CASE REPORT

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A Case Report of Intraductal Papillary Neoplasm Associated with an Extrahepatic Bile Ducts

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ABSTRACT Choledochal cysts are congenital cystic dilations of the extrahepatic and/or intrahepatic bile ducts and are rare. Adults with choledochal cysts are at risk of developing biliary carcinoma. Our case is a 31-year-old female patient who presented with right upper quadrant pain. In magnetic resonance imaging/magnetic resonance cholangiopancreatography, there was a fusiform enlargement of the extrahepatic bile duct and cystic duct and it was evaluated as compatible with Type 1 choledochal cyst. Extrahepatic biliary tract excision-cholecystectomy-Roux En Y hepaticojejunostomy procedure was performed. The pathology result was intraductal papillary neoplasm (IPN). IPN is a rare entity in the biliary tract and because it is precancerous, early diagnosis and surgical excision are critical.

Keywords: Choledochal cyst; bile ducts extrahepatic; bile duct neoplasm

Choledochal cysts are congenital cystic dilations that can be seen in the intra/extra hepatic biliary system. Choledochal cysts are considered as premalignant lesions and the mechanism of carcinogenesis has not been fully elucidated yet.[1] The risk of malignancy in choledochal cysts determines the treatment protocol. Excision of all affected bile ducts, cholecystectomy, and bilioenteric anastomoses form the basis of the treatment protocol and are also applied prophylactically in asymptomatic patients.[2] Intraductal papillary neoplasm (IPN) of the biliary tract describes neoplasia characterized by papillary proliferation of dilated biliary epithelium. IPN is a precursor lesion for cholangiocarcinoma.[3-5] In this case, the clinical, radiological and pathological features and surgical treatment of a 31-year-old female patient with Todani Type 1 choledochal cyst and diagnosed as IPN after surgical resection are presented.

CASE REPORT

A 31-year-old female patient was admitted to our clinic with right upper quadrant pain. There was no significant feature in the physical examination findings of the patient, who had no history of surgery other than ovarian cyst surgery, no comorbidities, smoking and alcohol use. Laboratory values were 4.180/qL white blood cells, 4.2 g/L albumin, 0.2 mg/dL total bilirubin, <0.1 mg/dL direct bilirubin, 57 IU/L aspartate transaminase, 82 IU/L alanine transaminase, 110 IU/L gamma-glutamyl transferase and 91 IU/L alkaline phosphatase. In magnetic resonance imaging/magnetic resonance cholangiopancreatography (MRI/MRCP), there was a fusiform enlargement of the extrahepatic bile duct and cystic duct and it was evaluated as compatible with Type 1 choledochal cyst (Figure 1, Figure 2). Because choledochal cysts are precancerous lesions, total extrahepatic bile duct excision was planned for the patient. Intra operative ascites, metastases, and peritoneal spread were not observed. The patient underwent total extrahepatic bile duct excision-cholecystectomy-Roux En Y Hepaticojejunostomy at the bifurcation level. As a result of the postoperative pathology, intracholecystic papillary neoplasm was

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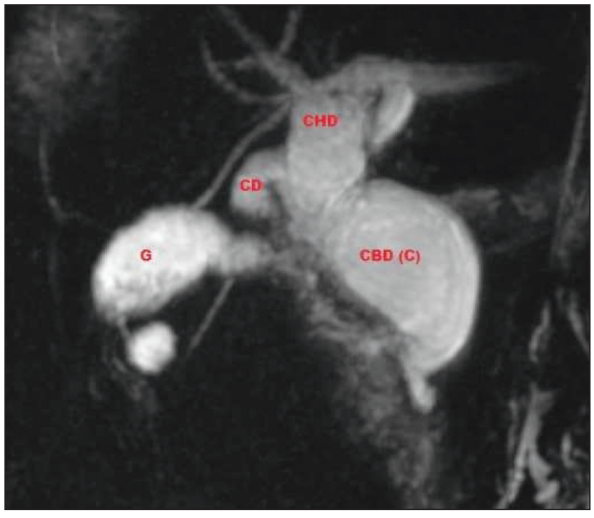

FIGURE 1: Gallbladder (G), cystic duct (CD), common hepatic duct (CHD), common bile duct (CBD) or choledoch (C) in the coronal magnetic resonance cholangiopancreatography image; Fusiform cystic enlargement associated with the cystic duct and extrahepatic bile duct.

