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Schwannoma in the Hepatic Hilum with Portal Vein Compression: Case Report and Literature Review of 31 Cases

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1. Abstract

Schwannomas originating from the Schwann cell nerve sheath are rare in the hepatic hilum, with only a few cases reported in the literature. We present the case of a 47-year-old male of mixed ethnicity with an incidental lesion in the hepatic hilum detected during routine abdominal ultrasound. Subsequent imaging studies revealed a hypovascularized mass compressing the portal vein, posing a diagnostic challenge due to its atypical location. Following multidisciplinary discussion, surgical resection was performed, and histopathological examination confirmed the diagnosis of schwannoma. This case highlights the importance of considering schwannoma in the differential diagnosis of hepatic hilar masses and underscores the diagnostic and therapeutic challenges posed by such lesions. Surgical resection remains the main treatment, with careful preservation of the capsule to minimize recurrence risk. Long-term follow-up is essential to monitor for potential recurrence, even in benign cases. This report contributes to the understanding of the clinical presentation, diagnostic approach, and management of hepatic hilar schwannomas.

2. Introduction

Schwannomas are neuroectodermal tumors arising from Schwann cells of the nerve sheaths. They most commonly occur in the head and neck region, vestibular system, and extremities. Hepatic hilum schwannoma is a benign tumor, although it is rare and easily misdiagnosed as cholangiocarcinoma [1,2]. In the present study, we sought to describe a case of schwannoma in the hepatic hilus with portal vein compression.

3. Case Presentation

3.1. Patient Information

A 47-year-old man of mixed ethnicity, with no prior symptoms who presented with an incidental finding of a lesion in the pancreas during a routine abdominal ultrasound. The patient was subsequently referred for evaluation at our biliopancreatic service. His medical history was significant for type 2 diabetes mellitus, for which he takes metformin. He does not use any other medications, has not undergone any surgery, and has no family history of neoplasia.

3.2. Clinical Findings

On physical examination, the patient was in good general condition and anicteric. Laboratory blood exams revealed the following elevated results: alkaline phosphatase (112 IU/L), gamma-glutamyl transferase (142 IU/L), total bilirubin (2,1 mg/dL), and direct bilirubin (1,6 mg/dL). Tumor markers were measured, and the results were negative.

3.3. Diagnostic Assessment

A computed tomography (CT) scan was performed (Figure 1), revealing a hypovascular and heterogeneous mass located in the "porta hepatis". The mass was positioned posteriorly to the proper hepatic artery and anteriorly to the portal vein. Its upper portion was in contact with the anterior surface of the hepatic caudate lobe, and its lower portion extended to the pancreatic plateau. Additionally, the left hepatic artery was found to be branch of the left gastric artery, traversing the fissure of the venous ligament (an anatomical variation). This mass measured approximately 7.5 cm at its longest axis. The pattern of the mass resembled that seen in neurogenic lesions. A magnetic resonance imaging (MRI) was subsequently conducted to assess the lesion more accurately. The same pattern was observed: a lesion measuring approximately 7 cm between the portal vein and the hepatic artery, hypovascular, with a high heterogeneous signal on T2-weighted sequences, bulging inferiorly to the superior surface of the pancreatic neck and maintaining an anterior relationship with the duodenal bulb.

3.4 Therapeutic Intervention

The case was extensively discussed at a multidisciplinary meeting of the biliary-pancreatic group at our institution, and surgical resection of the lesion was considered the best management. The patient then underwent a laparotomy, during which a lesion measuring 7 cm in diameter was noted between the portal vein and the proper hepatic artery, pushing the structures towards the tumor, but not invading them. Complete resection of the lesion was possible, in addition to lymphadenectomy of chains 8A, 12A, and 12P (Figure 2). The resected specimen sent further anatomopathological analysis.The pathological and immunohistochemical examination of the surgical specimen revealed a mass weighing 67.2 grams and measuring 7.5 x 6.0 x 4.0 cm, covered by a smooth and shiny capsule, strongly positive for S100 protein, with clear resection margins and lymph nodes free of neoplasia.

