Biliary Papillomatosis: case report

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Biliary papillomatosis is exceedingly rare. Only sporadic cases were reported since the first description of this disease by Caroli in 1959. The disease is characterized by multiple and recurrent papillary adenoma in the biliary tree. Jaundice and cholangitis are usually the main presenting symptoms. Although biliary papillomatosis is generally considered a benign disease, varying degree of dysplastic change in the epithelium is always present and progression to malignancy has been reported. The disease ultimately leads to death from sepsis or liver failure. In this paper, we present a new patient of biliary papillomatosis and describe the imaging findings including sonography, CT scan, MRI and ERCP.

Key words: Bile duct; Neoplasm; Papillary adenoma

CASE REPORT

An 76-year-old female patient presented in January 2000 with epigastric pain, poor appetite, and abdominal fullness for several days. Sonography performed in the emergency room showed gall bladder stone, CBD stone with biliary tree dilatation. Unusual thickening of the walls of common bile duct (CBD) and bilateral intrahepatic ducts (IHD) was noted at the same time.

On laboratory examination, the electrolytes were normal. The GOT was 279 U/L, GPT 497 U/L, alkaline phosphatase 570 U/L, total bilirubin 6.51mg/dl, and direct bilirubin 3.21 mg/dl. The white blood cell count was 7930 /ul, the hemoglobin was 14.5 g/dl, and the platelet count was 256000/ul. The alpha-fetoprotein was 1.7 IU/ML, and the CA-199 was 46.8 U/ml. She denied any history of previous hepatobiliary disease. She had no DM, hypertension or other systemic disease. She also had no smoking or alcohol drinking history. The review of system was unremarkable. The physical examination was generally unremarkable except mild tenderness in the epigastric area.

Under the impression of biliary tree stones with suspected cholangitis and tumor growth in the ductal wall, CT scan and MRI study were performed. CT scan showed distal end CBD stone and left side IHD stone with biliary tree dilatation. Diffuse wall thickening with soft tissue density in the common hepatic duct (CHD) and right side IHD was found (Fig 1). Cystic dilatation of left IHD was noted also. MRI study showed the similar findings to those of CT scan. The ductal wall lesions showed nearly isosignal intensity to liver parenchyma in T1WI (Fig 2) and mild high signal intensity in T2WI (Fig 3, 4). Gadolinium injection was not performed. MRCP study showed dilatation of the biliary tree with ductal wall thickening and irregularity (Fig 5). Under the impression of biliary tree stones with cholangitis, the patient received choledocholithotomy, cholecystectomy and T-tube placement. The operative findings showed dilated CBD and IHD with stones and diffuse villous tumor over CBD, CHD and IHDs. Biopsy of these villous tumor showed papillary adenoma with severe dysplasia. The patient had a smooth hospital course.
afer the operation and then discharged.

One and half years later, the patient came to our emergency room again due to fever and jaundice in recent one month. The laboratory examinations showed the WBC 9220/ul, total bilirubin 3.95 mg/dl, direct bilirubin 3.00 mg/dl, GOT 160 U/L and GPT 166 U/L. The CA-199 was 66.7 U/ml and CEA 1.0 ng/ml. CT scan showed dilatation of CBD and bilateral IHDS, but no obvious obstructing lesion or biliary tree stones could be found. Diffuse wall thickening in CBD, CHD and IHDS was still noted and there was no remarkable change when comparing with previous CT scan at the first time of admission. ERCP study showed dilatation of biliary tree with diffuse wall irregularity and shaggy appearance (Fig 6). Both of sono-guide liver biopsy of IHDS’s wall lesion and intraoperative biopsy of CBD’s wall lesion showed papillary adenoma. Microscopically, the papillary adenoma showed papillary configuration and cribriform pattern. Severe dysplasia of the epithelium could be found. The patient was admitted again 6 months later due to the same symptoms of fever and jaundice. Endoscopic biopsy of bile duct also showed papillary adenoma.

The final diagnosis of this patient was biliary papillomatosis with recurrent cholangitis.

Figure 1. a. Contrast-enhanced CT scan showed dilatation of CHD and IHDS. Diffuse wall thickening with soft tissue density in the CHD can be found (arrow). b. Dilatation of IHDS with abnormal wall thickening and soft tissue density over right IHDS can be noted (arrow).

Figure 2. a. MR study with axial T1WI showed dilatation of CHD with abnormal wall thickening which showed nearly isosignal intensity to normal liver parenchyma (arrow). b. Dilatation of right IHDS with abnormal wall thickening (arrow).
Biliary papillomatosis is a very rare disorder and only a small number of cases have been reported in the literatures. It was first described by Caroli and colleagues in 1959 [1]. The disease was characterized by multiple and recurrent papillary adenoma involving extensive area of the biliary tree. Extrahepatic bile ducts were involved in the majority of cases, but intrahepatic bile duct, cystic duct, gall bladder or pancreatic duct may also be involved.

