

A young lady with post-partum jaundice and right upper quadrant lump abdomen: an unusual etiology

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Abstract

Intraductal papillary mucinous neoplasm-biliary type is the biliary counterpart of intraductal papillary mucinous neoplasm-pancreatic type. We report a rare case of intraductal papillary mucinous tumor arising from extrahepatic biliary system. The diagnosis was established on histopathological analysis following endoscopic retrograde cholangiopancreatography-guided biopsy. Isolated papillary adenoma of the bile duct is extremely rare, and in this unusual case the patient was a 22-year-old young lady who had delivered a healthy infant 6 weeks previously.

Keywords IPMN-B, ERCP, CECT

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Introduction

Contrary to well-documented flat type neoplasms, biliary papillomatosis or intraductal papillary mucinous neoplasm-biliary type (IPMN-B) is relatively rare and a recently diagnosed disease entity. Papillary neoplasm of the bile ducts are rare, pathology is characterized by diffuse papillary proliferation of the bile duct epithelial cells. They account for 3-5% of cholangiocarcinomas and can arise from any portion of the intra or extra hepatic bile ducts [1]. Biliary intraductal neoplasms are proposed to be of two types: flat and papillary [2,3]. Papillary cholangiocarcinoma is believed to have a better clinical course than non-papillary cholangiocarcinoma, just as malignant intraductal papillary mucinous neoplasm of the pancreas (IPMN-P) has a better prognosis than pancreatic ductal adenocarcinoma.

Case report

A 22-year-old young lady was referred to the Gastroenterology department by her local physician with 4 weeks history of yellowish discoloration of urine and sclera. Lumpy upper abdomen and significant loss of weight (approx. 5 kg) were present for 3 weeks. There were no prodromal symptoms and

she denied any history of pruritus or passage of light colored stool. Her drug and family histories were unremarkable.

She had normal full-term pregnancy with transvaginal delivery of a healthy infant 6 weeks previously. There were no complications during pregnancy and no evidence of jaundice or cholestasis during pregnancy. Patient had a sensation of a lump in the right upper hypochondrium which had been gradually increasing in size over the past 3 weeks. It was associated with colicky pain lasting for 15-20 min and then subsided gradually. The patient also had intermittent spikes of fever with chills for one week.

On general examination there was pallor, jaundice but no lymphadenopathy. Tenderness was present in the right hypochondrium and a globular bulge was felt 4 cm below the right costal margin. Hepatomegaly was present with left lobe enlargement more than the right lobe; margins were sharp with firm consistency. Routine investigations confirmed normocytic normochromic anemia (hemoglobin 6.4 g/dL) with conjugated hyperbilirubinemia (10.5/6.8 mg/dL) and deranged liver function test (Table 1). Ultrasonography was done showing grossly dilated common bile duct with large filling defect throughout. Gross dilatation of intrahepatic biliary radicals and common hepatic ducts were seen.

Contrast-enhanced computed tomography (CECT) study of abdomen was done showing grossly dilated gallbladder with moderate to gross dilatation of intrahepatic biliary radicals and common bile duct (Fig. 1). A large polypoidal intraluminal multifocal enhancing lesion filling almost the entire common bile duct, cystic duct was also seen multifocally in the gallbladder. Multiple small hypodense lesions seen in both lobes showing peripheral rim enhancement clustered around central intra hepatic biliary radical dilatations were suggestive of cholangitic abscess.

Endoscopic retrograde cholangiopancreatography was performed under conscious sedation and after easy cannulation

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Table 1 Summary of investigations

Date	Hemoglobin (g/dL)	Total Leukocyte Count (cells/mm ³)	Differential Leukocyte Count	Bilirubin (Direct/ Indirect) (mg/dL)	Aspartate/ Alanine transaminase (IU/L)	Alkaline Phosphatase (IU/L)	CA19.9 (U/mL)
20.4.12	6.4	11700	N-84 L-12	10.5/6.8	95/70	680	
23.4.12	7.2	12900	N-87 L-5	12.7/10.2	117/112	659	
28.4.12							18

grossly dilated common bile duct was noted with obstruction and shouldering at mid common bile duct on air cholangiogram. A biopsy was taken from the mass lesion and sent for histopathological examination. Two plastic biliary stents 10 Fr x 10 cm were placed across the mass. Choledochoscopic image was taken by ultrathin neonatal endoscope (Fig. 2).

Histopathology analysis of the soft tissue revealed multiple papillae lined by complex epithelial proliferation with severe dysplastic changes. Early invasion of stroma was seen with neutrophilic infiltration and dilatation of blood vessels (Fig. 3A, B). These findings were in consistence with intraductal papilloma with severe dysplasia.

Discussion

Two types of biliary intraductal neoplasms preceding invasive cholangiocarcinoma have been identified so far; a flat type neoplastic lesion called BilIN, which develops into non-papillary cholangiocarcinoma, and a papillary type called

IPMN-B with malignancy potential. IPMN-B comprises a histological spectrum that ranges from benign to malignant adenoma, borderline tumor, carcinoma *in situ*, and invasive carcinoma [4]. The current WHO classification and some authors recognize biliary papillomatosis, as well as BilIN, or biliary epithelial dysplasia, as precursor lesion of cholangiocarcinoma [5-8]. The pathogenesis of the disease is not yet known, but has been thought to be related to chronic biliary ductal inflammation from pancreatic juice reflux resulting in excessive proliferation of the bile duct epithelium followed by dysplasia-carcinoma sequence [7].

