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Primary hepatic schwannoma: A case report

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ABSTRACT

INTRODUCTION: A hepatic schwannoma is extremely rare and difficult to diagnose preoperatively.
PRESENTATION OF CASE: We report the case of a 47-year-old male patient who was referred to our hospital for the close investigation of a hepatic tumor which had not been detected two years earlier. An enhanced computed tomography revealed a well-circumscribed and encapsulated tumor with a size of 50 mm which was adjacent to the inferior vena cava (IVC) and the right hepatic vein. The tumor was heterogeneously enhanced until the equilibrium phase. A magnetic resonance image showed a hypointense area on a T1-weighted image and a hyperintense area on a T2-weighted image. These findings differed from those of common malignant hepatic tumors, such as hepatocellular carcinoma and colorectal liver metastases. The tumor was most likely a mucus-producing tumor or a liquefactive degenerated adenocarcinoma. Although we could not confirm an exact diagnosis of the tumor, we performed a surgical resection in view of the possibility of malignancy. The patient underwent a limited liver resection with resection of the IVC. Histologically, the tumor was diagnosed as a benign schwannoma comprised of Antoni A and B areas. The nuclear palisading formation of the tumor showed Verocay bodies.
DISCUSSION: 15 cases of hepatic schwannoma are reviewed to clarify the typical radiological features. The radiological findings of the present case were consistent with those of the hepatic schwannoma when considering retrospectively.
CONCLUSION: A precise preoperative diagnosis of hepatic schwannoma is difficult, and liver resection is recommended when a hepatic schwannoma is suspected.

1. Introduction

Schwannoma is a benign nerve sheath tumor originating from Schwann cells. These tumors can emerge at any part of the peripheral nerves. Common sites include the head and neck region, the flexor surfaces of the upper and lower extremities, and the trunk [1]. Vestibular schwannomas account for about 8% of primary brain tumors. These lesions are sometimes incidentally diagnosed because of the widespread use of magnetic resonance image (MRI) and computed tomography (CT). Their radiological features are well known: an encapsulated tumor with a clear margin, which is heterogeneously enhanced, and a hypointense and hyperintense appearance on T1-weighted and T2-weighted MRI findings, respectively. However, the radiological features of primary hepatic schwannomas are unknown because of their extreme rarity. We treated a 47-year-old male patient with a hepatic schwannoma that could not be diagnosed preoperatively. Thus, we attempted to gain insight into the possibility of a precise preoperative diagnosis of primary hepatic schwannoma based mainly on radiological modalities by reviewing past case reports.

Abbreviations: MRI, magnetic resonance imaging; CT, computed tomography; US, ultrasonography; IVC, inferior vena cava; RHV, right hepatic vein; EOB-MRI, gadolinium-ethoxybenzyl-diethylenetriamine penta-acetic acid-enhanced magnetic resonance imaging; MPNST, malignant peripheral nerve sheath tumor; NF-1, neurofibromatosis type 1.

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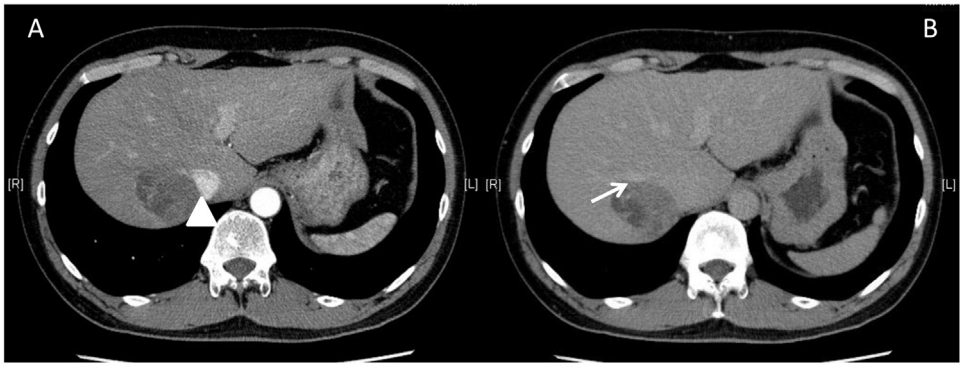

Fig. 1. Dynamic contrast-enhanced CT image showing an approximately 5-cm, well-circumscribed tumor adjacent to the IVC (arrowhead in A) and the RHV (arrow in B) in segment 7. A) Heterogeneous enhancement was visible during the arterial to portal phase of the CT examination. B) The heterogeneous enhancement persisted until the equilibrium phase.