3.5. Follow-Up and Outcomes

The patient's postoperative course was uneventful. Laboratory values normalized, and he was discharged on the third postoperative day. Long-term follow-up was recommended to monitor for recurrence, given the potential for late recurrences even in benign schwannomas.

4. Discussion

Schwannomas, also known as neurilemmomas are mesenchymal tumors with low malignant potential, originating from the Schwann cells, which are responsible for producing the myelin sheaths surrounding nerves [3,4]. About 90% of schwannomas are non-

cancerous, accounting for approximately 5% of all benign soft tissue tumors [4,5]. The disease can occur anywhere in the body however, primary schwannoma in the hepatic hilus is rare, with its likely origin being sympathetic and parasympathetic nerve fibers running through the hepatic artery via the hepatoduodenal ligament, as well as through the bile ducts and gallbladder wall [6,7].In 2012 Fonseca et al.6 published a review of 16 cases of biliary Schwannoma. The ages of the patients ranged from 15 to 64 years, and the patients were predominantly female (12/15).6 Later in 2020, Wang et al.4 published a review of the 5 cases of Schwannoma in the hepatic ligament. The male-to-female ratio in the study was 3:2, and the age range of 29 to 62 years. Three patients were asymptomatic [4]. Our literature review 31 cases published over past decades, as show in Table 12,7-9,11-25,33-44, with a mean age of 45 years (± 15). Female patients constituted 71% of cases, and the most frequently reported symptom was abdominal pain (48.39%). Additionally, seven patients were asymptomatic at the diagnosis.Preoperative diagnosis of schwannoma remains challenging due to the absence of a gold standard diagnostic method, none of the cases described in the literature were diagnosed pre-operatively.2 The most commonly utilized imaging modalities are CT and MRI. CT scans typically show a well-defined, round or oval, inhomogeneous low-density mass, often with degenerative changes such as cysts and calcifications. On MRI, schwannomas appear well-circumscribed, hypointense on T1-weighted images, and hyperintense on T2-weighted images [2,26-28].In cases similar to the one we presented, where the lesion is closely related to the structures of the hepatic hilum, it is tough to perform a confirmatory biopsy. One option is fine needle biopsy guided by endoscopic ultrasound. However, some authors express concern regarding the possible spread of malignant cells due to biopsy and therefore do not recommend it. However, there are no reports of dissemination after biopsy in the literature [29].The definitive diagnosis is pathological, histological, and immunohistochemical. Classically, Schwannomas can present histologically as Antoni type A or type B tissue. Antoni type A consists of compact cellular lesions, while Antoni type B consists of loose hypocellular myxoid lesions with microcystic spaces.30 Immunohistochemical analysis typically shows positivity for S-100 protein and negativity for desmin, smooth muscle actin (SMA), CD34, and CD117 [18,24].Surgical resection is the treatment of choice, ensuring preservation of the tumor capsule to minimize recurrence risk.31 Yin et al. [7].Reviewed 15 hepatic hilar schwannoma cases, all treated with surgical resection, though only seven cases achieved complete resection. Um two cases, invasion of the hepatic artery and common bile duct was reported. In only one case was it decided not to indicate surgical resection. Our case aligns with this treatment approach, achieving complete resection without capsule violation. A conventional laparotomy was preferred due to the absence of a preoperative pathological diagnosis. Complete surgical resection is curative, and recurrence is uncommon, eliminating the need for adjuvant therapy. The overall prognosis is excellent [4,32].

5. Conclusion

Schwannomas of the hepatic hilum are extremely rare and are often only definitively diagnosed postoperatively. Surgical resection with preservation of the capsule is crucial for reducing recurrence risk. While histologically benign, careful long-term follow-up is recommended due to the potential for recurrence. This case contributes to the growing understanding of hepatic hilar schwannomas, their clinical presentation, diagnostic challenges, and optimal management strategies.

