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Periportal Schwannoma: Two Case Reports and Review of Periportal Mass

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Abstract

Introduction: We present two cases of periportal schwannoma and discuss management of periportal mass.

Materials and Methods: The first case is a 55-year-old female who presented with a six-month history of abdominal pain and fifteen-pound weight loss. Magnetic Resonance Imaging (MRI) showed a 5cm complex periportal mass. The mass originated from the proper hepatic artery from which it was carefully dissected away and removed laparoscopically. The second patient was a 33-year-old female who presented with three weeks of upper abdominal and back pain. MRI showed cholelithiasis and an incidental 6.2cm mass in liver segment IV-B extending into the periportal region. The mass was enmeshed with surrounding biliary structures. Intraoperative frozen section of the periportal mass confirmed it as a benign schwannoma. This patient underwent laparoscopic cholecystectomy but the mass was not resected.

Results: The first patient was discharged on postoperative day two. Histopathological examination confirmed the mass as a benign cystic schwannoma. She remains asymptomatic and without any signs of recurrence after twelve months follow-up. The second patient was discharged on postoperative day one. Final biopsy pathology showed a plexiform schwannoma. She remains asymptomatic after three years of follow-up.

Conclusion: To our knowledge, our first case is the only reported case of laparoscopic resection of periportal schwannoma arising from the proper hepatic artery. If intraoperative frozen section can identify a schwannoma as benign, the mass can be left alone, and a major operation can be avoided, as prognosis is excellent and malignant transformation of periportal schwannoma has not been reported.

Keywords: Schwannoma; Periportal mass; Schwannoma laparoscopic resection

Abbreviations

MRI: Magnetic Resonance Imaging; CT: Computed Tomography; IOC: Intraoperative Cholangiogram

Introduction

Schwannomas are benign tumors that arise from Schwann cells of peripheral nerve myelin sheaths. Initially reported in 1988 by Daimaru et al. [1], gastrointestinal schwannomas have an excellent prognosis after surgical resection. Patients with periportal schwannomas can present with abdominal pain or concerning features such as weight loss, jaundice or anorexia raising suspicion for a malignant tumor [2]. Schwannomas characteristically undergo cystic degeneration due to vascular thrombosis and subsequent necrosis [3]. Computed Tomography (CT) scan shows a well-defined, hypodense, heterogeneous mass with peripheral enhancement [4] making the differentiation from a malignant tumor even more difficult. Only 15 cases of periportal schwannomas have been described in the literature, and preoperative diagnoses could not be made in any of them [2]. All of these patients underwent open surgical resection.

We present two cases of periportal schwannomas approached laparoscopically. The first patient underwent laparoscopic resection of a periportal schwannoma arising from the proper hepatic artery. To our knowledge, this is the first reported case of laparoscopic resection of periportal schwannoma. The second patient underwent a diagnostic laparoscopy, and the periportal mass was confirmed as a benign schwannoma by intraoperative frozen section. The tumor was encased around the extrahepatic biliary tree, so it was left in-situ to avoid unnecessary extensive biliary surgery.

Materials and Methods

Case 1

A 55-year-old Caucasian female presented with six months of severe abdominal pain, early satiety, bloating, steatorrhea and a fifteen-pound weight loss. Past surgical history was significant for laparoscopic cholecystectomy. Her laboratory studies (complete blood count, liver function test) and tumor markers including Carcino Embryonic Antigen (CEA) and Cancer Antigen 19-9 (CA 19-9) were within normal limit. Both upper and lower endoscopies were normal. Abdominal CT showed a 5cm well-defined hypodense mass in the porta hepatis (Figure 1a). No enlarged hilar lymph nodes were identified. This was confirmed on an MRI as a 5cm complex,

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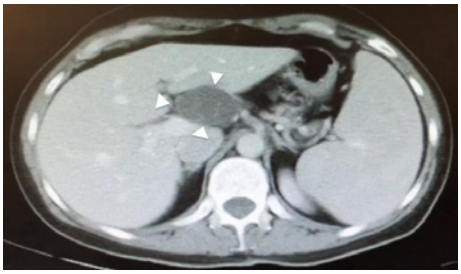

Figure 1a: Portal phase of CT abdomen showing a mass in the porta hepatis (arrowhead).