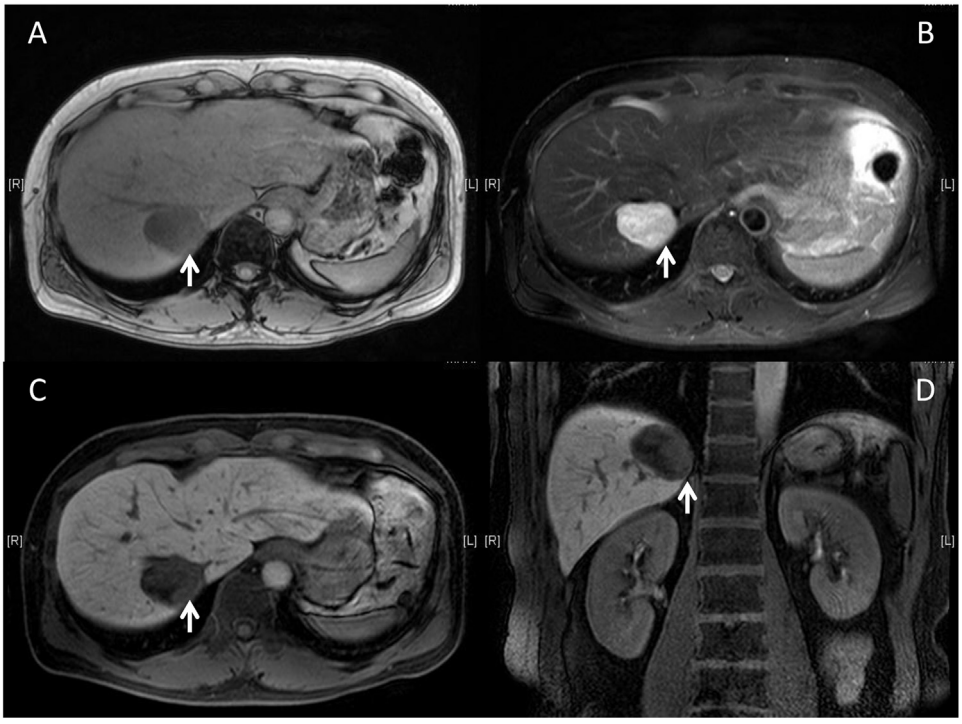

Fig. 2. A, B: Magnetic resonance images. A) A T1-weighted image shows a hypointense tumor (arrow). B) A T2-weighted image shows a hyperintense tumor (arrow). C, D: Gadolinium-ethoxybenzyl-diethylenetriamine penta-acetic acid-enhanced (EOB) MRI shows a defect (arrow) during the hepatobiliary phase. **C) Axial image. D) Coronal image.**

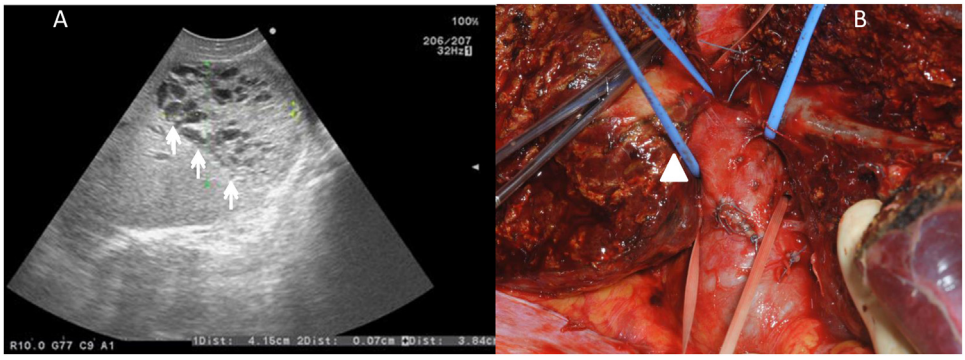

Fig. 3. A) US shows a multicystic tumor with numerous septa (arrow). B) Even after the division of the liver parenchyma, the specimen was still attached to the IVC (arrow head).

