

Biliary tract schwannoma: A rare case of obstructive jaundice in a middle aged gentleman.

Vishakha Kalikar, Sachin Wani, Roy Patankar, S.K. Mathur

Department of General Surgery, Zen Hospital, India

Abstract: Schwannomas are derived from Schwann cells, the most common sites being upper extremities, trunk, retroperitoneum, head and neck, mediastinum. However they can arise from the gastrointestinal tract too, including the biliary system. We present a case of a 40 year old gentleman, who presented with obstructive jaundice whose computed tomography of the abdomen was suggestive of mass lesion at the porta causing intra hepatic biliary radicle dilatation. And MRCP s/o similar findings with an endosonography s/o hydatid cyst. He underwent local resection with biliary enteric reconstruction and the histopathological examination confirmed a biliary duct schwannoma. On follow at 6 months, the patient is asymptomatic and shows no signs of recurrence. We have also reviewed the literature of biliary tract schwannomas.

Keywords: Biliary Tract, Hilarcholangiocarcinoma, Schwannoma

I. Introduction

Schwannomas are rare tumors of the digestive tract although other non-epithelial tumors such as adenomas, lymphomas can mimic a cholangiocarcinoma. (1,2). The aim of this case report was to put forth a case of a common bile duct schwannoma that caused obstructive jaundice in a middle aged male patient. It was diagnosed during surgery, hence allowing a local resection with reconstruction to be carried forward.

II. Case

We had a 40 year old gentleman, clinically diagnosed with obstructive jaundice, history of hepatitis twice in the past 1 year with 12kgs weight loss in the past 3 months. No history of any addictions. On clinical examination there was no organomegaly. Laboratory studies: Total bilirubin: 12 with direct being 8. SGOT /SGPT: 153/ 168 respectively, alkaline phosphatase being 600. Tumor markers were within normal limits. Computed tomography of the abdomen with oral/iv contrast: 32x29 mm mass at the porta causing intra hepatic biliary radicle dilatation. The adjacent vascular structures and fat planes were preserved. Magnetic resonance cholangiography s/o thin walled cystic mass at the porta. Endosonography s/o hydatid cyst at the porta. We opted for surgical resection of the mass and began with a laparoscopic approach. Intra operatively: a large solid cystic mass arising from the common bile duct with splaying of the CBD anterior to the mass. Decision was taken to convert to open. During dissection we found a well-encapsulated, solid cystic mass with regular margins with no signs of infiltration into the surrounding structures. A cholecystectomy was performed with en bloc resection of the mass along with CBD and a Roux en Y hepaticojejunostomy.

Post operative course of the patient was uneventful. Patient was discharged on day 7 in a healthy condition. Histopathological confirmed it to be a primary biliary schwannoma with free margins and node negative with immunohistochemistry being positive for S100. It was negative for CD 117 kit and CD 34. At a follow up after 6 months the patient is asymptomatic without any signs of recurrence.



Fig 1: Intraoperative findings: solid cystic mass lesion at the porta

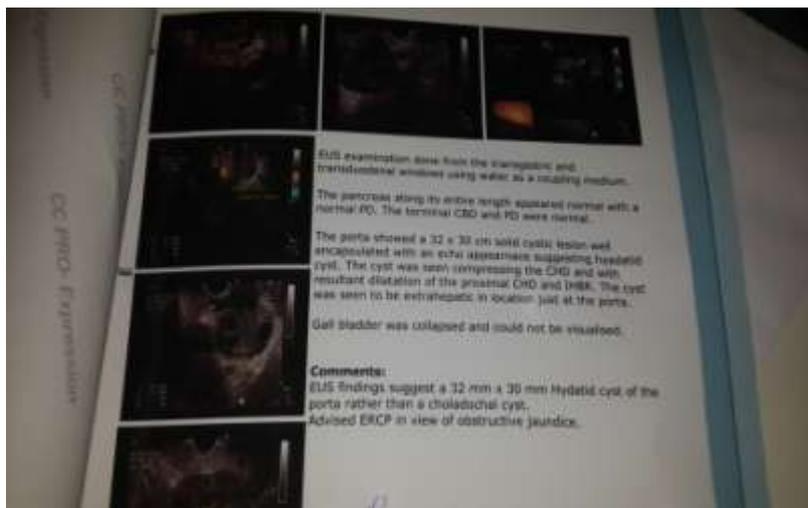


Fig2:EUS report suggestive of hydatid cyst

We found 15 cases in literature of the same as depicted in the table below.

