Common Bile Duct Schwannoma Associated with a Choledochal Cyst

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Introduction:
Myelin sheath tumors are neurogenic tumors which include schwannoma, neurofibroma, neurofibromatosis and neurogenic sarcoma. They are most commonly present during middle age and 90% are benign. It has been reported that, they are twice as common in women as in men, but there are few reports of neurogenic tumors in the biliary tract and fewer than 10 cases of schwannoma of the extrabiliary tract.

The most common neurogenic tumor is the Schwannoma (neurilemoma) arising from perineural Schwann cells in the nerve sheaths of the peripheral nerves. They are benign, slow-growing tumors, well circumscribed with a defined capsule. The most common locations are the central nervous system, extremities, neck, mediastinum, and retro-peritoneum. Schwannomas occurring in the biliary tract are extremely rare and have unique characteristics. We present a case of common bile duct schwannoma associated with a choledochal cyst.

Case Report:
A 52-year old Bangladeshi male with a past history of Type 2 diabetes mellitus and hypertension presented with intermittent upper abdominal pain for two weeks, high grade fever with chills and rigors, sweating and weight loss. Both clinical review and family history were unremarkable. The physical examination showed fever of 39°C. There was no hepatomegaly or splenomegaly. There was neither palpable mass nor enlarged lymph nodes in the neck or at the groin.

Laboratory results were: White blood count 5.1x10^6 /ul (normal range (NR) 4-11); total bilirubin 17.5 umol/L (NR 3.5-24); serum pancreatic amylase 34 U/L (NR 3-53); serum lipase 45 U/L (NR 13-53); alkaline phosphatase 176 U/L (NR 40-129); alanine aminotransferase (ALT) 143 U/L (NR 0-40); aspartate aminotransferase (AST) 152 U/L (NR 0-37); and lactate dehydrogenase (LDH) 940 U/L (NR 240-480). The coagulation profile was normal.

Abdominal ultrasonography demonstrated a complex mass adjacent to the head of the pancreas leading to suspicion of either an infected pancreatic pseudo-cyst or an infected choledochal cyst.

A CT scan of the abdomen (Figure 1) showed a well-defined (approximately 5 x 4 cm) hypodense cystic lesion related to the posterior aspect of the head of the pancreas and inseparable from the mid-portion of the common bile duct. There was no intrahepatic biliary dilatation. The possibilities were either a pancreatic pseudocyst or an infected choledochal cyst.

Figure 1: A CT scan of the abdomen showed a well-defined 5 x 4 cm hypo dense cystic lesion related to the posterior aspect of the head of the pancreas.

MRI of the upper abdomen supplemented with MR cholangiography (Figure 2) showed an abnormal cystic mass measuring approximately 6 x 5 cm related to the middle part of the common bile duct and superior and posterior to the pancreatic head. It had a high signal intensity on T2 weighted images. The lower part of the common bile duct was not displaced laterally as would be expected from a pancreatic head neoplasm. Radiologically, it was consistent with a choledochal cyst. The gallbladder was well visualized and appeared normal.

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Figure 2: MRI of the upper abdomen supplemented with MR cholangiography revealed abnormal cystic mass measure 6 x 5 cm related to the middle part of the common bile duct

An exploratory laparotomy showed a fusiform Type 1 choledochal cyst. There was marked thickening of the wall of the choledochal cyst and common bile duct. The choledochal cyst was resected along with the gall bladder and the biliary tract was reconstructed with a Roux-En-Y hepaticojejunostomy.

Histopathologic examination disclosed a cystic lesion which morphologically showed a cellular spindle cell lesion with central palisading (Verocay bodies) (Figure 3).

Figure 3: A medium power view of hematoxylin and eosin (H&E) stained tissue from the resected cyst wall showing a cellular spindle cell lesion with central palisading (Verocay bodies). Antoni Type A

Immunohistochemical staining using anti S100 protein antibodies demonstrated diffuse strong cytoplasmic staining of the tumor cells which is consistent with a benign schwannoma arising in a choledochal cyst (Figure 4).

