Extrahepatic Bile Duct Neurilemmoma Mimicking Klatskin Tumor

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Neurilemmoma rarely develops in the biliary tree. Here, we report a 39-year-old Iranian woman with neurilemmoma in the extrahepatic bile duct presenting with progressively deepening jaundice. On the basis of clinical and radiological features, this tumor was initially suspected as Klatskin tumor. Histologically, the tumor was a typical neurilemmoma. Immunostaining showed that tumor cells were strongly and diffusely positive for S-100 protein, which supported the diagnosis of neurilemmoma. Neurilemmoma should be considered in the differential diagnosis of obstructive jaundice.

Keywords: Extra-hepatic bile duct • Klatskin tumor • neurilemmoma • schwannoma

Introduction

Neurilemmoma of the extrahepatic bile duct is a rare tumor. Three cases have been previously described in the English literature.1-3

On the basis of clinical and radiological findings, preoperative diagnosis of neurilemmoma is difficult. Most of common bile duct (CBD) obstructions are caused by stones and malignant epithelial derived tumors. Benign mesenchymal tumors of the biliary tree are uncommon.4,5

The present case is a 39-year-old woman with hyperbilirubinemia because of extrahepatic bile duct neurilemmoma mimicking a Klatskin tumor.

Case Report

A 39-year-old Iranian woman was admitted to the gastroenterology Department because of a three-month history of progressively deepening jaundice. She complained of loss of appetite, weight loss, dark urine, and colorless stool. Her medical history was unremarkable. On physical examination, yellow colored bulbar conjunctiva and epigastric fullness, without organomegaly were noted.

Laboratory findings

White blood cells count was 6200/mm³ (normal range: 4800 to 10,800/mm³) with normal range of differential count; red blood cells, 2620 x 10⁶/mL (4200 – 5400 x 10⁶/mL); hemoglobin, 8.4 g/dL (12 – 16 g/dL); hematocrit, 25.5% (37 – 47%); and platelet count, 272 x 10⁹/mL (130 x 10⁹ – 400 x 10⁹ /mL).

Serum total bilirubin level was 8 mg/dL (0.1–1.2 mg/dL); direct bilirubin level, 5.6 mg/dL; aspartate amino transferase, 308 IU/L (up to 31 IU/L); alanine amino transferase, 442 IU/L (up to 31 IU/L); alkaline phosphatase, 4372 IU/L (64 – 306 IU/L); lactate dehydrogenase, 35 IU/L (up to 480 IU/L); amylase, 132 IU/L (up to 22 IU/L); blood urea nitrogen, 17 mg/dL (8 – 20.1 mg/dL); creatinine, 0.7 mg/dL (0.7 – 1.2 mg/dL); and glucose, 95 mg/dL (60 – 110 mg/dL).

Negative laboratory tests including HBsAg, IgM anti-HBc, IgM anti-HAV, and HCV RNA ruled out viral hepatitis. Urinalysis showed dark yellow-colored urine with acidic pH. Antinuclear antibody (ANA), antimitochondrial antibody (AMA), and antismooth muscle antibody (ASMA) tests were also negative.
Radiological findings

Computed tomography of the abdomen showed that intrahepatic bile ducts of the left liver lobe and the bile duct on porta hepatica region were slightly dilated, but the cause of obstruction was not visible (Figure 1). So, endoscopic retrograde cholangiopancreaticography (ERCP) and magnetic resonance cholangiopancreaticography (MRCP) were recommended.

ERCP demonstrated a stricture at the hepatic duct bifurcation with dilation of intrahepatic ducts. The most probable differential diagnosis was cholangiocarcinoma.

MRCP revealed moderately dilated intrahepatic ducts. The left hepatic duct was about 7.5 mm in diameter and the right one was 6 mm. There was an irregularly outlined stenotic lesion through proximal portion of the common hepatic duct with a diameter of about 2 mm. Pancreatic ducts, middle, and lower third of the extrahepatic bile ducts were normal. The findings were interpreted as an obstructive lesion in the porta hepatis, indicating Klatskin tumor.