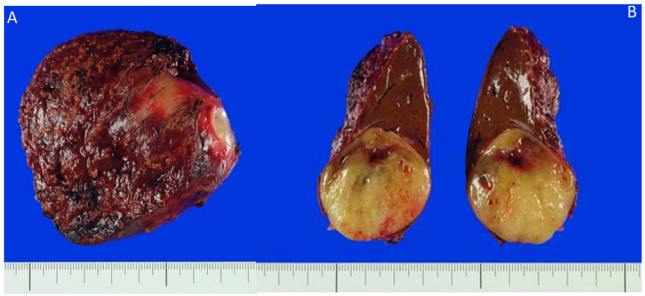

Fig. 4. Macroscopic image of the tumor A) A white tumor was resected together with a portion of the IVC. B) The cut surface showed multiple cysts.

2. Presentation of case

A 47-year-old male patient was referred to our hospital because of a hepatic tumor (50 mm) that had been discovered during an ultrasonography (US) examination as part of a regular medical check-up. The tumor had not been detected during a check-up performed 2 years earlier. The patient had no particular past history of illness or congenital disease. Laboratory data revealed that serological markers for hepatitis B virus and C virus were negative. A complete blood count and blood biochemistry tests showed normal values. No elevations in the serum carcinoembryonic antigen, carbohydrate antigen 19–9, alpha-fetoprotein, or des-γ-carboxy prothrombin levels were noted. The patient underwent a dynamic contrast-enhanced CT examination that revealed a well-circumscribed tumor with a size of 50 mm that exhibited internal heterogeneity and prolonged enhancement. The lesion was adjacent to the inferior vena cava (IVC) and the right hepatic vein (RHV) (Fig. 1A and B). Gadolinium-ethoxybenzyl-diethylenetriamine penta-acetic acid-enhanced MRI (EOB-MRI) scans revealed a well-circumscribed tumor that was hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 2A and B). After the injection of EOB as a contrast agent, a defect during the hepatobiliary phase was observed (Fig. 2C and D). A gastrointestinal fiberscopy and colon fiberscopy examination showed no evidence of a tumor. These findings differed from the typical findings for hepatocellular carcinoma and colorectal liver metastases. A biopsy was not performed because of the fear of dissemination. The lesion was thought to contain a heterogeneous liquid compartment and to have possibly undergone rapid growth over the previous two years. Thus, the lesion was considered to most likely be a mucus-producing tumor or a liquefactive degenerated adenocarcinoma. Even though no convincing data was obtained to indicate whether the tumor was benign or malignant, we decided to perform surgery since the tumor had not been detected two years earlier. Consequently, the patient underwent a limited liver resection combined with an IVC resection. The operation time was 330 min, and the estimated intraoperative blood loss was 610 g. A whitish tumor was found beneath the diaphragm, behind the RHV, and to the right of the IVC. Intraoperative US showed a multicystic tumor with numerous septa (Fig. 3A). Even after the complete division of the liver parenchyma, the detachment of the tumor from the IVC was still difficult (Fig. 3B). Since we suspected that the tumor might have originated from the IVC itself or from tissue surrounding the IVC, we removed the tumor in conjunction with a partial resection of the IVC.