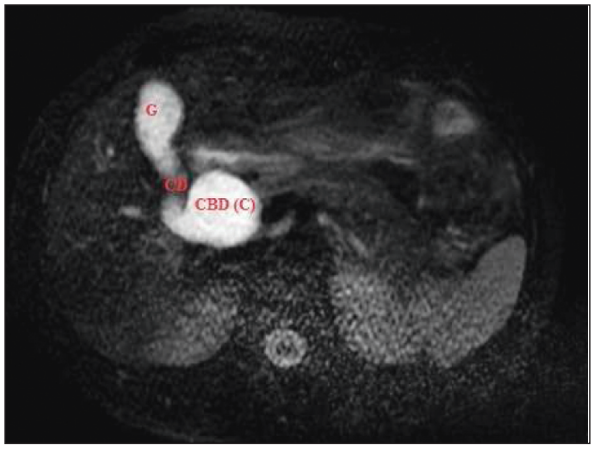

FIGURE 2: Gallbladder (G), cysticduct (CD) and choledoch (C) in the axial magnetic resonance imaging image.

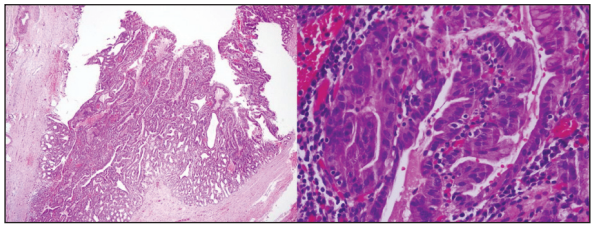

FIGURE 3: In the choledoch pathology specimen stained with hematoxylin&eosin, a neoplastic formation forming papillary structures growing towards the enlarged cystic lumen at low magnification (x40) drawsattention. At high magnification (x400), columnar-type cells with eosinophilic cytoplasm located around the fibrovascular core and without significant atypia and loss of polarity are observed.

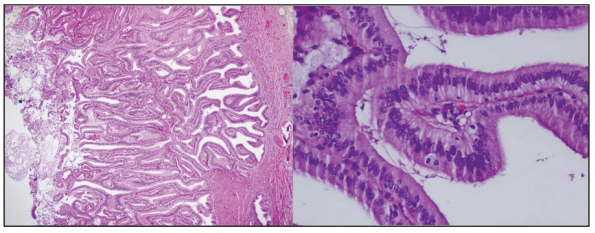

FIGURE 4: In the gallbladder pathology specimen stained with hematoxylin&eosin, epithelial proliferation that forms papillary structures that grow towards the lumen of the gallbladder is remarkable (x100). At larger magnification (x400), this proliferation is observed to consist of true papillae with fibrovascular cores. Columnar cells with intracytoplasmic vacuoles around these cores and cilia-like structures are observed on the apical surfaces of these cells.

detected in the gallbladder and no invasive carcinoma foci was observed. There was no papillary neoplasm at the surgical margin of the cystic duct. IPN was detected in the extrahepatic bile duct excision material and there was no invasive carcinoma foci. No papillary neoplasm was observed in the proximal and distal surgical margins. As a result of intraoperative and final pathology, it was observed that there were no affected regional lymph nodes (Figure 3, Figure 4). The patient had no post operative complications and was discharged on the 7th day.

Informed consent was obtained from our patient and she signed permission to publish her story.

