THAI J GASTROENTEROL 2005
Vol. 6 No. 1
Jan. - Apr. 2005

Intraductal Papillary Cholangiocarcinoma: Case Report and Review of the Literature

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ABSTRACT

A case of papillary cholangiocarcinoma is presented. A 64-year-old Thai female presented with progressive painless jaundice and significant weight loss within 3 months. CT scan showed dilated intrahepatic duct bilaterally and a soft tissue mass filled in dilated common bile duct. Endoscopic finding revealed bulging of the ampulla and a lobulated polypoid mass was extruded followed standard sphincterotomy. Cholangiogram showed irregular contour, intraluminal filling defect within common bile duct. 1 week after nasobiliary tube drainage, bile flow rate slowly decreased and bilirubin started to rise from obstructed tube. Surgical exploration revealed liver metastasis and hepaticojejunostomy was done. Finally, the patient was discharged home and remained free of jaundice 3 months after operation.

Key words : Papillary cholangiocarcinoma, Intraductal, Mucin hypersecreting

[Thai J Gastroenterol 2006; 6(1): 45-49]

INTRODUCTION

Papillary cholangiocarcinoma is the rarest type of cholangiocarcinoma[1]. There were few reports about its presentation and natural course. Prognosis is rather better than other types of cholangiocarcinoma according to previous literatures[2,3,9]. This is a case report of papillary cholangiocarcinoma with unresectable tumor mass since initial manifestration.

CASE REPORT

A 64-year-old Thai female presented with progressive, painless jaundice and 10 kgs weight loss within 3 months. She had no significant underlying disease. Physical examination revealed moderately icteric sclera with mild hepatomegaly. There was no liver stigmata found. Other systems was unremarkable. Liver function test (LFT) was abnormal (total bilirubin 28 mg/dL, direct bilirubin 23 mg/dL, aspartate aminotransferase 69 U/L, alanine aminotransferase 79 U/L, alkaline phosphate 293 U/L). CT scan showed dilated intrahepatic ducts (IHD) bilaterally and a soft tissue mass filled in dilated common bile duct (CBD) as shown in Figure 1A, 1B, 1C. Endoscopic finding during ERCP revealed bulging of the ampulla. Standard sphincterotomy was done and a lobulated polypoid mass was extruded through the opening of the distal CBD, as shown in Figure 2. Cholangiogram with vigorous injection of contrast into the CBD showed

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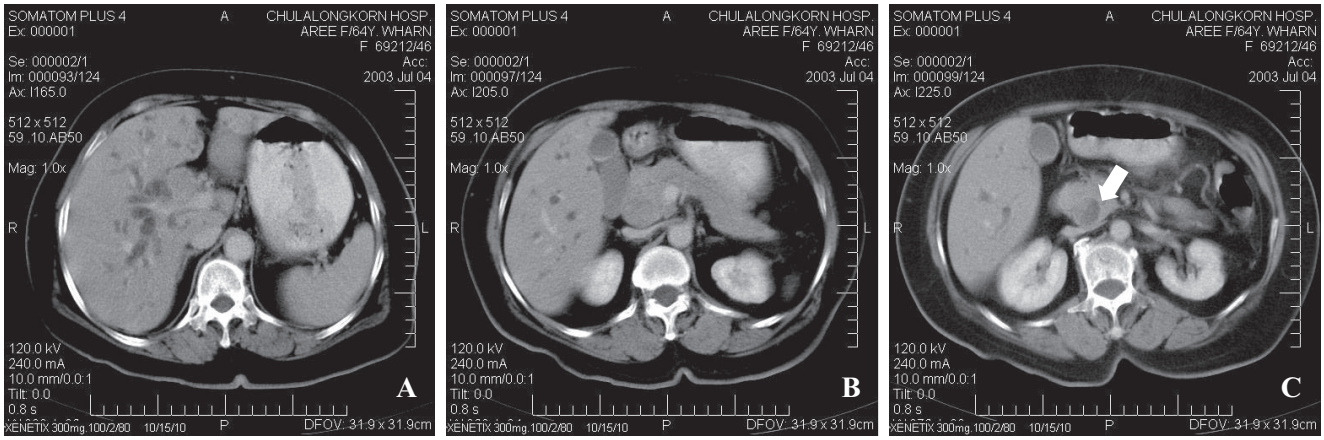

Figure 1 Contrast-enhanced CT images show diffuse intrahepatic bile duct dilatation (A, B) with evidence of a soft tissue mass within the dilated CBD (arrow, C)

irregular contour, intraluminal filling defect within CBD causing dilatation of the upstream intrahepatic bile ducts, as shown in Figure 3A-B. Biopsy of the polypoid mass was performed. Histopathology showed papillary structure, lining with high-grade, malignant epithelium, covering fibrovascular core, as shown in Figure 4. It was consistent with papillary cholangiocarcinoma. Soehendra dilator, number 8.5 Fr, was used to dilate the stricture. After then, a nasobiliary tube, number 8.5 Fr, was placed in order to drain bile proximal to the obstruction. Good bile flow via nasobiliary tube was observed, indicating proper positioning of the tube.