Macroscopically, a clearly demarcated white-gray tumor with focal cystic changes was observed (Fig. 4A and B). Histologically, the tumor consisted of spindle-shaped cells and included the presence of Antoni A and Antoni B areas. The Antoni A area was composed of spindle-shaped cells arranged in interlacing fascicles. The Antoni B area was composed of a myxoid component in which the tumor cellular density was relatively low (Fig. 5A). The nuclear palisading formation of the tumor exhibited Verocay bodies and there were few nuclear atypias and mitotic counts (Fig. 5B). Based on these findings, the tumor was diagnosed as a typical benign schwannoma

The patient's postoperative course was uneventful. He was discharged from our hospital on the 10th postoperative day. The patient is presently well and has shown no signs of recurrence as of 6 months after the operation.

3. Discussion

Schwannoma is a benign mesenchymal tumor emerging from a nerve sheath. Its malignant counterpart, so-called malignant peripheral nerve sheath tumor (MPNST), tends to arise following irradiation [2] or in association with neurofibromatosis type 1 (NF-1)[3,4]. Schwannomas are frequently detected in patients between the second and fifth decades of life and are found most commonly in the head and neck region, the flexor surfaces of the upper and lower extremities, or the trunk [1]. Although a schwannoma may arise from any organ that contains peripheral nerves, primary hepatic schwannomas are extremely rare. According to English medical literature, only 19 cases of hepatic schwannoma, including the presently reported case, have been documented (Table 1) [3–18]. These 19 cases consisted of 9 men (47.1%) and 10 women (52.9%), with a median age of 56 years (range, 21–83 years). Two patients had a history of NF-1. In 9 patients (47.1%), MPNSTs were diagnosed based on pathological findings: nuclear atypia, cell density, and mitotic count. Although a few cases that have not been associated with either irradiation or NF-1 have been reported, 8 of the 10 patients who were diagnosed as having hepatic MPNST had no history of congenital disease or irradiation [5,9,10,13,16–19].

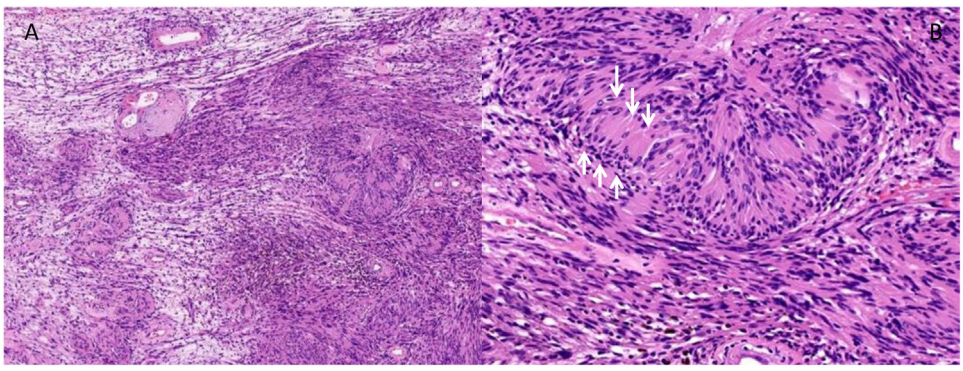

Fig. 5. Microscopic image of the tumor A) The tumor consisted of Antoni A and B areas. The Antoni A area was rich in spindle cells. The Antoni B area contained a myxoid component with a low density of tumor cells. B) Verocay bodies with nuclear palisading (arrows) are visible.

Table 1
Previous reports about hepatic schwannoma and MPNST.

