Intraductal papillary mucinous neoplasm of the intrahepatic bile ducts: a case report and literature review

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Case report

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Abstract

Background: Intraductal papillary mucinous neoplasm of the bile ducts (IPMN-B) is a rare malignant tumor originated from the epithelium of the bile duct. It can secrete a large amount of mucin to cause biliary obstruction. This disease has just been recognized in recent years.

Case presentation: We found a 60-year-old woman with intermittent right upper abdominal pain. Imaging examination showed that the left intrahepatic bile duct was dilated with a solid mass. We had left hepatectomy. During the operation, there was colloidal mucus in the bile duct. There was a papillary mass on the wall of the bile duct. -to have developed into invasive adenocarcinoma. At a postoperative 6 months follow-up, the patient had no recurrence and is in good conditions.

Conclusion: In our case, IPMN-B has underwent a malignant transformation at the early stage. Therefore, we think that surgical resection should be done as early as possible after the diagnosis of IPMN-B, so as to get a favorable prognosis.

Background

Intraductal papillary tumors of the bile ducts (IPNB) is a kind of precancerous pathological state of cholangiocarcinoma which has been gradually recognized in recent years. In 2010, it was defined by the World Health Organization (WHO) as an independent biliary tumor disease [1]. The disease can occur in both intrahepatic and extrahepatic bile ducts, which are connected with bile ducts. About 1/3 of IPNB can secrete mucus, which is called intraductal papillary mucinous tumor of the bile ducts (IPMN-B) [2, 3]. At present, the description of IPMN-B is rarely reported. Because of the clinical symptoms and signs are not obvious and the imaging performance is not typical, it often leads to missed diagnosis and misdiagnosis of IPMN-B. One case of intrahepatic cholangiocarcinoma was found due to abdominal pain. The postoperative pathological diagnosis was intrahepatic IPMN-B infiltrating adenocarcinoma. The diagnosis and treatment process of the patient was reviewed, and the current research progress of the disease was discussed in combination with relevant literature.

Case Presentation

A 69-year-old woman presented at the outpatient clinic for intermittent pain in her right upper abdomen. She was healthy without smoking or drinking. An abdominal ultrasonography showed a mixture of cystic and solid mass in the dilated left hepatic duct, and there are many gallstones in the gallbladder. The liver function and tumor marker tests were normal: alanine Transaminase, 17.4 IU/L; aspartate transaminase, 20.2 IU/L; and serum bilirubin, 20.3 mg/dL; γ-glutamyl transpeptidase, 39.2 IU/L; alkaline phosphatase, 88 IU/L. alfa-fetoprotein, 1.27 IU/ml; carcinoembryonic antigen 19 – 9, 9.14 IU/ml; and carcinoembryonic antigen, 2.47 IU/ml.

Contrast-enhanced computed tomography revealed a tumor in the dilated left intrahepatic bile duct. The lesions in the hepatic artery phase and portal vein phase are all moderately enhanced. The hepatic artery

phase lesions are enhanced more than the liver parenchyma, showing a slightly higher density lesions; the portal vein phase lesions are inferior to the liver parenchyma, showing a relatively low density lesion. Magnetic resonance imaging and magnetic resonance cholangiopancreatography showed significant dilation of the peripheral biliary ducts in the left lobe of the liver and an 1.9 × 1.5 cm tumor in ducts. The right intrahepatic bile duct and extrahepatic bile duct are normal (Fig. 1).

On intraoperative inspection, we found that the thick jelly-like substance was flowing out of the intrahepatic bile duct, and a mass of about 2 × 2 cm in size was seen in the left hepatic duct, which was papillary and had a soft pedicle. Because of it was suspected malignancy, we made a decision to perform a left hepatectomy. Pathology reports from the postoperative specimens were intraductal papillary neoplasm with high-grade intraepithelial neoplasia, part of invasive mucinous adenocarcinoma. Immunohistochemistry showed that CK19 was strongly positive in tumor tissue. Other positive markers are CK8/18, CK19, CAM5.2 and CK, partially positive for the CEA. The proliferative index based on Ki-67 staining was 30%. The tumor cells were negative for CK7, CDX-2, CK20, AFP, GCDFP-15, CA125 and P53. The patient had an uneventful recovery and was discharged from the hospital 14 days later. No recurrence was found after 6 months of follow-up (Fig. 2).