Cases studies of biliary schwannoma in the literature

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

F: Female; M: Male.

III. Discussion

Schwannomas are tumors derived from Schwann cells i.e. from the inner portion of the nerve sheath. (3).The most common locations are upper extremities, head and neck, trunk, retroperitoneum, mediastinum and pelvis. (4). Schwannomas of the gastrointestinal tract are uncommon, the most common sites being the stomach and colon. (5), though they can develop along the biliary tract because of the abundance of the sympathetic and parasympathetic supply along the wall of the gall bladder and the CBD (6) These have a female preponderance and most commonly present with obstructive jaundice.

We have no conclusive diagnostic imaging studies for the same as schwannomas simulate findings similar to other tumors especially cholangiocarcinoma. Histopathologically on gross examination they are usually large, solitary cystic and the nerve of origin doesn't penetrate the tumor. In the mediastinum it is also known as a dumbbell tumor.

Microscopically there are Antoni A which compact and highly cellular, containing spindle cells and Antoni B areas, which consist of loose myxoid tissue containing spindle cells, with degenerative changes. (7) There are 4 types of schwannomas: cellular, glandular, epithelioid and ancient. The cellular type contains Antoni

a type cells, epitheloid and granular contains cells according to their name and ancient type: bizarre hyper chromatic nuclei with cyst formation, calcification and hyalinization.

Immunohistochemistry is essential to distinguish schwannomas from other tumors like neurofibromas, liposarcomas, stromal tumors. Schwannomas are strongly positive for S100 and vimentin but negative for CD 117 kit, which are found in smooth muscle cell tumors. (8) CD 34, which is expressed in the stromal cell tumors, is usually negative in schwannomas. Our patient tested positive for S 100 but negative for CD 34.

IV. Conclusion

The diagnosis of schwannomas finally required histopathological examination. (9). Resection is the treatment of choice for such cases. (10). Schwannomas have an excellent prognosis after surgical resection. To date there has been no data that they can be potentially malignant. (6). biliary tract schwannoma is extremely rare and is mostly seen in the extra hepatic bile duct. A radiological diagnosis is difficult

References

- [1]. Eslick GD. Epidemiology of gallbladder cancer. *Gastroenterol Clin North Am.* 2010;39:307–330.
- [2]. Malhi H, Gores GJ. Cholangiocarcinoma: modern advances in understanding a deadly old disease. *J Hepatol.* 2006;45:856–867.
- [3]. Hajdu SI. Peripheral nerve sheath tumors. Histogenesis, classification, and prognosis. *Cancer.* 1993;72:3549–3552.
- [4]. Kulkarni N, Andrews SJ, Rao V, Rajagopal KV. Case report: Benign porta hepatic schwannoma. *Indian J Radiol Imaging.* 2009;19:213–215.
- [5]. Iiettinen M, Shekitka KM, Sobin LH. Schwannomas in the colon and rectum: a clinicopathologic and immunohistochemical study of 20 cases. *Am J Surg Pathol.* 2001;25:846–855.
- [6]. ung JH, Joo KR, Chae MJ, Jang JY, Lee SG, Dong SH, Kim HJ, Kim BH, Chang YW, Lee JI, et al. Extrahepatic biliary schwannomas: a case report. *J Korean Med Sci.* 2007;22:549–552.
- [7]. Honjo Y, Kobayashi Y, Nakamura T, Takehira Y, Kitagawa M, Ikematsu Y, Ozawa T, Nakamura H. Extrahepatic biliary schwannoma. *Dig Dis Sci.* 2003;48:2221–2226.
- [8]. Hou YY, Tan YS, Xu JF, Wang XN, Lu SH, Ji Y, Wang J, Zhu XZ. Schwannoma of the gastrointestinal tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. *Histopathology.* 2006;48:536–545.
- [9]. de Diego Rodríguez E, Roca Edreira A, MartínGarcía B, HernándezRodríguez R, Portillo Martín JA, GutiérrezBaños JL, CorreasGómez MA, del Valle Schaan JI, Villanueva Peña A, Rado Velázquez MA, et al. [Retroperitoneal benign schwannoma. Report of a new case] *ActasUrol Esp.* 2000;24:685–688.
- [10]. Fenoglio L, Severini S, Cena P, Migliore E, Bracco C, Pomero F, Panzone S, Cavallero GB, Silvestri A, Brizio R, et al. Common bile duct schwannoma: a case report and review of literature. *World J Gastroenterol.* 2007;13:1275–1278.