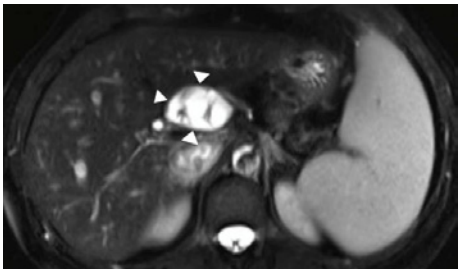

Figure 1b: T2 weighted MRI showing a 5cm complex, septated cystic mass adjacent to porta hepatis separate from the pancreas and biliary tree (arrowhead).

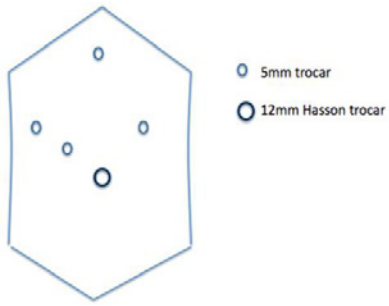

Figure 2: Placement of laparoscopic trocars.

septated cystic mass, hyperintense on T2 signal, adjacent to porta hepatis but separate from the pancreas and biliary tree (Figure 1b). There was no intra or extra-hepatic bile duct dilatation. A decision was made to pursue laparoscopic resection of the periportal mass based on the extent to which the symptoms affected the patient's quality of life. Intraperitoneal access was gained through a Hasson cannula and abdomen was insufflated with CO_2 pneumoperitoneum to a pressure of 12mmHg. Four 5-mm trocars were placed in the upper abdomen (Figure 2). After introduction of the laparoscope (KARL STORZ, Tuttlingen, Germany), the mass was easily identified posterior to the hepatic artery (Figure 3a and 3b). It was dissected carefully from surrounding structures using an ultrasonic dissector (SonoSurg, Olympus, Tokyo, Japan). A network of peripheral nerves was seen to be entering the tumor. The mass appeared to arise from the proper hepatic artery. The cystic duct stump from prior cholecystectomy was identified by the presence of surgical clips (Figure 4a). An intraoperative cholangiogram (IOC) through the cystic duct demonstrated normal filling of the bile duct and no communication with the periportal mass (Figure 4b). The incision

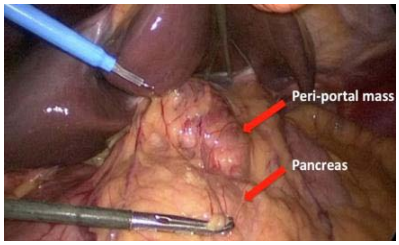

Figure 3a: Laparoscopic view of periportal mass.

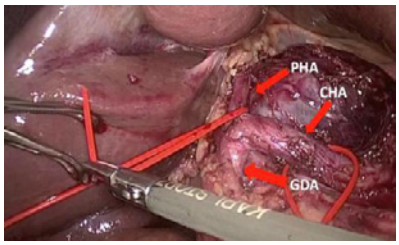

Figure 3b: After initial dissection, the mass can be visualized posterior to the hepatic artery. (PHA: Proper hepatic artery, CHA: Common hepatic artery, GDA: Gastro-duodenal artery).

on the cystic duct stump was closed with a 3-0 vicryl suture (Figure 4c). A clip was placed across the stalk of the mass where it seemed to be arising from the proper hepatic artery. The mass was removed safely in an endoscopic specimen retrieval bag. The duration of the operation was 180 minutes, and blood loss was around 25mL.

Case 2

A 33-year-old female with a history of scoliosis and recent submandibular schwannoma resection presented with three weeks of upper abdominal and back pain. Abdominal CT scan reported a cystic lesion measuring 4.6cm in the porta hepatis, which was later confirmed on MRI. There was no intra or extrahepatic biliary duct dilation, ruling out a choledochal cyst. Preoperative labs including complete blood count, liver function tests, and tumor markers were normal. After obtaining informed consent, the patient was taken to the operating room given her intractable and chronic symptoms. She subsequently underwent diagnostic laparoscopy, cholecystectomy, and biopsy of the periportal mass. Intraoperatively, the mass was seen near the Calot's triangle, separate from the liver parenchyma and biliary tree as confirmed by IOC. A frozen section of the biopsy reported it as a benign-appearing schwannoma. Since the mass was enmeshed with the surrounding structures in the hepatoduodenal ligament, a decision was made not to perform the resection. The total operative time was 165 minutes with an estimated blood loss of 50mL.