Discussion And Conclusion

The definition of intraductal papillary mucinous neoplasm of the bile ducts (IPMN-B) is derived from intraductal papillary mucinous neoplasm of the panceas (IPMN-P). Both the bile duct and the pancreatic duct originated from the foregut mesoderm in embryology, which made IPMN-B and IPMN-P have many similar characteristics. They can both secrete mucus and cause corresponding blockages in the ducts. Most importantly, they are precancerous lesions [3]. In recent years, IPMN-P has been gradually recognized, but the understanding of IPMN-B is just the beginning.

IPMN-B is rare in western countries, mainly in China, Japan, South Korea and other Asian countries, there is no big data to provide the incidence in the population. The age of onset of this disease is 55–65 years, and there is no significant difference in incidence between men and women [4, 5].The pathogenesis of the disease is not clear. But some scholars think that chronic inflammation of stones, clonorchis sinensis and chronic inflammation of bile duct may increase the risk of the disease [6, 7]. The clinical manifestations of B are atypical and non-specific. In the early stage, it only showed intermittent upper abdominal pain. As the disease progresses, the tumors increase and mucus secretion can cause fever, jaundice, and even secondary pancreatitis. In addition, about 10% of patients are asymptomatic [8, 9].

As far as the current understanding of IPMN-B is concerned, it is still difficult to make a clear diagnosis before surgery. In laboratory tests, it is often found that abnormal liver function indexes and elevated CA-199 are caused by obstruction of the bile duct. Among them, the increase of CA-199 could not identify its benign and malignan [10]. Imaging tests also lack specificity for the diagnosis of IPMN-B. Ultrasound is the most economical inspection method. It can show bile ducts that are dilated inside and outside the liver, and uneven echo masses in the bile ducts. However, misdiagnosis is often caused by intestinal

flatulence and bile duct stones. The most commonly used diagnostic test for IPMN-B is computed tomography (CT), which is typically manifested as local or extensive bile duct dilatation, and the mass in the bile duct can be seen. The computed tomography (CT) arterial phase shows a slight intensification of the mass in the parenchyma, and no significant enhancement in the delay period. Magnetic resonance imaging (MRI) and magnetic resonance cholangiopancre- atography (MRCP) can clearly show the bile duct tree, and can find filling defects in the bile duct. Ritchie et al. have collected literature and concluded that typical imaging features of IPMN-B include bile duct dilatation and bile duct wall mass [11]. Positron emission tomography-computed tomography (PET-CT) is currently the most accurate method for diagnosing benign and malignant tumors and determining whether they are metastatic. Ikeno et al. comparative study found that the preoperative PET-CT can differentiate non-invasive IPMN-B from invasive IPMN-B [12]. Eendoscopic retrograde cholangiopancreatography (ERCP) can detect mucus adherence and outflow at the duodenal papilla opening in some patients, and biliary filling defects can be found by angiography. Biopsy can be taken in some patients with choledochoscope, which is a clear diagnosis method, but because of its technical difficulty large and easy to induce iatrogenic pancreatitis, and less clinical application. According to its pathological features, IPMN-B can be divided into four types: pancreaticobiliary duct type, gastric type, intestinal type, and eosinophilic type [13]. According to the degree of atypia, it can be divided into: low or medium grade intraepithelial tumors, high grade intraepithelial tumors, related invasive cancers [14]. Therefore, IPMN-B is considered to be an important precursors for the development of cholangiocarcinoma. The rate of malignancy has not been accurately reported, Kubota et al. found that 119 patients with IPMN-B diagnosed with invasive cancer developed pathologically after surgery, accounting for about 36% [8]. Therefore, if the disease is clinically suspected, it should be actively seek treatment. IPMN-B needs to be distinguished from other types of cholangiocarcinoma, bile duct stones, and other cystic diseases, especially mucinous cystic tumors (MCN).

Radical surgical resection remains the preferred treatment for IPMN-B. The extent of resection depends on the location and degree of development of the lesion. For lesions that are localized in the intrahepatic bile duct and have negative frozen resection margins during surgery, hepatic segment or lobe resection should be performed. In patients with tumors involving the extrahepatic bile duct, pancreatoduodenectomy should be feasible. For patients with extensive bile duct invasion by B, liver transplantation is the only treatment [9, 15]. Lymph node metastasis of IPMN-B is rare, so it is still controversial whether to perform regional lymph node dissection. However, for tumors in the hilar and lower bile ducts, Jarnagin et al. recommend regional lymph node dissection [15]. For patients who cannot undergo radical surgical resection, biliary drainage or stent placement in the bile duct should be performed to control biliary infections, improve liver function, and reduce the incidence of complications.Studies show aggressive surgical treatment,the 1-, 5- and 10-year survival rates for patient with M-IPNB were 96%, 84% and 81% [8].