The majority of patients presented between age of 50 and 60 years old. The men and women were equally affected [1]. The disease often presented with signs and symptoms of biliary obstruction that was often complicated with cholangitis. Death often ensued within 5-6 years from sepsis, liver failure or malignant transformation into invasive adenocarcinoma [4].

The disease was characterized by papillary proliferation of bile duct epithelium. It was considered as a benign entity, but varying degrees of dysplastic change in the epithelium could be found. It had the greater potential for malignant transformation than a solitary adenoma. Malignant transformation into adenocarcinoma in 35% of cases had been reported [4]. Histologically, there revealed biliary tree dilatation and multiple papillary adenomas. The epithelium of papillary adenoma was composed of mucin-secreting columnar or cuboidal cells with basal nuclei. A fibrovascular core supporting the epithelium could be found [1,4,5]. Grossly, multifocal papillary growing tumor in the extrahepatic bile duct with or without intrahepatic bile duct involvement could be found. The affected bile ducts were dilated and intraluminal mucin may be visible. Papillary adenomas were typically tan, soft and friable polyps, and usually did not demonstrate gross invasion of bile duct wall. Significant amount of mucin in the bile duct may have doughy or jelly-like consistency [1].

Most patients presented with intermittent obstructive jaundice and cholangitis. Two different features could be observed [3]. In most cases, the biliary obstruction was caused by the neoplasm. In the rare cases, biliary obstruction was not due to polypoid mass but due to a large amount of mucin within the

DISCUSSION

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lumen of bile ducts. The lesion may produce copious amounts of jelly-like mucus, filling and obstructing the biliary tree and leading to symptom.

Radiologic feature of papillary adenoma including sonography, ERCP, CT scan and MRI were reported in the literatures [1-7]. Sonography showed dilated intra- and extrahepatic bile ducts. Sometimes, single or multiple non-shadowing, intermediate echogenic intraluminal masses could be found. Low level echoes within lumen may represent mucin sludge or debris. Percutaneous cholangiography or ERCP study showed multiple, irregularly marginated, polypoid filling defects within dilated intra- and extrahepatic bile ducts. Irregular, granular or shaggy margin of bile duct's wall may represent small adenoma or inflammatory change from secondary cholangitis. Large floating filling defects corresponding to mucus secretion could be found. Lack of motility during irrigation helped to distinguish the papillary adenoma from intraductal stone or mucus plug. CT scan demonstrated intra- and extrhepatic duct dilatation with thickening of bile duct wall. Hypoattenuating intraductal soft tissue mass before and after contrast medium injection could be found. MRI findings were rarely reported in the literatures. The lesion was hypointense to liver parenchyma in T1WI, slightly hyperintense in T2WI and showed no significant enhancement on dynamic image after gadolinium injection [1].

The differential diagnosis of biliary papillomatosis may include polypoid adenocarcinoma, biliary metastasis, myoblastoma, lipoma, fibroma, cystadeno-noma, cystadenocarcinoma and carcinoid. But in fact, all the lesions mentioned above were rarely seen clinically. A well-defined lesion and absence of invasive feature could help to differentiate papillary adenoma from malignant lesion [3].

Surgery remains the primary treatment for biliary papillomatosis [1,2,4,5]. But complete surgical excision is difficult and local recurrence is common. Curettage and drainage of bile duct has been associated with high incidence of recurrence. Bile duct resection, pancreaticoduodenectomy and hepatic lobectomy can be performed when the involved area can be resected completely. However, even after complete resection with adequate clear margin, recurrence still can occur in the remaining intrahepatic duct. Frequent recurrence after surgery may suggest incomplete evaluation of the disease extent. Small papilloma may not be detected by conventional radiology and these undetected ones, usually remote from the main tumors, may be the foci of recurrence. So careful assessment of biliary tree by PTC is essential in determining the extent of surgery. Intraluminal brachytherapy may have some benefit for patients who are too frail for surgery [8]. The only definitive cure is total hepatectomy with liver transplantation. The prognosis is usually bad. In one report, the median survival was less than 18 months with a 5-year survival rate 12%[8]. Biliary sepsis is the most common cause of death.
REFERENCES

膽道多發性乳頭狀瘤：病例報告

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膽道多發性乳頭狀瘤相當少見，自從Caroll在1959年首次描述這個疾病以來，只有少數的病例被報導。這個疾病的特徵為在膽道中有許多乳頭狀腺瘤，並且在治療後容易復發，在臨床表現上主要為黃疸及膽管炎。雖然一般認為膽道多發性乳頭狀瘤為一良性疾病，但其上皮細胞常有不同程度的發育異常，甚至有報告顯示會演變成惡性腫瘤。這個疾病最終將造成病人死亡，而原因多為敗血症或肝臟功能衰竭。我們將報告一新病例，並呈現其在影像學上的變化。

關鍵詞：膽管，腫瘤，乳頭狀腺瘤