

Biliary tract schwannoma: A rare cause of obstructive jaundice in a young patient

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did not recur in any of the resected cases.

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Abstract

Schwannoma is a tumor derived from Schwann cells which usually arises in the upper extremities, trunk, head and neck, retroperitoneum, mediastinum, pelvis, and peritoneum. However, it can arise in the gastrointestinal tract, including biliary tract. We present a 24-year-old male patient with obstructive jaundice, whose investigation with computed tomography abdomen showed focal wall thickening in the common hepatic duct, difficult to differentiate with hilar adenocarcinoma. He was diagnosed intraoperatively schwannoma of common bile duct and treated with local resection. The patient recovered well without signs of recurrence of the lesion after 12 mo. We also reviewed the common bile duct schwannoma related in the literature and evaluated the difficulty in pre and intraoperative differential diagnosis with adenocarcinoma hilar. Resection is the treatment of choice for such cases and the tumor

INTRODUCTION

Although it is considered to be a rare tumor, adenocarcinomas are the most common malignant neoplasms of extrahepatic bile ducts^[1,2]. However, other non-epithelial tumors can develop, whether they are malignant, such as lymphomas and neuroendocrine cancer^[3], or benign, such as adenomas, lipomas, fibromas and schwannomas^[4,5], which sometimes appear similar to hilar adenocarcinomas (cholangiocarcinomas).

The goal of this case study was to present a rare case of common bile duct schwannoma that simulated a hilar adenocarcinoma. It was diagnosed during surgery, which allowed only a local resection.

CASE REPORT

A 24-year-old male patient was clinically diagnosed with

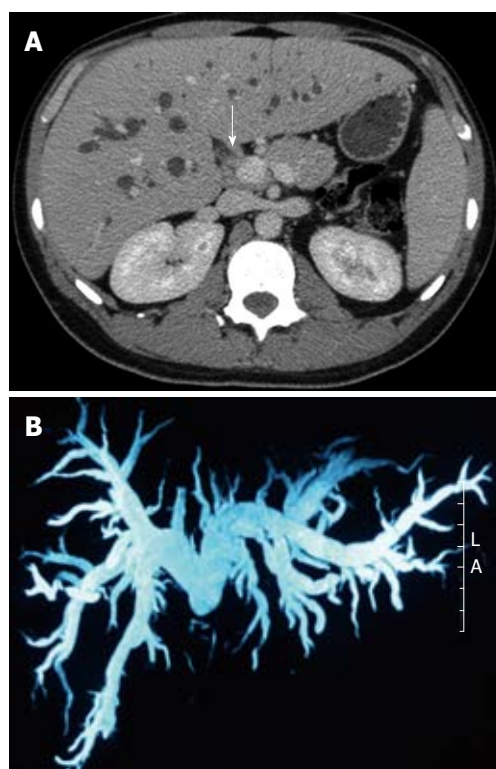


Figure 1 Large dilatation close to hepatic duct. A: Stenosis and thickening of the common hepatic duct (arrow) with upstream biliary dilation; B: Magnetic resonance cholangiography showing large dilatation close to the lesion in the common hepatic duct. A: Ahead; L: Left.

obstructive jaundice, had 2 mo of epigastric pain and vomiting, and lost 15% of his body weight. The patient had a history of smoking and social drinking. A physical examination confirmed that the patient had jaundice with a flaccid, painless abdomen and palpable liver 4 cm below the right costal margin. There were no other relevant findings.

The laboratory studies revealed: total bilirubin 23.8 mg/dL; direct bilirubin 22.9; serum alkaline phosphatase 298 IU/L; and serum gamma-glutamyl transpeptidase 1052 IU/L; serum aspartate aminotransferase 121 IU/L; serum alanine aminotransferase 249 IU/L; and carbohydrate antigen 19-9 was above normal range, 62.7 U/mL. Others laboratory tests were normal.

Imaging studies began with abdominal ultrasound sonography test, which revealed increased liver volume and dilation of the intra- and extrahepatic bile ducts up to the hepatic hilum.

A computed tomography scan of the abdomen showed focal wall thickening in the common hepatic duct 1.5 cm from the hepatic duct confluence. As a result, the diameter of the lumen was reduced, and there was upstream dilation (Figure 1A). The vascular structures and adjacent fat planes were preserved.

A magnetic resonance cholangiography showed biliary tract dilation with abrupt obstruction in the common hepatic duct (Figure 1B).