In conclusion, as a rare biliary tract tumor, IPMN-B is still unknown. For elderly patients, imaging studies suggest that patients with bile duct wall masses and bile duct dilatations need to be alert to the

possibility of this disease. For early radical resection of this disease, good prognosis can be obtained by removing biliary obstruction.

Declarations

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Not applicable.

Authors' contributions

J.F., L.-Q.Z., Y.-L.W., Y.L., X.-L.M., G.-F.C., B.R., G.-B.J. and Y.-D.C. participated in the clinical management of the patient; G.-B.J. and Y.-D.C. performed the surgeries described in this report; Y.Z. and X.-J.G. conducted histological diagnoses; and F.J. and Y.Z. wrote the manuscript. All authors have read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed are included in this published article.

Ethics approval and consent to participate

Not applicable.

Consent for publication

The patient provided written consent to publish this case report.

Competing interests

The authors declare that they have no competing interests.

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Figures

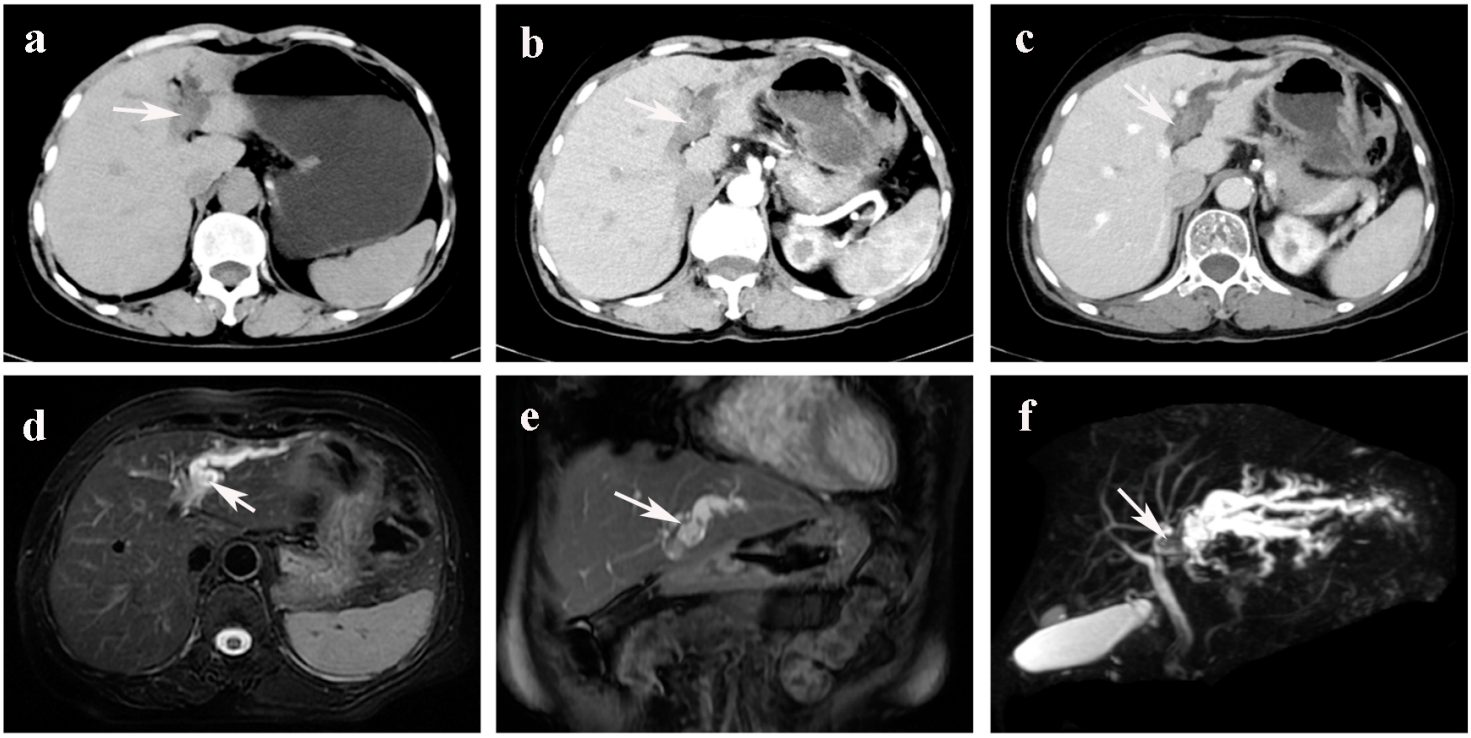

Figure 1

Preoperative CT scan: a Dilation of the left hepatic duct can be seen, as seen on plain CT. b, c The enhancement of intraductal mass can be seen in the contrast-enhanced image (white arrowhead). Magnetic resonance imaging: d, e Axial and coronary MRI image showed a cystic-solid mass in the left hepatic duct (white arrowhead). f On MRCP, a mass blocked the left hepatic duct (white arrowhead).

