremities. Consequently the spinal roots and the cervical sympathetic nerves, vagus, peroneal and ulnar nerves are most commonly effected (1). A schwannoma can develop infrequently in the gastrointestinal tract or retroperitoneal cavity as well as any other part of the body. However, a primary benign schwannoma of the liver is extremely rare (2). In this paper we report a case of a benign schwannoma of the liver parenchyma, which was at first misdiagnosed as degenerated hydatid cyst.

Case Report
A 56-year-old woman was referred to our hospital with epigastric pain and for further evaluation of a 9 cm-sized mass in the liver detected by abdominal ultrasonography at a private clinic. The patient initially presented to the clinic with epigastric discomfort for four weeks. No specific findings were noted in the medical or family history. On physical examination, none of the vital signs on admission were found to be abnormal. The patient appeared alert and the Eastern Cooperative Oncology Group (ECOG) performance status was 1. There were no pathologic lymph nodes palpable. The patient had anicteric sclerae and normal conjunctivae. Chest, back and extremities showed no particular pathology on physical examination. Her abdomen showed no distension, rebound tenderness or organomegaly. On inspection no pathologic dermatologic signs were seen. The laboratory findings on admission were as follows; hemoglobin 11 g/dL, white blood count 5.7 10³/μL, platelet 286 10³/μL, glucose 103 mg/dL, blood urea nitrogen 29.9 mg/dL, creatinine 0.54 mg/dL, aspartate aminotransferase 17.6 IU/L, alanine aminotransferase 12 IU/L, total bilirubin 0.71 mg/dL. A computed tomography (CT) of the abdomen revealed a -10 cm cystic lesion in the left lobe of the liver including multiple calcified foci within its highly dense fluid. It was encapsulated with a highly fibrotic capsule and exophytic invasion of the mass to omentum minus was also reported. There were no enlarged lymph nodes found near the mass and no other abnormal findings on the abdominal CT scan (Fig. 1A, B). In the ultrasound exam of the patient, dimensions and parenchymal echo of the liver was normal. A 10×10 cm necrotic mass was seen in the posteroinferior part of the left liver lobe.

With the prediagnosis of degenerated hydatid cyst, the patient underwent a subcostal incision; the consequential intraoperative exploration finding was a mass (approximately...
Figure 1A. In abdominal computed tomography an 6×7 cm cystic well capsulated mass including calcified foci in sagittal sections.

Figure 1B. In abdominal computed tomography an 6×7 cm cystic well capsulated mass including calcified foci in transverse sections.

15×10×10 cm) with exophytic nodules and haemorrhagic necrosis. On pathologic macroscopic examination there were four hemorrhagic, brown coloured, irregular surfaced masses with elastic density sized 2×1×0.3 cm, 2×1.5×1 cm, 3×1×2 cm and 6×3×2 cm from the smallest to largest. In total the neoplastic tissue weighed 278 grams and the cross-sectional surfaces of the material was solid and grayish coloured. Adjacent liver parenchyma was macroscopically normal. The operation proceeded to trisegmentectomy which included segments 2, 3 and 4. Microscopic examination of the surgical specimen revealed two different growth patterns in the neoplastic tissue: First is a Antoni A region which is highly cellular and contains a relatively scarce matrix and highly cellular (Fig. 2A). In cellular areas nuclei were palisating to form verocay bodies in a random order (Fig. 2B). Second of the two above-mentioned growth patterns is Antoni B region with loose myxoid stroma and with poor cellularity (Fig. 2C). Tumoral tissue was composed of cells with spindle nuclei in spurred cytoplasm. No mytotic figures were seen. In immunohistochemical staining NSE and S-100 reactions were positive (Fig. 2D), and SMA (smooth muscle actin) reaction was negative. In some previous reports CD-117 staining was also performed while schwannomas are thought to be a subgroup of gastrointestinal stromal tumors (3). However, in the present case CD-117 was not performed as this receptor is not valuable in a mesenchymal liver tumor. The patient is healthy and free from recurrence at her last follow up, 22 months after the surgery.