We opted for surgical treatment without biliary intervention for decompression or endobiliary biopsy, with a

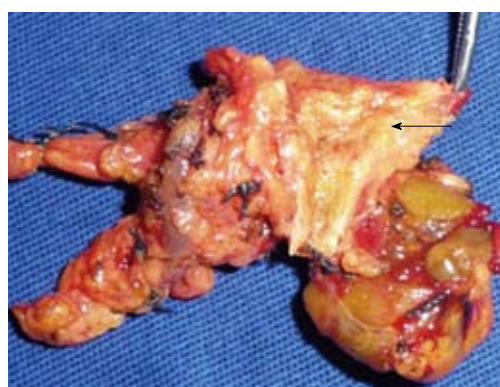


Figure 2 Hepatocolochochal tumor opened longitudinally with the affected area (arrow).

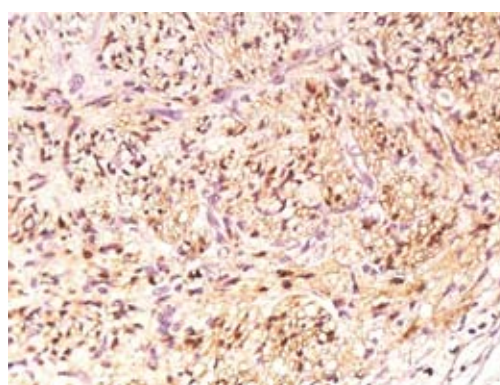


Figure 3 Immunohistochemical staining for the S-100 protein (40 ×).

preoperative diagnosis of hilar adenocarcinoma. In the cavity, we discovered that the liver was enlarged and appeared to be cholestatic. There was a 1-cm nodular lesion that was palpable in the common hepatic duct; additionally, the proximal bile duct was dilated. A cholecystectomy dissection was performed from the common bile duct above the pancreas to the nodular lesion in the common hepatic duct. During the dissection of the affected area, we observed that the tumor was well defined and regular, with no signs of having infiltrated adjacent tissues (Figure 2). We performed frozen sections, which showed a mesenchymal tumor with free margins in the hepatic duct. In view of these findings, we opted for local resection and Roux-en-Y hepaticojejunostomy.

The patient recovered and was discharged in good condition on the 7th postoperative day. A pathological examination showed a 0.8-cm lesion in the common hepatic duct, which was diagnosed as benign schwannoma with free margins in the extrahepatic bile duct. The diagnosis was made through microscopic examination and immunohistochemistry (Figure 3). The patient was re-examined 12 mo after surgery and was in excellent general health with no signs of recurrence.

DISCUSSION

Benign schwannoma, which is also known as neurile-

Table 1 Cases studies of biliary schwannoma in the literature

Observations/references	Age (yr)	Sex	Signals/ symptoms	Initial diagnosis	Location of tumor	Preoperative tissue acquisition
Oden <i>et al</i> ^[17]	40	F	Abdominal pain + obstructive jaundice	Choledocholithiasis	Common bile duct	No
Whisnant <i>et al</i> ^[18]	15	F	Abdominal pain + weight loss + obstructive jaundice		Distal portion of the common bil duct	No
Complicated by liver abscess, treated with drainage/Balart <i>et al</i> ^[19]	56	F	Abdominal pain + obstructive jaundice	Cholangiocarcinoma or extrinsic compression of the bile duct	Common hepatic duct	No
Jakobs <i>et al</i> ^[20]	37	M	Abdominal pain + obstructive jaundice	Intra-ductal benign tumor	Common hepatic duct	Yes
Honjo <i>et al</i> ^[13]	48	F	Obstructive jaundice	Benign non-epithelial tumor	Common bile duct	Yes (transpapillary brush cytology, non-diagnostic)
Otani <i>et al</i> ^[21]	59	F	Abdominal pain		Remnant bile duct (pancreatic portion)	No
Park <i>et al</i> ^[22]	53	F	Asymptomatic		Porta hepatis	No
Vyas <i>et al</i> ^[23]	29	F	Abdominal pain + obstructive jaundice		Common bile duct	Yes (non-diagnostic)
Kamani <i>et al</i> ^[24]	39	F	Jaundice + weight loss	Klatskin tumor	Proximal portion of the common hepatic duct	No
Fenoglio <i>et al</i> ^[16]	41	F	Obstructive jaundice + weight loss		Middle segment of the common bile duct	No
Jung <i>et al</i> ^[4]	64	F	Asymptomatic		Proximal portion of the common bile duct	No
Madhusudhan <i>et al</i> ^[5]	46	M	Obstructive jaundice	Variable polypoid cholangiocarcinoma	Intrahepatic bile duct	Yes
Kulkarni <i>et al</i> ^[7]	38	M	Abdominal pain + weight loss + jaundice		Common bile duct/ porta hepatis	No
Patient has von Recklinghausen's disease/De Sena <i>et al</i> ^[25]	58	F	Obstructive jaundice	Biliary schwannoma	Extrahepatic bile duct	No
Previous malignant melanoma/Panaït <i>et al</i> ^[26]	54	F	Gastroesophageal reflux symptoms	Recurrent metastatic melanoma	Porta hepatis	Yes (non-diagnostic)

