

Schwannoma in the hepatoduodenal ligament with portal vein invasion

A case report

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Abstract

Rationale: Schwannomas are mesenchymal tumors with low malignant potential that originate from Schwann cells. They can occur in most parts of the body, such as the head, neck, and extremities. Schwannoma in the hepatoduodenal ligament is extremely rare, and only four cases have been reported in the literature.

Patient concerns: Herein, we describe a 58-year-old female who presented with right epigastric pain for 10 days. Preoperative computed tomographic (CT) revealed a 4.5 cm × 3.8 cm tumor in the hepatic hilar area.

Diagnoses: Schwannoma in the hepatoduodenal ligament with portal vein invasion.

Interventions: Intraoperative findings revealed that the tumor was identified in the hepatoduodenal ligament, and the left branch of the portal vein was compressed. Complete tumor resection with reparation of the portal vein was performed for the patient. Postoperative pathological examination confirmed the final diagnosis of benign schwannoma, characterized by abundant spindle-shaped cells and positive reactivity for S-100 protein.

Outcomes: The patient had a good prognosis and had no recurrence after 37 months of follow-up.

Lessons: Our case of schwannoma in the hepatoduodenal ligament is unique owing to the portal vein invasion, aimed at helping recognize the difficulty of preoperative diagnosis.

Abbreviations: CT = computed tomography, SMA = smooth muscle actin.

Keywords: hepatoduodenal ligament, portal vein invasion, schwannoma

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J-KW and QW have contributed equally to the study and were the co-first authors.

Ethical review and written informed consent: The ethical approval from the Ethics Committee was not necessary to report this case because the case was reviewed retrospectively. Written informed consent was obtained from the patient for publication of this case report and all accompanying images.

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All data generated or analyzed during this study are included in this published article [and its supplementary information files].

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1. Introduction

Schwannomas originate from Schwann cells, which cover peripheral nerves.^[1] It seems that these tumors can occur at all ages with no obvious sex difference. Most schwannomas are benign, and they comprise approximately 5% of benign soft-tissue tumors.^[2,3] They can occur in any part of the body, although the most common parts are the head, neck, and extremities.^[4] Schwannomas usually have a single tumor origin, while 10% originate from multiple locations.^[5] They rarely occur in the abdominal cavity; in particular, only 4 cases of schwannomas in the hepatoduodenal ligament have been reported in the literature.^[6–9] The preoperative diagnosis of fascioliasis is difficult because of nonspecific clinical manifestations. Surgical excision with negative margins is the most effective treatment. Following complete tumor excision, patients with benign schwannomas generally have a relatively good prognosis.^[10]

Herein, we describe a case of schwannoma in the hepatoduodenal ligament with portal vein invasion in a 58-year-old female patient who was diagnosed and treated in our department. This case is of interest because of the difficulty surrounding the preoperative diagnosis and the unique extent of tumor invasion.

2. Case report

A 58-year-old woman was admitted to our department on June 7, 2016, complaining of right epigastric pain for 10 days. She had a medical history of chronic gastritis for over 20 years but no

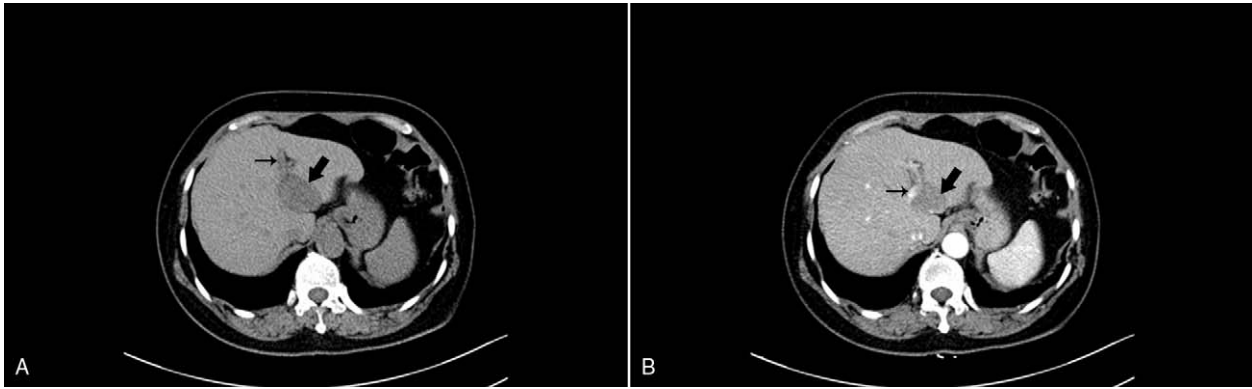


Figure 1. Abdominal CT showed a well-defined hypodense lesion (thick arrow) in the hepatic hilar, compressing the left branch of the portal vein (fine arrow); (A) plain scan phase; (B) arterial phase. CT=computed tomography.

history of abdominal surgery. Her family history had no significant disease. Mild tenderness of the upper abdomen was detected during physical examination. Routine laboratory tests, including routine blood tests, biochemical analyses, and tumor marker levels, were within the reference ranges. Plain computed tomography (CT) revealed a 4.5 cm × 3.8 cm, well-defined hypodense lesion in the hepatic hilar area (Fig. 1A); contrast-enhanced CT revealed mild enhancement, and the left branch of the portal vein was compressed by the tumor (Fig. 1B). According to the CT examinations, an abdominal mass was primarily considered.

Although a definitive preoperative diagnosis was lacking, exploratory laparotomy was performed for the patient because of the continuous epigastric pain. Intraoperative findings revealed that a mass surrounded by a fibrous capsule was identified in the hepatoduodenal ligament, and the left branch of the portal vein was infiltrated. The mass was completely resected with reparation of the left branch of the portal vein.

Macroscopically, the tumor was a capsulated yellowish-white mass, 4.8 cm × 4.2 cm × 3.0 cm in size. Histopathological examination revealed that the tumor was mainly composed of spindle-

shaped cells, and no atypical cells were observed (Fig. 2A). Immunohistochemically, the tumor cells were strongly positive for S-100 protein (Fig. 2B) but negative for desmin and CD117. Based on the above findings, the final diagnosis was primary schwannoma in the hepatoduodenal ligament. The postoperative recovery of the patient was good. She was followed for 37 months in our outpatient department and has no recurrence as of the time of this report.

3. Discussion

Originating from the Schwann cells of nerve sheaths, schwannomas are mesenchymal neoplasms with low malignant potential.^[1] The secondary degenerative changes due to schwannomas may sometimes present as cyst formation, calcification, hemorrhage, and hyalinization.^[11] More than 90% of schwannomas are benign, accounting for approximately 5% of benign soft-tissue neoplasms.^[2,3] The disease can occur in most parts of the body; however, the most common locations are the head, neck, spinal cord, and extremities.^[4] Schwannomas have also been found in abdominal viscera, such as the retroperitoneum,^[12] stomach,^[13]