Figure 4: Immunohistochemical staining using anti S-100 protein antibodies demonstrating diffuse strong cytoplasmic staining of the tumor cells

Discussion:

Cystic dilation of the biliary ducts, also known as a choledochal cyst, is an uncommon but serious condition that requires surgical treatment. However, benign tumors of the biliary tract are extremely rare with schwannoma being the most common. A Schwannoma (Neurilemmoma) is a benign tumor derived from Schwann cells (the lining cells of the nerve sheath) presenting in young to middle-aged adults of both genders equally. This tumor can occur almost anywhere in the body, but the most common sites are the central nervous system, flexor surfaces of extremities, neck, mediastinum, and retroperitoneum. Involvement of the gastrointestinal tract (stomach, colon, and small intestine), liver, and pancreas has been reported and the bladder and abdominal wall are involved sporadically. Schwannoma of the biliary tract can develop because there is an abundant network of autonomic nerve fibers to the wall of the gallbladder and bile duct.

The characteristic of a schwannoma (neurilemmoma) is the mixture of two growth patterns of alternating Antoni A and B areas. The type A area is cellular, composed of spindle cells, often arranged in a palisade fashion; the "nuclear-free zones" of processes that lie between the regions of nuclear palisading are termed Verocay bodies. The type B areas are far less orderly and less densely cellular with a loose meshwork of cells. A variety of degenerative changes may be found in schwannomas, including cyst formation, calcifications, hemorrhages and hyalinization.

Immunohistochemical methods are essential to differentiate Schwannomas from neurofibromas, gastrointestinal stromal tumors (GISTs), or even leiomyomas. Most Schwannomas are strongly positive for S100 protein. Gastrointestinal tract schwannomas are different from soft tissue schwannomas. They are not encapsulated and have an intramural growth pattern. In addition, they
are different both histologically and immunohistochemically from peripheral schwannoma. Malignant transformation is unusual.

Gastrointestinal schwannoma, are usually negative for CD34, CD117 and muscle cell markers, whereas they are strongly positive for vimentin and S100 protein. This typical combination differentiates schwannoma of the gastrointestinal tract from GIST. This differentiation is of clinical significance.

Gastrointestinal schwannoma are benign and then associated with a good prognosis and post-operative recurrence is unusual, even when treated only with enucleation. Gastrointestinal stromal tumors tend to recur and have malignant potential.(2,4,7)

The digestive tract schwannoma are generally asymptomatic and are often discovered incidentally. When symptomatic they compress adjacent structures. Pre-operative diagnosis of the tumor is very difficult because early clinical detection is limited until it gives a palpable mass or compresses the surrounding organs. Diagnosis often awaits intra-operative discovery and definitive histological confirmation.

Radiologically, the digestive tract schwannoma usually appear in CT as a homogenous, round or oval, well-defined mass with heterogeneous contrast enhancement and often show secondary degeneration such as a cystic change, cavity formation, necrosis, or calcification.(2,4)

The MRI findings of schwannomas are mainly masses with low signal intensity on the T1-weighted images and heterogeneous high signal intensity on the T2-weighted images owing to alternation of the Antoni A and B areas and secondary degeneration.(2,4)

In the present case CT illustrated a well defined hypodense cystic lesion. The MRI images revealed an abnormal cystic mass measuring 6 x 5 cm related to the middle part of the common bile duct with high signal intensity on T2 weighted images.

Surgical resection of these tumors remains the treatment of choice(1,2,4,7) and Schwannomas of the digestive tract have an excellent prognosis after a surgical resection. To date, there is no evidence that these tumors have a malignant potential.(2,4,7)

In summary, biliary schwannoma with choledochal cyst are extremely rare and a pre-operative diagnosis is often difficult. However, these potentially curable tumors should be included in the differential diagnosis of tumors arising in the biliary system.

References:


