Schwannoma of the biliary tract resembling cholangiocarcinoma: A case report and review

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ABSTRACT

Schwannomas are benign tumours derived from Schwann cells and are extremely rare in the biliary tract. We present the case of a 62-year-old patient with a common bile duct schwannoma that resembled a cholangiocarcinoma. We also review all 17 previously published cases of schwannoma of the biliary tract and discuss the challenges of preoperative diagnosis.

KEYWORDS

Bile ducts – Cholangiocarcinoma – Differential diagnosis – Obstructive jaundice – Schwann cells

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Although cholangiocarcinoma is the most common biliary neoplasm causing obstructive jaundice, other malignancies, such as lymphoma or neuroendocrine tumours can be implicated. Currently, only 17 cases of biliary schwannoma, a benign tumour of the bile ducts, have been reported in the literature. We present the case of a patient with a biliary schwannoma that resembled a cholangiocarcinoma and caused jaundice.

Case report

A 62-year-old man presented with obstructive jaundice and abdominal pain. His past medical history included moderate aortic insufficiency, hepatic steatosis and cholecystectomy. Laboratory tests revealed hyperbilirrubinemia (7.56 mg/dL, normal range 0.2–1.3 mg/dL), raised levels of aspartate transaminase 618 U/L (normal range 4–38 U/L), alanine transaminase (924 U/L, normal range 5–41 U/L), gamma-glutamyl transferase (1034 U/L, normal range 7–32 U/L) and alkaline phosphatase (189 U/L, normal range 4–129 U/L), and an international normalised ratio of 1.05 (normal range <1.3).

Abdominal ultrasound showed mild dilation of the intrahepatic biliary tree, with normal extrahepatic biliary ducts. Magnetic resonance cholangiopancreatography (MRCP) revealed both intrahepatic and extrahepatic bile duct dilation secondary to a focal stenosis near the confluence of the right and left hepatic ducts. Subsequent endoscopic retrograde cholangiopancreatography confirmed the stenosis with proximal dilation (Fig 1). Biliary brushings were benign on cytology and a stent was placed. Computed tomography scan revealed no further information as to extent of the disease. Tumour marker levels were 0.68 ng/mL (normal range 0–3 ng/mL) for carcinoembryonic antigen, 1.85 ng/mL (normal range <10

ng/mL) for alpha-fetoprotein and 30.53 U/mL (normal range <30 U/mL) for carbohydrate antigen 19-9.

The patient was discussed at the multidisciplinary team meeting and a presumptive diagnosis of cholangiocarcinoma was reached. Management options were discussed with the patient, and it was decided to proceed to operative intervention.

At the operation, the biliary tree appeared unremarkable, with no palpable masses. The liver was normal and no

Figure 1 Endoscopic retrograde cholangiopancreatography showing a local stenosis at the main bile duct

evidence of metastatic disease was identified (Fig 2). We performed a resection of the common hepatic duct and choledocus, along with a lymphadenectomy of the hepatic pedicle and a Roux-en-Y anastomosis. A frozen section of the surgical specimen margin was intraoperatively diagnosed as biliary intraepithelial neoplasia stage (BilIN) 2–3, so we widened the resection upstream in the biliary tract.

The postoperative course was uneventful and the patient was discharged after 7 days.

Pathology findings revealed a 0.9 cm plexiform schwannoma of the biliary tract, along with reactive inflammatory cell infiltration (Fig 3 and 4). The proximal surgical margin was finally classified as BilIN-2.

At 18-months follow-up, the patient remained asymptomatic, with no evidence of recurrence.

Discussion

Schwannomas, also known as neurilemmomas, are benign tumours that arise from the Schwann cells, the main component of the neural sheath. They typically arise in the soft tissues of the upper limbs, trunk, head, neck, retroperitoneum, mediastinum and pelvis,^{1,2} but are rarely seen in the digestive tract. Within the gastrointestinal system, most tumours are reported in the stomach, followed by the colorectum and oesophagus. Biliary schwannomas are extremely rare, although the region is densely innervated by both the sympathetic and parasympathetic systems.

Only 17 previous cases have been reported in Medline (Table 1),^{2,5} with a female predominance (12/17), and an average age of 43 years (range 15–64 years). The most common symptom at presentation was jaundice (13/17), followed

Figure 2 Exploratory laparotomy

Figure 3 Histological examination showing the biliary tract (*) and the schwannoma (^)

Figure 4 Histological examination showing a two-component growing pattern

by abdominal pain (9/17). Preoperative diagnosis was only suspected in the case published by De Sena et al, as the patient was already known to suffer from type-2 neurofibromatosis, which is associated with schwannomas.² Unlike cholangiocarcinoma, in which vague abdominal pain is present only in 20% of patients, schwannomas often include this symptom at presentation (9/17), as in our case. This seems rather interesting, even more so when considering that pain in cholangiocarcinoma is associated with advanced stages of disease, whereas it has no relationship with tumour size in schwannomas. Abdominal pain as initial presentation

Table 1 Published biliary schwannoma case reports

Reference	Age	M/F	Presentation	Preoperative diagnosis	Tumour location
Whisnant 1974 ⁶	15	F	Abdominal pain + obstructive jaundice + weight loss		Distal common bile duct
Balart 1983 ⁷	56	F	Abdominal pain + obstructive jaundice	Cholangiocarcinoma vs extrinsic compression	Common hepatic duct
Oden 1995 ⁸	40	F	Abdominal pain + obstructive jaundice	choledocolithiasis	Common bile duct
Jakobs 2003 ⁹	37	M	Abdominal pain + obstructive jaundice	Intraductal benign tumour	Common hepatic duct
Honjo 2003 ¹⁰	48	F	Obstructive jaundice	Benign non-epithelial tumour	Common bile duct
Otani 2005 ¹¹	59	F	Abdominal pain	Remnant choledochal cyst	Intrapancreatic bile duct
Park 2006 ¹²	53	F	Asymptomatic		Porta hepatis
Vyas 2006 ¹³	29	F	Abdominal pain + obstructive jaundice		Common bile duct
Kamani 2007 ¹⁴	39	F	Jaundice + weight loss	Klatskin tumour	Proximal common hepatic duct
Fenoglio 2007 ⁵	41	F	Obstructive jaundice + weight loss	Pancreatic tumour	Middle common bile duct
Jung 2007 ¹	64	F	Asymptomatic		Proximal common bile duct
Madhusudhan 2009 ¹⁵	46	M	Obstructive jaundice	Cholangiocarcinoma	Intrahepatic bile duct
Kulkarni 2009 ¹⁶	38	M	Abdominal pain + jaundice + weight loss	GIST vs lymph nodal mass	Porta hepatis
De Sena 2009 ¹⁷	58	F	Obstructive jaundice in patient with type-2 neurofibromatosis	Biliary schwannoma	Extrahepatic bile duct
Parameshwarappa 2010 ³	38	M	Abdominal pain + obstructive jaundice + weight loss	GIST vs lymph nodal mass	Common bile duct
Panait 2011 ¹⁸	54	F	Gastroesophageal reflux in patient with history of melanoma	Metastatic melanoma	Porta hepatis
Fonseca 2012 ²	24	M	Abdominal pain + obstructive jaundice + weight loss	Klatskin tumour	Proximal common hepatic duct

F = female; GIST = gastrointestinal stromal tumor; M = male

might be explained by the neural origin of schwannomas, although there is no current data to support this.

Tumour markers are usually normal and imaging tests do not render specific findings either. Although ultrasound appears as the initial procedure in most cases, it does not seem to be helpful for diagnosis. CT generally reveals a homogeneous mass with irregular contrast enhancement. Secondary degenerative changes can sometimes be seen, in what Ackerman and Taylor first called ‘ancient schwannomas’ in 1951.⁴ These changes occur with tumour growth and include calcification, cystic degradation, bleeding and hyalinisation.^{1,4} However, schwannomas of the biliary tract have generally been diagnosed at an early stage, and these features will therefore not show up on CT. In our case, only a punctual stenosis was evident on CT, and no signs of macroscopic tumour could be seen.

MRCP findings disclose tumours with low-to-moderate signals in T1-weighted images and heterogeneous high-intensity signals in T2 sequences.⁵ This heterogeneity can be explained by the two-component growing pattern observed under a microscope. On the one hand, Antoni type A areas are hypercellular zones in which cells sometimes develop a nuclear palisading pattern known as Verocay bodies. In contrast, Antoni type B areas show less cellularity, with cuboid cells that are widely scattered over a myxoid matrix.²

Immunohistochemistry can be useful for the differential diagnosis.² Being nerve sheath-derived tumours, schwannomas show positivity for neural markers such as vimentin and S-100 protein. They are negative, however, for muscle markers such as actin and desmin, as well as for CD-117 (c-kit) and CD-34 tests, thus differentiating them from gastrointestinal stromal tumors.⁵

Most schwannomas are diagnosed postoperatively on histological examination, as preoperative diagnosis is almost impossible and these lesions mimic more invasive tumours. As schwannomas are universally benign, long term outcomes are excellent, with no recurrences reported in the literature. Thus, operative resection seems to be curative.

In our case, even if a preoperative diagnosis of a benign biliary lesion had been made, operative resection would still have been advocated, given the progressive nature and location of the patient's tumour to prevent obstructive liver failure.

Conclusions

Schwannomas are benign lesions that can rarely affect the biliary tree and cause obstructive symptoms. They tend to mimic more worrying tumours and there is a lack of preoperative pathognomonic features, which results in an overtreatment of these lesions. All reported cases within the literature have been published following pathological assessment of the surgical specimen. As schwannomas are universally benign, resection should be considered curative as in this case.

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