Results

In the case of the first patient, pathology showed mixture of hyper-cellular cells with a palisading pattern and hypo-cellular cells confirming it as schwannoma with cystic degenerative changes. On immunohistochemistry, the mass stained positive for S-100 and hematin. It was negative for HMB-45, desmin, and keratin. All these findings supported the diagnosis of a benign periportal schwannoma. The patient's clinical course was uneventful, and she was discharged on postoperative day two with complete symptom resolution by postoperative day ten. No relapse has been observed after twelve

Figure 4a: The cystic duct was identified by the presence of cystic duct clip placed during previous laparoscopic cholecystectomy.

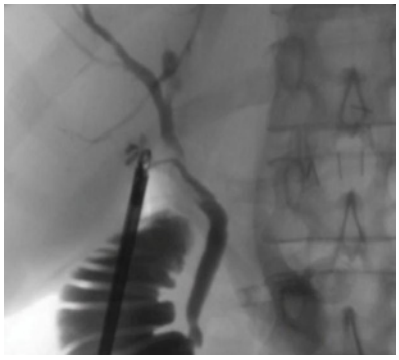

Figure 4b: An intraoperative cholangiogram performed through the cystic duct demonstrated normal filling of the bile duct and no communication with the periportal mass.

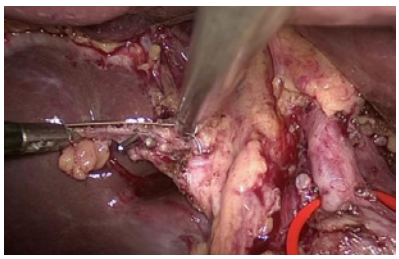

Figure 4c: The cystic duct was closed laparoscopically with a 3-0 vicryl suture.

months follow-up.

In the second case, the patient was discharged on postoperative day one. Repeat ultrasound at three and six month's demonstrated stable size of the schwannoma. She reported complete resolution of her symptoms at six weeks postoperatively. She remains asymptomatic at three years follow-up.

Discussion

Schwannomas, also known as neurilemomas, are benign tumors that arise from the Schwann cells of peripheral nerve myelin sheaths. These tumors are relatively uncommon with a poorly defined incidence and can technically occur in any part of the body except in optic and olfactory nerve tissue. They are usually found in the head, neck, trunk and flexor surfaces of extremities [5]. Rarely do they occur in the visceral abdomen and the retroperitoneum, with an incidence rate less than 1%. Less than fifty cases of pancreatic schwannoma [6] and less than twenty cases of extra-hepatic biliary

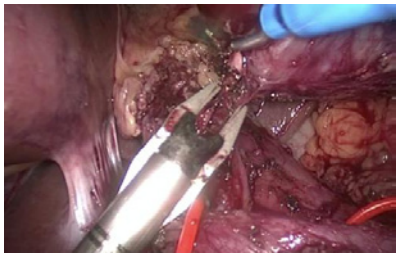

Figure 5: Mass stalk connected to the proper hepatic artery is being clipped. Red vessel loop is pulling the common hepatic artery.

schwannoma have been reported in the literature [7-9]. Otani et al reported a schwannoma arising from the remnant choledochal cyst [10]. Schwannoma stemming from the porta hepatis is extremely rare, and only fifteen such cases have been described over the last thirty years [2]. Schwannoma arising from an abdominal artery is rare but possible because of network of sympathetic and parasympathetic nerve fibers present on the vessel wall. Only one case of schwannoma originating from the proper hepatic artery has been reported before our case [11]. We report 2 cases of periportal schwannoma (one arising from the proper hepatic artery) from our institution.

Gastrointestinal schwannomas are slow growing and tend to occur more frequently in middle-aged women between the ages of 30-60 years of age [7]. With periportal schwannomas, patients usually are initially asymptomatic but may present later with symptoms of obstructive jaundice due to mass effect. Radiological appearance is highly variable and correlates with the ratio of organized cellular components to myxoid tissues as well as the amount of cystic degeneration present [12]. On ultrasound, schwannomas appear as complex cystic masses with hyperechoic solid areas [13]. On CT imaging, schwannomas appear sharply demarcated homogenous mass, and those with high amounts of Antoni A areas appear targetoid with central enhancement [3,14]. On MRI, schwannomas show hyperintensity on T2- weighted imaging and hypointensity on T1-weighted imaging [15]. Faster growth, lack of demarcation and presence of invasion, contrast enhancement, irregular borders, and vascular thrombosis suggest malignancy [16]. Gastrointestinal schwannomas have avid uptake on the PET scan and can be misinterpreted as a malignant lesion [17].

Periportal masses on imaging can have a wide differential. However, some of the common and important differentials include gastrointestinal stromal tumors (GIST), cholangiocarcinoma, choledochocele, primary or secondary lymphoma, enlarged lymph nodes and low-grade sarcoma [18-20]. When patients present with abdominal pain or jaundice, and a periportal mass is detected in image scans, the diagnosis of a cholangiocarcinoma or a GIST may be considered first. Hilar cholangiocarcinoma is most commonly infiltrative and less commonly mass-forming or polypoidal. MRCP demonstrates a hilar stricture with biliary enhancement. A hypo- to iso-attenuating or isointense mass is characteristically seen on arterial phase and venous phase images, with progressive enhancement in the delayed phase [13]. GIST has a more heterogeneous appearance on the CT compared to schwannoma [14]. Primary and secondary lymphomas of the gastrointestinal tract may have a similar CT appearance to schwannomas, but lymphomas are commonly

Table 1: Common differential diagnosis of periportal mass and their specific features.

	Peri-portal Mass	Clinical features	CT	MRI	Histology	IHC
1.	Periportal Schwannoma	Abdominal pain, Jaundice, Anorexia, Nausea NF1 or NFII	Well-defined hypodense heterogenous Contrast enhancement often showing secondary degeneration	T1 – hypointense T2 – hyperintense, heterogeneous	Macroscopic – Spheroidal, cystic degeneration, Hemorrhage Microscopic - Antoni A area: hypercellular, nuclear palisading and Verocay bodies, Antoni B area: hypocellular	S-100 positive CD-117 positive
2.	GIST	Abdominal pain, anemia, emesis, gastric outlet obstruction	Heterogenous, peripheral enhancement with central non-enhancement due to necrosis	T1 – low signal intensity, solid component with peripheral enhancement T2 – high signal intensity, solid component	Spindled nuclei with pale eosinophilic cytoplasm. Cells are frequently palisading with variable mitotic activity	CD 34 and CD 117 positive
3.	Choledochal Cyst	Abdominal pain, right upper quadrant mass, jaundice	Segmental or diffused dilatation of CBD. Water-like hypodense attenuation due to filling with bile, may have thickened wall if cholangitic	Coexistent underlying anomalous pancreaticobiliary junction. T2 - hyperintense signal with contrast filling of cyst	Fibrosis of cyst wall columnar epithelium, lymphocyte infiltration and increased epithelial metaplasia	
4.	Cholangio-carcinoma	Abdominal pain, jaundice, weight loss	Mass forming – Arterial phase shows ragged rim enhancement; Portal phase shows gradual centripetal enhancement Periductal infilterating – thickening of the periductal parenchyma with narrowed or dilated duct Intraductal tumor – Alteration in duct caliber filled with intraductal mass which enhances with contrast CT	Mass forming – T1 – hypointense T2 – hyperintense Capsular retraction, heptolithiasis Periductal infilterating – equilibrium phase - periductal enhancement around the narrowed or dilated intrahepatic duct Intraductal tumor – Alteration in duct caliber filled with intraductal mass which enhances with contrast MRI	Tubules, acini and solid nests of adenocarcinoma embedded in desmpolastic stroma Tumor spreads along hepatic plates, duct walls and via nerves	CK 7 and CK19, positive CK 20 and CEA negative
5.	Lymphoma	Fever, chills, night sweats, lymphadenopathy, weight loss, fatigue	Single dominant solid hypoattenuating lesion Adenopathy in the supporting mesenteries and retroperitoneum	T1 -hypointense T2 - variable intensity	Depends upon the type	
6.	Secondary Metastasis	Blood loss anemia, weight loss, fatigue, abdominal pain	Hypoattenuating on non-contrast CT	T1 - Moderately hypointense T2 - Mild to moderately hyperintense	Depending upon the primary source of metastases	

CBD: Common Bile Duct; NF: Neurofibramatosis; GIST: Gastrointestinal Tumor; IHC: Immunohistochemistry.

accompanied by adenopathy in the supporting mesenteries and retroperitoneum [14]. Choledochal cyst shows segmental or diffuse dilatation on imaging and MRCP also depicts a coexistent underlying anomalous pancreaticobiliary junction [13]. Adenoma typically involves the extra-hepatic bile duct, and it is seen as an intra-luminal polypoidal mass isoechoic to the liver with possible dilatation of the biliary tree on imaging [21]. Granular cell tumor, are usually small tumors (<3cm) and imaging typically shows a short segment biliary stricture with proximal dilatation though CT may show a small heterogeneous soft-tissue mass [21]. Sarcoma can present as a large mass with heterogeneous attenuation, signal intensity, and enhancement; surgical excision is required for definitive diagnosis [13].

It becomes challenging to get a specimen by fine needle aspiration when the mass is high up in the porta-hepatis surrounded by blood vessels. Retrieving a sufficient quantity of cells has been a problem, as schwannomas tend to be hypocellular [22]. Even adequate specimens may yield nonspecific cytology [23]. Intraoperative tissue biopsy with frozen section can be done safely under vision [6,24], as in our second case.

Histologically, schwannomas originating in the gastrointestinal tract are composed of cellular Antoni A areas that do not show the nuclear palisading pattern found in conventional schwannomas of the soft tissues [1]. These are interspersed with other areas of loosely organized gelatinous and myxoid components with scattered thick walled blood vessels – Antoni B areas [25]. Moreover, schwannomas of the digestive tract were recently reported to lack neurofibromatosis-II genetic alterations, which support the theory that schwannomas of the gastrointestinal tract are unique tumors distinct from conventional schwannomas [26]. On immunohistochemistry, they are positive for S-100 staining, attributable to spindle cells in Antoni B areas, which helps in distinguishing them from neurofibromas [27]. They are also positive for Glial Fibrillary Acidic Protein (GFAP) [2]. Schwannomas are negative for CD-34 and CD-117, which helps to rule out GIST though there are few case reports where gastrointestinal schwannomas were CD-34 positive [28]. Negative staining for desmin and keratin virtually eliminates tumors of mesenchymal origin.

There are about eighteen cases of laparoscopic resection of gastrointestinal schwannoma arising from the retroperitoneum, but none of these were arising from the porta hepatis [29]. The first case in our series is the first reported case of laparoscopic resection of periportal schwannoma. Moriya et al analyzed forty-seven cases

of pancreatic schwannoma reported in the literature, of which five had malignant changes [6]. Even though they concluded large tumor and associated cystic changes to be associated malignant changes but on detailed analysis, only one case represented true malignant pancreatic schwannoma [30]. Madusudhan et al reported a rare case of biliary schwannoma involving both intra and extra-hepatic biliary tract. Authors did not resect the tumor because of extensive involvement of the biliary tract [21]. The management is similar to our second case in the series where we decided not to resect the tumor because of extensive involvement of the biliary tract. Periportal schwannomas have an excellent prognosis after a surgical resection, similar to conventional schwannomas. There is no evidence to date that these tumors have a malignant potential [2,18,31] and they can be monitored closely if they are encasing important biliary structures to avoid an unnecessary radical biliary surgery.

Conclusion

To our knowledge, this is the first reported case of laparoscopic resection of periportal schwannoma. With proper identification of anatomy using IOC and careful sharp dissection, a periportal schwannoma can be resected laparoscopically safely with quick postoperative recovery. If the schwannoma is encasing important biliary structures then it may require extensive biliary surgery. However, if intraoperative frozen section can confirm it as a benign schwannoma then the mass can be left alone and a major operation can be avoided, as it has an excellent prognosis and malignant transformation of periportal schwannoma has not been reported.

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