DISCUSSION

Although bile duct cysts are in a rare condition, they are important because of their malignant potential. The incidence of bile duct cancer in patients with choledochal cysts increases with age. It can be seen in 15% of patient solder than 20 years and 35% of patient solder than 60 years of age. Approximately 70% of tumors are seen in the biliary tract and 30% in the gallbladder.[6]

Bile duct cysts are divided into types according to the Todani anatomical classification. Type 1 is the most common type with a frequency of 50-90%, and Type 4 is the second most common type with a frequency of 15-42%. Dilation of Type 1 extrahepatic bile ducts (1a-Cystic, 1b-Focal/segmental, 1c-Fusiform), Type 2 choledochal diverticulum, Type 3 choledococele, Type 4 choledochal cyst with multiple dilation of intra-extrahepatic bile ducts (4a-dilation

in intra and extrahepatic bile ducts, 4b-dilation only in extrahepatic bile ducts), Type 5 is characterized by Caroli's disease, which is characterized by multiple cysts in the intrahepatic biliary tract (Figure 5).[2] In our case, fusiform cystic enlargement associated with the cystic duct and extrahepatic bile duct and Todani Type 1 choledochal cyst was the most common. It is accepted that long-term reflux may lead to malignant degeneration of the biliary epithelium in Todani Type 1 and Type 4 choledochal cysts, in which pancreatic secretions reflux into the biliary tract for a long time.[7]

Biliary IPN is characterized by dilated bile ducts filled with papillary or villous neoplastic biliary epithelium.[8,9] Risk factors also choledochal cyst, primary sclerosing cholangitis, and Gardner's syndrome. It is more common in the 6th decade of life and in men. Although it can be seen in all bile ducts, it is most commonly seen in the intrahepatic localization or in the hepatic hilum.[3,4,8] Although they are generally non invasive lesions, they may have an invasive component and may accompany adenocarcinoma.[10] In our case, the patient with choledochal cyst as a risk factor had intraductal and intracholecystic papillary neoplasm without an invasive component.

Bile duct IPN is clinically similar to other biliary tract pathologies. 35-88% of the cases present with right upper quadrant pain, 5-59% with recurrent acute cholangitis and 20-36% with obstructive jaundice. 5% of cases may be asymptomatic.[5,11] Our case also had abdominal right upper quadrant pain.

Radiological findings are characterized by in bile duct cysts and IPN enlarged bile ducts. Ultrasonography, MRI/MRCP, endoscopic retrograde cholangiography and percutaneous transhepatic cholangiography are used in bile duct imaging.[12,13]

The differential diagnosis of choledochal cysts includes duodenal diverticulum, pancreatic cystic lesions and cholangiocarcinoma, and differential diagnosis can be made with imaging methods. The differential diagnosis of bile duct IPN includes hepatic mucinous cystic neoplasm and cholangiocarcinoma.[14]