Discussion

Schwannoma is a benign mesenchymal tumor that originates from Schwann cells in the peripheral nerves (5). Schwannoma (neurilemoma) commonly arises along the peripheral nerve as well as in the cranial nerves and dorsal spinal nerve roots; however its very rare in the liver (6). The tumor is associated with neurofibromatosis in about 50% of cases. Malignant transformation of these tumors is very rare (5). However as Enzinger and Weiss claim in their textbook on pathology of soft tissue sarcomas benign schwannomas are usually sporadic lesions. They based their argument on a population-based study reporting that 90% were sporadic, 3% occurred in patients with neurofibromatosis 2 (NF2), 2% in those with schwannomatosis and 5% in association multiple meningiomas in patients with or without NF2. Rarely they occur as part of NF1 (1). They grow very slowly and are well encapsulated in most cases. Schwannomas are usually smaller than 5 cm at diagnosis (2). Larger schwannomas have a tendency to undergo secondary degeneration such as pseudocystic regression, hemorrhage and calcification which generally leads to a misdiagnosis of degenerated hydatid cyst on CT as in the present case. The incidence of a schwannoma in the gastrointestinal (GI) tract is very low; gastrointestinal schwannomas usually occur in the stomach (60%-70%) or colon (20%-30%) of older adults, and they are rare in the small intestine and in other GI locations. Nearly all gastric and most intestinal schwannomas are relatively small (<5 cm). Intramural spindle cell tumors are composed of bundles of spindle cells with focal atypia and often form a microtrabecular pattern intermingled with fibrovascular septa. Epithelioid variants occasionally present as colonic mucosal polyps (7). There are several reports of benign schwannomas in the stomach, duodenum, rectum, and retroperitoneum. The hepatobiliary nerves originate from the hepatic plexus in the hilum. They are composed of sympathetic and parasympathetic (vagal) fibers (6). Consequently benign schwannoma of the liver can originate from a variety of hepatic sympathetic and parasympathetic nerves distributed among the intralobular connective tissues and hepatic arteries. In the liver few nerves run to the liver cells and their terminals are uncertain (8). The schwannoma in the present case was surrounded by normal hepatic parenchyma. Thus, the tumor was diagnosed as a benign primary schwannoma of the liver. In 1993, Hytiroglou et al reported the first primary benign hepatic schwannoma diagnosed after surgical resection (9). We reviewed the clinical characteristics, location, size and secondary degeneration of primary benign schwannomas and the findings are shown in Table 1 (3, 4, 6, 9, 11, 13-17). They were found in ten women and two men. The age of presentation ranged between 38
and 70 years. Most patients were admitted with the complaint of pain or discomfort in the upper abdomen or epigastrium. The location of liver tumors was the right lobe in five cases and the left lobe in six cases (the location of the tumor in the first case reported in 1977 is not mentioned). The maximal diameter of the tumors reported varied between 4 and 30 cm. Tumors had secondary degeneration in most of the cases except the smaller ones were approximately 4 or 5 cm in diameter. Radiological findings showed degenerative changes with cyst-like characteristics, calcification and hemorrhage (10). To date, the CT appearance of schwannoma has not been precisely characterized. Consequently the literature includes a few attempts for the radiologic description of such lesions and differentiation from their malignant counterparts (10). However, the tumor size was much smaller than average. Generally, a plain CT depicts a schwannoma as a nonhomogenous area; an enhanced CT shows the margins clearly and the inside appears as an irregular pattern (6). Akin et al performed PET - CT for two of their breast cancer patients with liver lesions suspicious of metastasis in abdominal ultrasound. Although in PET - CT the lesion in the liver revealed pathologic fluoro-2-deoxyglucose (FDG) uptake, they confronted an incidental liver schwannoma in both of the patients in the postoperative setting (11). A definitive diagnosis of a hepatic schwannoma by radiological methods is difficult because it is such a rare finding. Therefore, pathological examination is essential for the diagnosis. Microscopic examination shows spindle cells like other stromal tumors. The histological diagnosis of a benign schwannoma is usually a simple procedure with standard Hematoxylin and Eosin staining. The distinction between a schwannoma and other spindle cell tumors or neurofibromas is based on the presence of a true capsule and Antoni A and Antoni B findings which are the two structural components of schwannoma (2). Antoni A area is a highly ordered cellular region; on the contrary the Antoni B area is a loose myxoid region. Immunohistochemical staining is diffusely and strongly positive for S-100 protein in a schwannoma consistent with the finding of a nerve sheath tumor. The tumor is also positive for the glial fibrillary acidic protein and CD57 (Leu 57) (12). Even though a
few gastrointestinal stromal tumors are positive for S-100, they are also positive for either CD34 or CD117. However, a schwannoma is negative for both CD34 and CD117. A leiomyoma would be negative for S-100 and positive for desmin or smooth muscle actin (7). The treatment for a benign primary schwannoma is a simple excision. The complete excision of the tumor is curative and most cases do not relapse; additional treatments are not necessary. The overall prognosis is very good (1).

In conclusion, we report a case of primary schwannoma of the liver parenchyma. The diagnosis was established after surgical resection of the tumor that was identified to have spindle cells with Antoni A and Antoni B areas and was strongly positive for the S-100 protein.

## References