Unlike traditional ductal adenocarcinoma, papillary tumors are low-grade malignancy. They are usually limited to the mucosa or spread along a mucosal membrane, although they can invade the ductal wall in the later stage. Once papillary cholangiocarcinoma shows stromal invasiveness, its prognosis is as poor as non-papillary cholangiocarcinoma. Most papillary tumors are peripheral in location and are nodular or papillary in shape. IPMN-B involves the extrahepatic ducts in 58%, both extra and intrahepatic ducts in 33% and intrahepatic ducts alone in 9% cases [9].

IPMNs are classified in 4 subtypes on the basis of histology and mucin expression, as recently published in a consensus study [10]. These are: 1) the gastric type, composed of columnar epithelial cells resembling cells of the gastric foveolae (MUC1-, MUC2-, MUC5+); 2) the intestinal type, characterized by

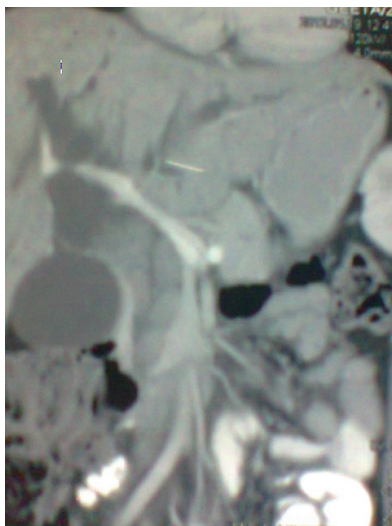


Figure 1 Contrast-enhanced computed tomography showing large polypoidal intraluminal multifocal enhancing lesion filling almost the entire common bile duct and cystic duct (white arrow)

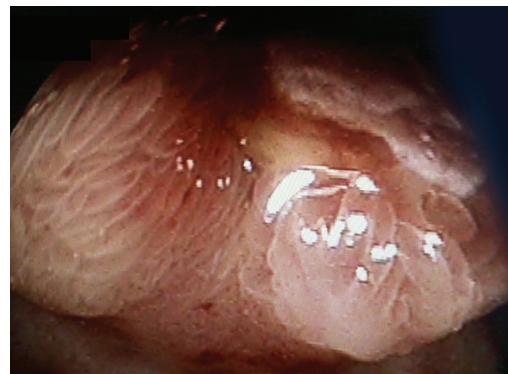


Figure 2 Choledochoscopy showing papillary soft tissue lesion projecting in the common bile duct

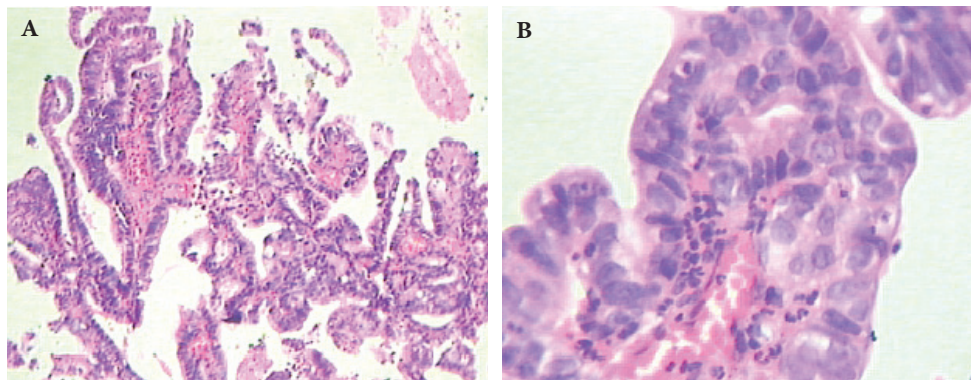


Figure 3 (A, B) Histopathological slides showing multiple papillae lined by epithelial proliferation with severe dysplastic changes

tall columnar cells and some goblet cells as found in intestinal adenoma or adenocarcinoma (MUC1-, MUC2+, MUC5+); 3) the pancreaticobiliary type, composed of columnar cells with moderate amphophilic cytoplasm and enlarged hyperchromatic nuclei resembling biliary or pancreatic epithelium (MUC1+, MUC2-, MUC5+); and 4) the oncocytic type, characterized by cells with abundant eosinophilic cytoplasm and large round nuclei (MUC1-/+ , MUC2-, MUC5+).

Biliary papillomatosis commonly affects adults older than 60 years, with a Male: Female ratio of 2:1 [11], but we report a case of IPMN-B in a young 22-year-old lady during the post-partum period. There is no literature available about a possible link between pregnancy hormones and IPMN-B. Cytological examination by epithelial brushing and biopsies during ERCP is an effective preoperative procedure for the diagnosis of papillary tumor of the bile duct.

In conclusion, intraductal papillary tumor of the bile duct runs a benign course and will benefit from early surgical intervention. When massive localised dilatation of the intrahepatic duct is seen on CT without an obvious cause, papillary mucinous tumor of the bile duct should be considered. ERCP is often diagnostic when a copious amount of mucus is present. Furthermore, for establishing the concept of IPMN-B, more continual reports and studies are warranted.

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