One week after nasobiliary tube was placed, bile flow rate slowly decreased and bilirubin started to rise. Therefore, percutaneous biliary drainage via left IHD was placed to assure adequate drainage. Follow up LFTs revealed a significant improvement of total bilirubin and finally her jaundice resolved. Subsequently, surgical drainage was performed due to recurrent blockage of the percutaneous tube. During surgical exploration, liver metastasis was detected. Hepaticojejunostomy was done and patient had a successful recovery. She was discharged home and remained free of jaundice 3 months after operation.

DISCUSSION

Cholangiocarcinoma (CCC) accounts for approximately 3 percent of all gastrointestinal malignancies, with a prevalence in autopsy studies of 0.01 to 0.46 percent[1]. The majority of cholangiocarcinomas (>90 percent) were adenocarcinomas. Squamous cell carcinoma was responsible for remaining of the cases. Adenocarcinomas are categorized into three types: nodular, sclerosing, and papillary. The rarest type is papillary cholangiocarcinoma and only small number of cases have been reported in the literature. Table 1 and 2 summarize clinical presentations, treatments and outcomes from previous publications in the English literatures[2-9].

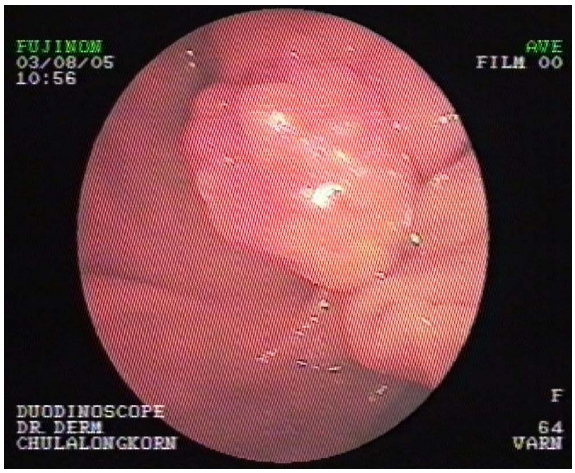

Figure 2 Endoscopic image shows a lobulated polypoid mass.

The largest series were from H.J. Kim, *et al*, which reported 9 cases of papillary tumors of bile duct[2]. In this series, 4 patients complained of biliary pain. Acute cholangitis and obstructive jaundice were observed in four other patients. In one patient , the lesion was incidentally found during a general check-up. ERCP revealed widely open ampulla of Vater with extrusion of mucin, and cholangiogram showed diffusely dilated intrahepatic and extrahepatic bile ducts with amorphous

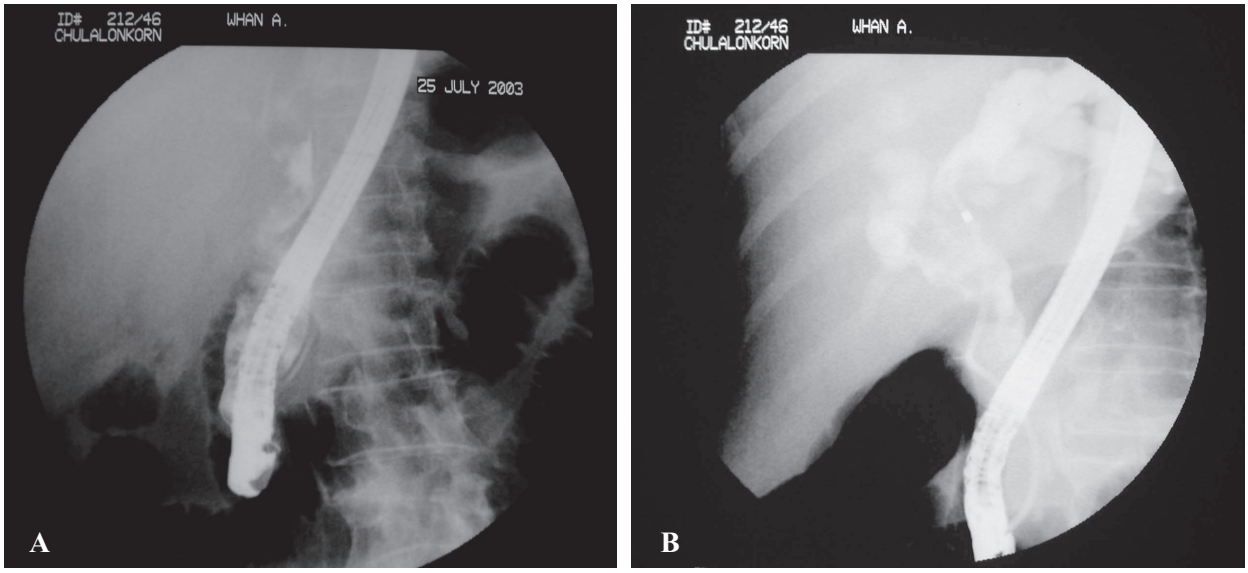

Figure 3 ERCP images show irregular contour filling defect within the dilated CBD.