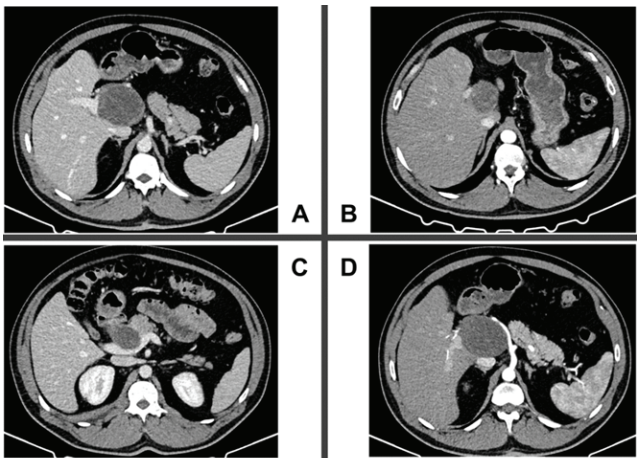

Figure 1: CT scans. (A) Area of contact between the mass and the portal vein b branch. (B) Area of contact between the mass and the caudate lobe of the liver. (C) Compression of the portal vein by the mass. (D) Tumorcontact with the hepatic artery.

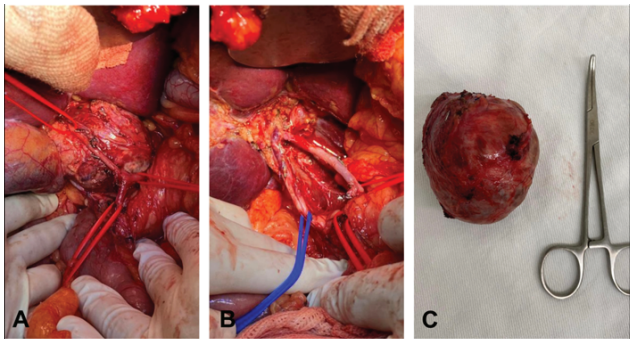

Figure 2: Laparotomy with resection of the mass. (A) Common hepatic artery, gastroduodenal artery, and proper hepatic artery encircled with red vessel loops anteriorly, with the tumor in the hepatic hilum posteriorly. (B) The mass has been resected, showing preserved arteries and the portal vein encircled with a blue vessel loop. (C) Resected tumor.

Table 1: Previous case reports characteristics.

Author	Year	Age (years)	Sex	Presentation	Size (cm)	Primary diagnosis	Site	Intraoperative characteristics
Hulme et al.	2022	37	F	Abdominal pain + palpable mass	14	Biliary cystadenoma or cystadenocarcinoma	Porta Hepatis	The lesion was found to be centered in the porta hepatis and adherent to but not involving the liver.Achieved complete resection.
Filizoglu et al.	2021	36	F	Abdominal pain + pruritus	9	Hilar mass	Porta Hepatis	Achieved complete resection.
Mourra et al.	2020	67	F	Gastric fullness	6.5	Hilar mass	Porta Hepatis	The lesion was extrahepatic, arising from the porta hepatis, and connected to the peri-hepatic artery tissue.Achieved complete resection.