Surgical procedure

On laparotomy, a friable mass was identified at the bifurcation site of the right and left hepatic ducts. The left one was unresectable, impressing a malignant tumor. Some biopsies were taken (by milking the lumen) and then a Nélaton tube was inserted into the right hepatic duct. Postsurgical cholangiography is shown in Figure 2.

Pathological findings

The specimen consisted of several soft tissue fragments measuring 1.5 x 1 x 0.5 cm in aggregate. Microscopically, the tumor was solid and composed of sheets of spindle cells arranged in short bundles or fascicles. The cytoplasm was eosinophilic and fibrillar and the nuclei were elongated and twisted with single, small, and round nucleoli. Mitotic figures were absent. Interlacing fascicles of spindle cells showed foci of palisading arrangement of the nuclei (Verocay body) (Figure 3). The hypocellular areas of the tumor showed myxoid change and perivascular hyalinization. Immunohistochemical staining for S-100 protein (DAKO, Denmark: dilution, 1:3000) was strongly and diffusely positive (Figure 4). The final diagnosis was obstructive jaundice caused by a neurilemmoma of the bile duct.

Clinical course

On the second operation, hepaticojejunostomy was performed. The postoperative course was uneventful and after six months, the patient was completely free of symptoms or any clinical signs of recurrence.

Discussion

Obstructive jaundice is a common finding that is caused by gallstones, neoplasms, or inflammatory disorders. Extrinsic compression can...
also induce obstructive jaundice.

Carcinoma of the biliary duct is a rare neoplasm, which accounts for less than 0.5% to as many as 4% of malignancies. According to its location, it is categorized as hilar, middle, or lower level on the basis of the level of surgical procedure. Klatskin tumor is nominated for those of the hilar type or those occurring at the bifurcation of the left and right hepatic ducts. This is the most common site for carcinoma. The usual finding is a well- to moderately-differentiated tubular adenocarcinoma.4 – 6

Benign tumors of the biliary tree are also uncommon. Three out of every 2500 cases examined are affected.7, 8 Adenoma is the most common lesion. The others include fibroma, leiomyoma, adenomyoma, hamartoma, lipoma, and neural derived tumors such as neurona, neurofibroma, neurilemmoma, and granular cell tumor.7, 8

Review of the literature shows several cases of granular cell Sc, granular cell neurofibroma, and granular cell myoblastoma, which are older terms for granular cell tumors. It is a benign neural tumor characterized by large granular-appearing eosinophilic cells.9

Three cases of schwannoma of the common bile duct have been described before in the available literature (Table 1).

Neurilemmoma is derived from Schwann cells, the lining cells of the nerve sheath. It is a benign nonrecurring tumor of adulthood with no sex predilection in its classical form.9 – 11

Its anatomical distribution is very wide, such divers locations as the cranial nerves, bone, or the gastrointestinal tract (stomach, colon, and small intestine), liver, and pancreas. In most cases it is developed in flexor surfaces of extremities, neck, mediastinum, and retroperitoneum.9 – 13

The hallmark of neurilemmoma is the pattern of alternating Antoni A and B areas, with varying relative amounts. The type A area is cellular, composed of spindle cells, often arranged in a palisading fashion, or in an organoid arrangement (Verocay bodies). The type B areas are far less orderly and less cellular. Cells are arranged haphazardly in the loosely textured matrix containing blood vessels with hyaline thickening of the adventitia.9 – 11

One reported case of extrahepatic bile duct neurilemmoma has been described with different histologic pattern. Lymphoid aggregates were present, which were different from usual type of
soft tissue neurilemmoma. This feature is usually seen in gastrointestinal neurilemmoma. 

Most neurilemmomas demonstrate S-100 protein strongly and diffusely. Immunohistochemical methods are required to distinguish neurilemmomas from neurofibromas, gastrointestinal stromal tumors (GISTs), or even leiomyomas.

Neurilemmoma behaves in a benign fashion. Malignant change is rare and can be discounted.

Based on clinical and radiologic features, the differential diagnosis of CBD tumor is broad. Definite diagnosis of neurilemmoma of CBD can only be established by histopathological examination. This potentially curable tumor should be included in the differential diagnosis of obstructive jaundice.

**References**