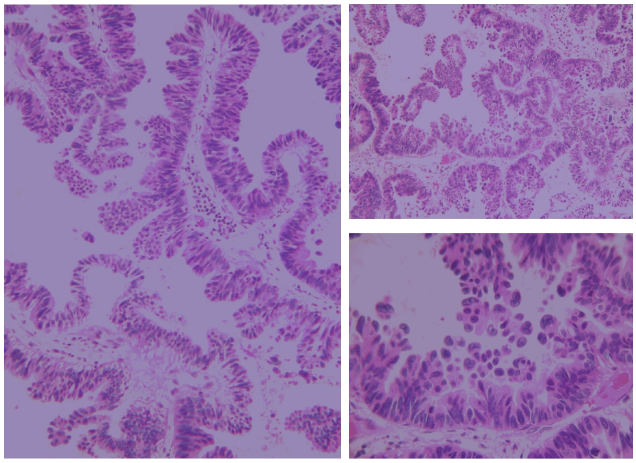

Figure 4 Histologic sections show papillary structure, lining with high-grade, malignant epithelium, covering fibrovascular core

filling defects. These findings were also noted in our case.

In Kim series, cholangioscopic examination revealed papillary mass or small mucosal lesion in the dilated bile duct containing thick viscus mucin[2]. All of his patients were referred for surgery. Curative resection was performed in eight of them. Exploratory laparotomy and biopsy were carried out in another patient with peritoneal seeding. Base upon histologic pathology, well differentiated adenocarcinoma in the background of benign hyperplasia and adenoma was documented in all patients except one, who showed pure adenoma. The authors concluded that this mucin-hypersecreting bile duct tumor can be characterized by a striking similarity to intraductal papillary mucinous tumor (IPMT) of the pancreas in clinical, radiologic, and pathologic features. In our patient, we demonstrated a similar histology with this series, unfortunately our patient had distant metastases and curative resection was not possible.

Two of nine patients from Kim's series showed intrahepatic cystic lesions on CT scan[2]. These cysts connected to peripheral dilated duct and became visible only after continuous forceful injection of contrast medium. These cystic lesions were very interesting manifestation. Martin, *et al* also reported 3 patients who presented with well-defined intrahepatic cystic masses, ranging in size from 7.2 to 21.1 cm[3]. The most prominent cells lining the epithelium were columnar with oncocytic features which showed abundant eosinophilic granular cytoplasm and centrally located nucleoli. Communication with ductal system was histologically confirmed in all patients. All three patients underwent resection and results of treatment were shown in Table 1.

Another unusual manifestation as an abdominal wall mass was reported by Ko, et al[8]. This mass was suspected to be abscess initially, but surgery revealed focal liver disruption with extrahepatic spread of biliary fluid to the abdominal wall via the ligamentum teres hepatis and to the lesser curvature of stomach via the gastrohepatic ligament. However, there was no fever or jaundice in this patient.

In conclusion, papillary cholangiocarcinoma is a rare type of cholangiocarcinoma and there are some

Table 1 Roengenographic findings, pathological findings and clinical outcomes of papillary tumors.

Ref.	Age	Sex	Cholangiogram from ERCP or CT	Pathology	Outcome
2	52	F	Diffuse CBD and IHD dilatation with amorphous filling defect	Intraductal CCC	Alive, 12 months
2	57	M	Diffuse CBD and IHD dilatation with amorphous filling defect	Intraductal CCC	Alive, 8 months
2	59	M	Diffuse CBD and IHD dilatation with amorphous filling defect	Invasive CCC infiltrating to hepatic parenchyma	Alive, 10 months
2	38	F	Diffuse CBD and IHD dilatation with amorphous filling defect Single cystic lesion communicating with large bile duct Dilatation of contiguous peripheral ducts distal to the lesion	Intraductal CCC	Alive, 28 months
2	66	F	Diffuse CBD and IHD dilatation with amorphous filling defect Cystic dilatation of LIHD and subtle dilatation of RIHD Dilatation of contiguous peripheral duct	Invasive CCC Peritoneal, carcinomatosis	Dead
2	42	F	CBD and IHD dilatation with amorphous filling defect	Intraductal CCC	Alive, 10 months
2	72	M	CBD and IHD dilatation with amorphous filling defect	Adenoma	Alive, 42 months
2	59	M	Diffuse CBD and IHD dilatation with amorphous filling defect	Invasive CCC infiltrating to hepatic parenchyma	Alive, 10 months
2	40	F	Diffuse CBD and IHD dilatation with amorphous filling defect	Invasive CCC limited to fibromuscular layer	Recurrence, 18 months
3	46	M	Unknown cholangiogram CT : cystic lesion centrally located in liver	Columnar cells with oncocytic features Communication with duct system	Alive, 36 months after resection
3	39	M	Unknown cholangiogram CT : lobulated enhancing mass within bile duct, biliary ductal dilatation and atrophy of segment 5 and 8.	Columnar cells with oncocytic features Communication with duct system	Alive, 24 months after right extended hepatectomy
3	50	M	Unknown cholangiogram CT showed large cystic mass with biliary ductal dilatation	Columnar cells with oncocytic features Communication with duct system	Alive, 18 months
4	58	M	Huge, multiloculated, septated cystic mass located in right lobe of liver from CT finding and connection to right IHD was demonstrated by intraoperative cholangiogram	Papillary adenocarcinoma of bile duct	Unknown
5	69	M	Laparoscopic ultrasonography showed mild right intrahepatic duct dilatation and exophytic mass measuring 1 cm at confluence.	A well-differentiated papillary adenocarcinoma of bile duct	Unknown
6	69	M	CT : Moderate dilatation of left IHD. ERCP : Obstruction in the proximal portion of left IHD.	Surgical specimen : papillary hyperplasia	Unknown
7	69	M	CT : Low density masses in hepatic ducts which extended to IHD. PTC : Tumor involved the junction of the anterior and posterior segmental bile duct	Papillary adenocarcinoma in hepatic ducts. (Exploration revealed swollen lymph nodes along common hepatic artery)	Died 10 months later because of pleural and perito neal dissemination
8	60	F	CT : Dilated CBD and left IHD filled with fluidlike content and multiple soft tissue density of papillary vegetation. ERCP : Depicted only part of distal CBD due to retrograde obstruction by the thick mucin.	Dilated IHD with papillary growth of mucin producing neoplastic cells arising from ductal epithelium	Unknown
9	40	M	CT : Cystic lesion with low attenuation near the porta hepatis. PTC : Cystic lesion filled with contrast material and having a slightly lobulated filling defect within it.	Tumor mass was located in the focal dilatation of IHD and communicated with the bile duct. Histology was consistent with well-differentiated papillary adenocarcinoma.	Alive with cancer free, 12 years follow up

Table 2 Presenting symptoms, endoscopic finding and unusual liver cysts in papillary tumors.

Ref.	No. of Patients	Presenting Symptoms (Number of Patients)	Diffuse IHD and CBD Dilatation with Amorphous Material	Widely open Ampulla of Vater with Extrusion of Mucoid Material	Cystic Lesion on CT scan
2	9	Cholangitis and jaundice(4) Biliary pain (4) Incidental finding (1) Recurrence cholangitis or biliary pain (4)	9	9	2
3	3	Recurrence pain (1) Spontaneously resolved jaundice (1) Abdominal mass (1)	Unknown	Unknown	3 (Size from 7.2-21.1 cm)
4	1	Fever RUQ pain Liver abscess like	0	Unknown	1
5	1	Painless jaundice	1 (Only right intrahepatic duct dilatation)	Unknown	0
6	1	Weight loss, abnormal radiography	1 (Only left IHD dilatation)	Unknown	0
7	1	Dyspepsia, abnormal radiography	1 (Only left IHD dilatation and intraductal masses)	1 (Soft, white masses with abundant mucus in IHD from surgical specimen)	0
8	1	Abdominal wall mass	1 (Dilated CBD and left IHD and intraductal papillary projection)	1 (Massive gelatinous mucin retention and papillary tumor out growths)	0
9	1	RUQ pain 1 Jaundice 1	0	Unknown	1
Total	**18**	**Jaundice or cholangitis** **7** **Biliary pain** **5** **Abscess like** **1** **Abdominal mass** **2** **Cystic lesion** **7** **Mucinous material** **11** **(Some patients had more than one presenting symptoms)**			

characteristic findings from imaging, cholangiogram, endoscopic and pathological findings which could be used for diagnoses. From previous case reports, they tentatively had a better prognosis compared to other types of cholangiocarcinoma. Unfortunately, our patient presented late and found to have liver metastasis, which precluded the patient from curative resection.

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