F: Female; M: Male.

oma, is a tumor derived from Schwann cells, which form the inner portion of the peripheral nerve sheath^[6]. Theoretically, the tumor can affect any organ or nerve trunk, except the optic and olfactory nerves, which lack Schwann cells^[7]. The most common locations are the upper extremities, trunk, head and neck, retroperitoneum, mediastinum, pelvis, and peritoneum^[8].

Schwannomas in peripheral nerves can be associated with neurofibromatosis type 2, while many schwannomas can occasionally be associated with neurofibromatosis type 1^[5,9].

Schwannomas in the digestive tract are relatively rare. These tumors are most common in the stomach followed by the colon/rectum and esophagus^[10-12]. Additionally, schwannomas can develop in the biliary tract because there is an abundant network of sympathetic and parasympathetic nerve fibers along the wall of the gallbladder and bile ducts, but these cases are extremely rare^[4].

Diagnostic imaging tests do not allow preoperative diagnoses of biliary schwannoma because the findings are similar to those observed in most common lesions, particularly central cholangiocarcinoma. The most relevant finding in our case was that the preoperative examinations revealed a well-defined ductal injury with no signs of adjacent structural involvement or distant metastases^[5].

A schwannoma is usually a macroscopically encapsulated solid globular or ovoid tumor. Degenerative cystic changes are occasionally observed within the tumor. Microscopically, the tumor has two components: a hypercellular component with areas of spindle cells forming palisades (Antoni type A) and a myxoid component containing cuboidal cells with clear cytoplasm (Antoni type B)^[13].

We found 15 case studies of extrahepatic biliary schwannomas in the literature, which are shown in Table 1. Of these cases, the ages of the patients ranged from 15 to 64 years (with an average age of 44 years), and the patients were predominantly female (12/15). The most common symptom was jaundice (11/15 patients) followed by abdominal pain (7/15) and weight loss (4/15); two patients were asymptomatic. A preoperative diagnosis of hilar adenocarcinoma occurred in 3/15 patients. The lesion was resected in all but one case in which resection was not conducted because of the extensive involvement of the tumor^[5]. In this case, endoscopic prosthesis was suggested, but the patient refused the procedure with clinical follow-up. The tumor did not recur in any of the resection cases.

Immunohistochemical analysis is necessary to distinguish schwannomas from neurofibromas, gastrointestinal stromal tumors and leiomyomas. Schwannomas are

strongly positive for vimentin and S100 protein and are negative for muscle cell markers and CD117 (kit), which are found in smooth muscle and gastrointestinal stromal tumors. The CD34 antigen is expressed by a distinct cell population in peripheral nerves, nerve sheath tumors, and related lesions. This antigen is also a useful parameter for the immunohistochemical diagnosis of gastrointestinal stromal tumors. Schwannomas in the digestive tract are usually negative for CD34, although Hou *et al*^[14] identified 3 gastrointestinal schwannomas with CD34-positive spindle cells in 33 analyzed cases. Our patient was positive for both the S100 protein, as was expected, and CD34, as in the 3 cases described by Hou *et al*^[14].

While there may be preoperative suspicion, the diagnosis of schwannoma requires intraoperative and histopathological confirmation^[15]. Despite the possible complications, resection is the treatment of choice for such cases^[16]. Schwannomas in the digestive tract have an excellent prognosis after surgical resection, as do schwannomas in other locations. To date, there is no evidence suggesting that these tumors are potentially malignant^[4]. In keeping with these findings, our patient remains asymptomatic after 12 mo of postoperative follow-up.

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