Ancient Schwannoma of the Gallbladder

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ABSTRACT

Schwannomas are benign tumors which arise in the Schwann cells of the peripheral nerves. They develop anywhere in the human body but are less frequent found in the digestive tract. A 67-year-old female patient was referred to our tertiary HPB department with an asymptomatic mass of the gallbladder. Contrast-enhanced computed tomography revealed a rather homogeneous, well-circumscribed, high-density ovoid mural mass. An open cholecystectomy was performed. A final diagnosis of schwannoma with degenerative atypia, so-called “ancient schwannoma”, was made. Clinical recovery and follow-up was uneventful, with no signs of recurrence.

INTRODUCTION

Schwannomas are benign tumors which arise in the Schwann cells of the peripheral nerves. These neurogenic tumors are benign and often develop in young to middle-aged patients.1,2 Schwannomas can develop everywhere in the human body, preferentially in the head, neck, and extremities but are less frequently found in the digestive tract.3–5 In the digestive tract, schwannomas are found in the stomach, colon, and rectum. This case report is the first description of the occurrence of an ancient schwannoma of the gallbladder.

CASE REPORT

A 67-year-old woman was admitted to the hospital because a mass was incidentally detected in her gallbladder during a routine health examination due to vague abdominal complaints. She had a history of renal insufficiency and atrial fibrillation but no previous abdominal surgery. There was no evidence of jaundice or abdominal tenderness.

The blood laboratory showed a leukocyte count of 10.9 × 10⁹/L, estimated glomerular filtration rate of 32 mL/min/1.73 m², hemoglobin of 8.6 mmol/L, total bilirubin of 5 μmol/L, serum alanine aminotransferase of 20.0 U/L, serum alkaline phosphatase of 17.0 U/L, and serum gamma-glutamyltranspeptidase of 59.1 U/L. The laboratory studies revealed an almost-normal liver function. Carcinoembryonic antigen was within the normal ranges. Ca 19-9 was 173 U/L.

An abdominal ultrasound revealed a well-defined isoechoic polypoid mural mass located in the body of the gallbladder. No intraluminal protruding polypoid mass located in the body of the gallbladder. No intraluminal protruding polypoid lesion was visible on color Doppler ultrasound. The overlying mucosa of the gallbladder was intact. Cholecystolithiasis in the neck was also appreciated. Consequently, an abdominal computed tomography scan with intravenous contrast was performed using a 64-slice scanner and revealed a rather homogeneous high-density ovoid mural mass with a clear boundary that measured 30 × 35 × 18 mm (Figure 1). No signs of central necrosis and no clear internal calcifications were seen other than the above mentioned; the gallbladder had a normal appearance. Differential diagnosis included adenomyomatosis of the gallbladder, gallbladder polyp, gastrointestinal stromal tumor, and gallbladder cancer.

An open cholecystectomy with a gallbladder bed resection and intraoperative frozen section was performed. The analysis of the frozen section revealed a probably benign polypoid lesion with a highly atypical stromal component and normal mucosa. The gross specimen showed an intraluminal protruding polypoid mass of 4.2 cm (Figure 2). The cut surface showed no signs of invasion.
Hematoxylin and eosin-stained slides showed the polypoid lesion consisted of highly atypical stroma, covered by normal mucosa. The stromal cells showed spindled wavy nuclei with tapered ends. There was a remarkable variation in nuclear size and shape, but mitoses were not seen. The cytoplasm was fibrillar, and there was nuclear palisading. Focally, the lesion consisted of looser more haphazardly arranged cells (Figure 3). Immunohistochemically, the lesion was S100 positive (Figure 4). Cytokeratin was negative, as was desmin, smooth muscle actin, and CD34. Ki67 showed staining in less than 1% of nuclei.

A final diagnosis of schwannoma with degenerative atypia, so-called the “ancient schwannoma”, was made. The word “ancient” can be added to the diagnostic schwannoma to indicate that the histological findings are degenerative in nature and are not worrisome. Degenerative changes in the schwannomas include hyalinization, calcification, hemorrhage, and severe nuclear atypia. These can be seen focally in “normal” schwannomas, but it is believed these changes occur predominantly in the long-standing schwannomas, hence the word ancient. It was first suggested in 1951 by Ackerman and Taylor when reviewing neurogenic tumors of the thorax.6

In the 2 months since the operation, the patient recovered well and is without signs of recurrence. No routine follow-up was scheduled.

DISCUSSION

To our knowledge, this is the sixth case in the literature of a gallbladder schwannoma and the first describes a case of an ancient schwannoma. Until now, only 5 cases of a “non-ancient” schwannoma have been reported. In these reported cases of gallbladder schwannomas, 2 cases revealed obstructive jaundice and vague pain and the other 3 cases were asymptomatic.7 Yamagiwa et al did not describe imaging of the tumor.8 Matsuoka et al reported about a traumatic neurinoma and not a schwannoma.9 Little is known on the other 3 previous (non-English) publications of Ren et al, Colovic et al, and Ohta et al.10–12 Various sizes were reported from 3 to 110 mm7. In our case, the ancient schwannoma was 42 mm in size.

Because of the high degree of atypia in ancient schwannomas, frozen sections can be very challenging and the evaluating pathologist should be aware of this. The polypoid shape of the lesion found in our patient is also a remarkable finding, given the fact that schwannomas are usually well-circumscribed, nodular masses. Because of this polypoid morphology, there was a high index of suspicion for dysplasia or malignancy preoperatively. Another differential diagnosis based on the histology alone is a sarcoma, but the fact that the lesion was positive only for S100 and showed no mitotic activity argues against a malignant nature. In addition, it is well-known bizarre nuclear atypia that is degenerative in nature can occur in schwannomas.

Normally a schwannoma is a benign tumor derived from the Schwann cells that encapsulate the nerve sheath. von Recklinghausen disease is associated with malignant schwannomas.
although these are extremely rare.11,13 Schwannoma can be
found in the extremities, head, neck, trunk, retroperitoneum, or
mediastinum. But they can also develop in the gallbladder be-
cause of the abundant anastomotic network of the sympathetic
and parasympathetic nerve fibers in the wall of the gallbladder and
bile duct.5,11

However, schwannomas arising in the gallbladder are ex-
tremely rare. Because of a lack of adequate knowledge of this
tumor and low incidence in clinical practice, correct pre-
operative diagnosis is hard to achieve, especially when such
a tumor is small. In our patient, the tumor was rather large but
the patient was asymptomatic; on the contrary, jaundice and
vague pain may have happened when tumor was large enough
to compress the surrounding organs. Schwannomas of the
digestive tract have an excellent prognosis after a surgical
resection, similar to conventional schwannomas, and there
is no evidence to date that these tumors have a malignant
potential.

DISCLOSURES

Author contributions: JK de Bakker and F. Daams analyzed and
interpreted the patient data. JK de Bakker performed radiologic
examinations. E. Witteveen performed the histological exami-
nation of the gallbladder and wrote the manuscript. All authors
read and approved the final manuscript.

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