Yoshida et al.	2018	80	M	Abdominal pain	7.0	Abdominal mass	Porta Hepatis	The lesion was seen to arise from the hepatoduodenal ligament, although there was no adherence to either the liver or the pancreatic parenchyma. The lesion was connected to the tissue around the right hepatic artery via a short stalk. Achieved complete resection.
Sebastian et al.	2017	22	F	Abdominal pain	3.5	Hilar mass	Porta Hepatis	Tumor in the porta hepatis without invasion of the vessels or bile ducts. Achieved complete resection.
Xu et al.	2016	31	F	Abdominal pain	11	Hilar mass	Porta Hepatis	Mass in the porta hepatis, wrapping the whole hepatoduodenal ligament, invading the common bile duct, left and right hepatic bile ducts, as well as the right and left hepatic arteries. Achieved complete resection.
Yin et al.	2016	57	F	Abdominal distension	3.5	Spindle Cell Neoplasia	Porta Hepatis	The tumor was located in the porta hepatis, tightly pressing the hepatoduodenal ligament, and the caudate lobe and the pancreas were not involved. Achieved complete resection.
Maruyama et al.	2015	79	M	Asymptomatic	2.8	Hilar mass	Porta Hepatis	Yellow tumor strongly adhered to the left gastric artery and common hepatic artery. Achieved complete resection.
Lopes et al.	2015	30	F	Abdominal pain	3.8	Hilar mass	Porta Hepatis	A full laparotomy revealed the tumor behind the portal vein. Achieved complete resection.
Huang et al.	2011	45	F	Abdominal pain	7.5	Hilar mass	Proper hepatic artery	the lesion was found to be in close contact with the common hepatic artery, the proper hepatic artery (which was totally encroached by the lesion), and the gastroduodenal artery. Achieved complete resection.
Panait et al.	2011	54	F	Asymptomatic	5.2	Metastases or Lymphoma	Porta Hepatis	The tumor was located at the base of segment 4, at the bifurcation of the right and left hepatic ducts, and originated from the common hepatic duct. Achieved complete resection.
Pinto et al.	2011	29	M	Asymptomatic	3.7	Spindle Cell Neoplasia	Hepatoduodenal Ligament	Multilocular cystic mass of the hepatoduodenal ligament. Achieved complete resection.
Kulkarni et al.	2009	38	M	Abdominal pain, jaundice and weight loss	4.5	Hilar mass	Porta Hepatis	Inseparable mass from the common bile duct. Achieved complete resection.
Zhang et al.	2009	42	F	Asymptomatic	2.2	Left hepatic lobe mass	Hepatoduodenal Ligament	NA
De Sena et al.	2009	58	F	Obstructive jaundice		Biliary schwannoma	Extrahepatic bile duct	NA
Madhusudhan et al.	2009	46	M	Obstructive jaundice		Variable polypoid cholangiocarcinoma	Intrahepatic bile duct	NA Surgical resection was not performed.
Kamani et al.	2007	39	F	Obstructive jaundice + weight loss		Klatskin tumor	Proximal portion of the common hepatic duct	NA
Fenoglio et al.	2007	41	F	Obstructive jaundice + weight loss			Middle segment of the common bile duct	The mass involved the intermediate tract of the common bile duct. Achieved complete resection.

Jung et al.	2007	64	F	Asymptomatic			Proximal portion of the common bile duct	Several well-demarcated nodules Ian the bile duct. Achieved complete resection.
Park et al.	2006	53	F	Asymptomatic	4.5	Hilar mass	Porta Hepatis	Yellowish, cystic, and encapsulated mass attached to portal vein, common bile duct, liver, and duodenal wall above the hepatoduodenal ligament. Achieved complete resection.
Wang et al.	2006	45	M	Abdominal distension + nausea	7	Klatskin's tumor	Porta Hepatis	NA
Vyas et al.	2006	29	F	Abdominal pain + obstructive jaundice			Common bile duct	NA Achieved complete resection.
Otani et al.	2005	59	F	Abdominal pain			Remnant bile duct (pancreatic portion)	NA Achieved complete resection.
Jakobs et al.	2003	37	M	Abdominal pain + obstructive jaundice		Intra-ductal benign tumor	Common hepatic duct	NA
Honjo et al.	2003	48	F	Obstructive jaundice		Benign non-epitelial tumor	Common bile duct	Tumor just anteriorly to the extrahepatic bile duct. Achieved complete resection.
Choi et al.	2001	37	M	Asymptomatic	5.0	Hilar mass	Porta Hepatis	The mass was found to be encasing the right hepatic artery and appeared to be arising from it. The mass was otherwise well separated from adjacent solid organs, bile ducts, and other major vessels. Achieved complete resection.
Huang et al.	1996	41	M	Abdominal pain	7.2	Abdominal mass	Hepatoduodenal Ligament	NA
Fang et al.	1995	33	F	Abdominal distension	4.5	Portal Hypertension and Hilar mass	Porta Hepatis	NA
Balart et al.	1983	56	F	Abdominal pain + obstructive jaundice		Cholangiocarcinoma or extrinsic compression of the bile duct	Common hepatic duct	Complicated by liver abscess, treated with drainage. Achieved complete resection.
Whisnant et al.	1974	15	F	Abdominal pain + weght loss + obstructive jaundice			Distal portion of the common bile duct	NA Achieved complete resection.
Oden et al.	1955	40	F	Abdominal pain + obstructive jaundice		Choledocholithiasis	Common bile duct	NA

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