Author	Year	Age/sex	Diameter	NF	MPNST/ Schwannoma	Radiological features
Young et al. [3]	1975	M/23	5 cm	+	MPNST	Autopsy
Tuder et al. [18]	1984	M/74	4.5 cm	–	MPNST	Autopsy
Lederman et al. [4]	1987	M/21	4 cm	+	MPNST	Autopsy
Hytiroglou P et al. [8]	1993	M/67	13 cm	–	Schwannoma	Plain CT: well circumscribed
Heffron TG et al. [7]	1993	F/38	5 cm	–	Schwannoma	Plain CT: low density area
Morikawa et al. [13]	1995	M/63	20 cm	–	MPNST	Autopsy
Sheikh MY et al. [16]	1996	F/35	–	–	MPNST	Enhanced CT: well defined, heterogeneously enhanced
Fiel MI et al. [5]	1996	M/49	4.5 cm	–	MPNST	Not mentioned in the article
Wada et al. [19]	1998	F/69	15 cm	–	Schwannoma	Plain CT: low density area
Wada et al. [19]	1998	F/64	4 cm	–	Schwannoma	Plain CT: low density area
Flemming et al. [6]	1998	F/57	Huge	–	Schwannoma	Plain CT: cystic lesion
Momtahen AJ et al. [12]	2008	F/52	4.4 cm	–	Schwannoma	MRI T1 hypointense, MRI T2 heterogenously hyperintense, Enhanced MRI: heterogenousely enhanced
Kóbori L et al. [10]	2008	F/22	26 cm	–	MPNST	Enhanced CT: well cicumscribed, heterogenously
Iddings DM [9]	2008	M/83	7 cm	–	MPNST	Enhanced CT: not enhanced
Lee WH et al. [11]	2008	F/36	5 cm	–	Schwannoma	Enhanced CT: well circumscribed, low-attenuating mass,containing central septation
Subramaniam et al. [17]	2012	M/71	21 cm	–	MPNST	MR, CT: complex solid and cystic right lobe liver lesion
Ozkan EE et al. [15]	2010	F/56	15 cm	–	Schwannoma	Enhanced CT: well capsulated mass including calcified foci
Yu Ota [14]	2012	F/72	6.4 cm	–	Schwannoma	Enhanced CT: heterogenously enhanced, MRI:T1 hypointense T2 heterogenously hyperintense
Our case	2016	M/47	5.0 cm	–	Schwannoma	Enhanced CT: heterogenously enhanced, MRI:T1 hypointense T2 heterogenously hyperintense

NF: Neurofibromatosis Diameter: longest diameter.

A precise preoperative diagnosis of primary hepatic schwannoma was not possible in any of the cases listed in Table 1. The radiological features of the 15 resected cases are summarized in Table 1. The most common CT findings were a low density during a plain phase and a well-demarcated lesion with heterogeneous enhancement. As for MRI, the most common findings were hypointensity on T1-weighted images and hyperintensity on T2-weighted images. These radiological findings are consistent with those of typical acoustic schwannomas or schwannomas originating in more common locations. Ota et al. [14] reported that contrast enhanced US was useful in diagnosing hepatic schwannoma and it showed minute arterial flow into the tumor along the nodal septum during the vascular phase and the delayed enhancement of solid areas during the postvascular phase. Although the radiological characteristics of the present tumor were similar to those of schwannoma when considered retrospectively, a precise diagnosis of primary hepatic schwannoma is still considered to be difficult because of the extreme rarity of this tumor and its radiological similarity to a mucus-producing tumor or adenocarcinoma. If a schwannoma is suspected, a radiological consultation is highly recommended.

Meanwhile, regarding the differentiation of hepatic schwannomas, there seems to be no difference in the radiological features of malignant and benign tumors. Thus, distinguishing these lesions based on preoperative CT and MRI findings without a biopsy would likely be difficult.

If the tumor is diagnosed as a schwannoma preoperatively, surgical resection is still required because of the difficulty in discriminating malignant tumors from benign tumors and the possibility of malignant transformation. Actually, malignant cases account for about half of all the reported hepatic schwannomas.

4. Conclusions

We treated a patient with an extremely rare tumor, a hepatic schwannoma. Although a precise diagnosis of this tumor is difficult, surgical resection is required in cases with suspected schwannomas.

Conflict of interest

The authors declare that they have no competing interests.

Informed consent

Written informed consent for the publication of this case report and all accompanying images was obtained from the patient and the patient's family. Copies of the written consents are available for review by the Editor-in-Chief of this journal.

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None.

Author contribution

MY, KH, JA, RM, NA, JK, YS, and NK participated in the clinical treatments. TW, NO and OK carried out the radiological diagnosis. AH, JS and MF carried out the pathological diagnosis.

Guarantor

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