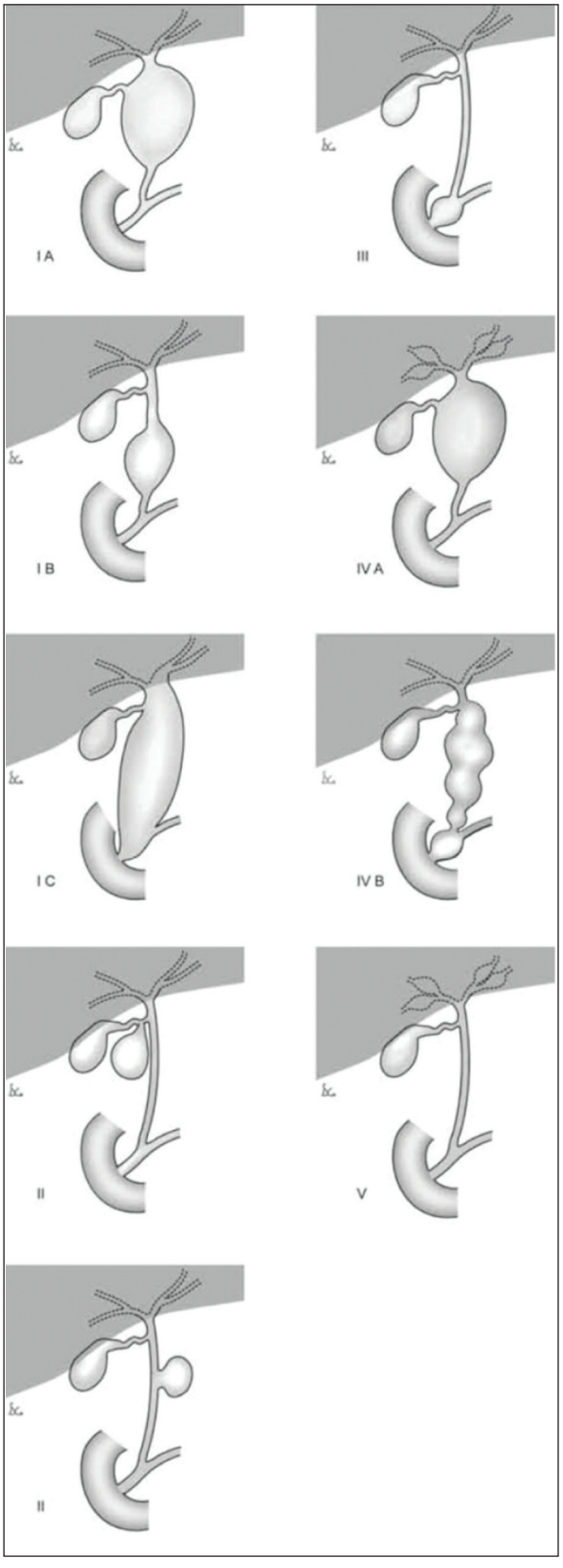

FIGURE 5: Modified Todani classification.[2]

The standard treatment for bile duct cysts is complete resection of the cyst. The risk of complications increases in the presence of pancreatitis, cholangitis, bile duct stones and malignancy. In Type 1 bile duct cyst, complete cyst resection with extrahepatic bile duct resection-cholecystectomy-Roux En Y biliojejunal anastomosis are the basic surgical principles. Simple excision is usually sufficient because the risk of malignancy is less in Type 2 common bile duct diverticulum. Endoscopic sphincterotomy is generally used for Type 3 choledochocel, especially in cases with a cyst diameter of less than 2 cm. Surgical resection may be more aggressive in cases with Type 1c, 4a and 5 bile duct cysts, liver resection or transplantation may be added to complete cyst excision due to intrahepatic spread. In the presence of malignancy, oncological surgical approach is applied. Cholecystectomy is included in the surgical procedure because the risk of gallbladder cancer increases in surgical resection of all types.[6]

In the follow-up of these patients abdominal ultrasound and parameters of cholestasis, cancer antigen 19-9 levels is recommended on annual basis.[15] We planned the postoperative follow-up with annual blood test and abdominal ultrasound, MRI/MRCP in a suspicious finding.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Sezai Tarhan, Enes Şahin;* ***Design:*** *Sezai Tarhan, Enes Şahin;* ***Control/Supervision:*** *Enes Şahin, İsmail Erdem Okay;* ***Data Collection and/or Processing:*** *Sezai Tarhan, Enes Şahin, Ahmet Tuğrul Eruyar;* ***Analysis and/or Interpretation:*** *Sezai Tarhan, Enes Şahin, İsmail Erdem Okay;* ***Literature Review:*** *Sezai Tarhan;* ***Writing the Article:*** *Sezai Tarhan;* ***Critical Review:*** *Enes Şahin, İsmail Erdem Okay;* ***Materials:*** *Sezai Tarhan, Enes Şahin;* ***Other:*** *Sezai Tarhan, Enes Şahin.*

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