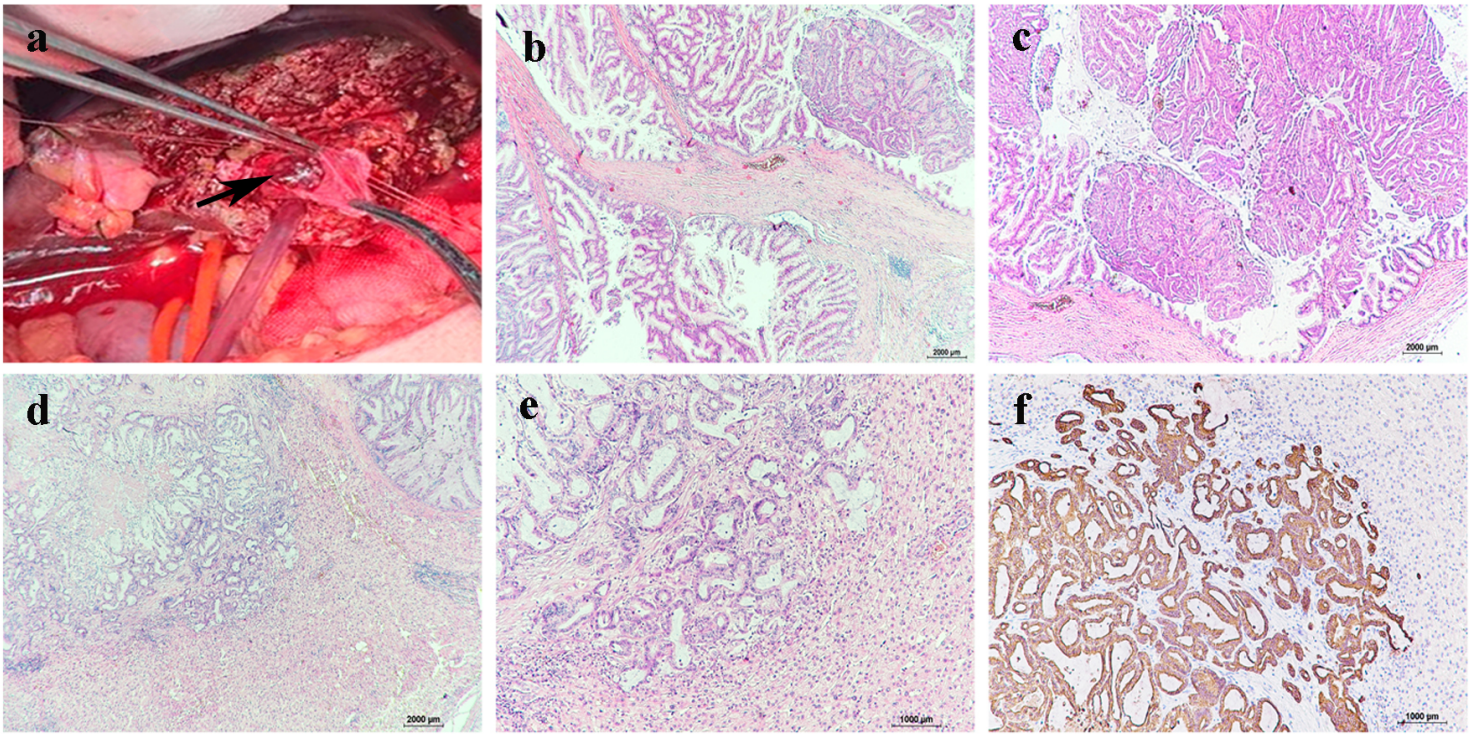

Figure 2

Surgery photos: a There was a papillary mass (approximately 2 × 2 cm) in the left hepatic duct (black arrowhead). Pathological findings. b, c In the duct, the tumors are arranged in papillary shape, and the fibrous vascular axis can be seen in the center (HE, a,b × 40). d, e The tumor presented adenoid structure, partially fused and infiltrated the surrounding liver tissue (HE, d × 40, e × 100). f Immunohistochemistry showed that CK19 was strongly positive in tumor tissue (f × 100).