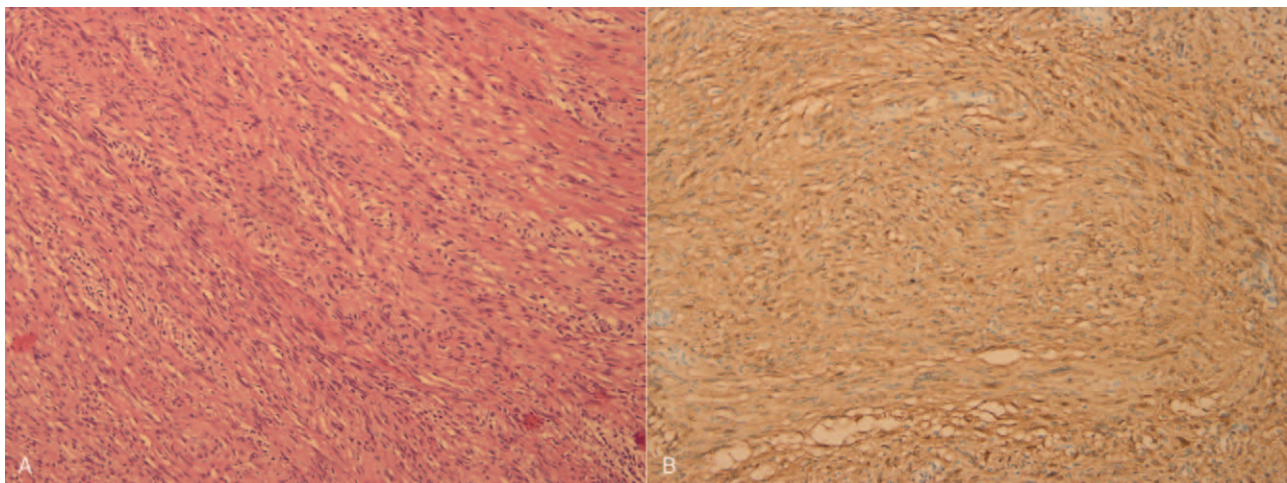


Figure 2. Pathological findings. (A) Microscopic examination showed that the tumor was mainly composed of spindle-shaped cells (HE ×200). (B) Immunohistochemical staining revealed the tumor cells were positive for S-100 protein (×200). HE=hematoxylin and eosin.

gallbladder,^[14] pancreas,^[15] bowel mesentery,^[16] colon,^[17] and common bile duct,^[5] although the incidence is extremely low. In the abdominal cavity, the retroperitoneum is the most involved site.^[18] Primary schwannoma in the hepatoduodenal ligament is extremely rare. To our knowledge, only 4 such cases have been reported in the English literature worldwide.^[9] The first case was reported by Nagafuchi et al^[6] in 1993. Herein, we report the fifth case of schwannoma in the hepatoduodenal ligament, and our case is unique because of the portal vein invasion by the tumor.

A literature review of reported cases of schwannoma in the hepatoduodenal ligament (including the present case) was performed. The male-to-female ratio was 3:2, and the age was between 29 and 62 years. Two patients presented with pain in the right abdomen, and the remaining patients were asymptomatic, with schwannomas discovered by routine physical examination. All 5 patients had solitary tumors. The diameter of the tumors ranged from 4.5 to 9.0 cm. The cause and mechanism of schwannoma are unclear due to its low incidence.

It is difficult to determine the location and obtain a precise diagnosis prior to surgery, although multiple imaging modalities. Ultrasound, CT, and magnetic resonance imaging may provide a reference for probable diagnosis. Color Doppler ultrasound usually shows well-defined hypodense lesions with no echoic enhancement.^[9] CT usually reveals a round or oval homogeneously attenuating, well-defined lesion with frequent signs of degeneration, such as cysts and calcifications.^[19] Endoscopic ultrasound guided fine needle biopsy may contribute to a definitive preoperative diagnosis^[20]; however, this invasive operation is not routinely used in clinical practice. The tumor in our case was found in the hepatoduodenal ligament during the operation, which did not match the preoperative CT findings. The definitive diagnosis of schwannoma can only be based on pathological examination, and schwannomas often present with spindle-shaped cells and without atypia.^[8] Immunohistochemically, schwannomas are usually positive for S-100 protein and negative for desmin, smooth muscle actin (SMA), CD34, and CD117.^[21] In our case, immunohistochemistry showed strong positivity for S-100 protein, which supported the benign nature of the tumor.

Surgical resection is currently the only method of determining the tumor location, and it can be a curative treatment for schwannomas.^[9] Of all the patients with schwannomas in the hepatoduodenal ligament, 4 underwent laparotomy, while 1 underwent laparoscopic surgery. In the present case, exploratory laparotomy revealed that the mass was located in the hepatoduodenal ligament, and the left branch of the portal vein was infiltrated. We carefully separated the portal vein with reparation around the tumor. Histopathological and immunohistochemical examinations of resected specimens showed typical features of schwannoma.

4. Conclusions

Schwannoma in the hepatoduodenal ligament with portal vein invasion has not been reported, and we present the first case with complete tumor excision. Following complete tumor excision,

patients with benign schwannomas generally have good prognosis.

Author contributions

Conceptualization: Fu-Yu Li.

Data curation: Jun-Ke Wang, Qian Wu.

Writing – original draft: Jun-Ke Wang, Qian Wu.

Writing – review & editing: Zhen-Ru Wu, Fei Liu, Cong-Dun Ran, Chang-Hao Yin.

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