

Case Report

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Intrabiliary papillary myxoma: A case report

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Abstract

In 2010, the World Health Organization (WHO) classified intra-ductal papillary mucinous tumor (ipmn-b) as a solid tumor of the liver producing mucinous bile, which is rare and difficult to diagnose. We reported a 71 year old female patient with ipmn-b due to abdominal pain. Through the description of the causes, diagnosis, treatment and prognosis, we can get a better understanding of the disease. Ipmn-b is considered as a kind of benign precancerous lesion of cholangiocarcinoma with good prognosis.

Keywords: papillary mucinous tumor; cholangiocarcinoma; neoplasm.

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Introduction

Intraductal papillary mucinous neoplasm of the bile duct (ipmn-b) is a kind of intraductal papillary neoplasm of the bile duct (ipnb). It is a rare benign lesion with high malignant transformation rate. It is characterized by intraluminal papillary mass with bile duct obstruction and dilatation. Some of these low-grade malignant tumors secrete large amounts of mucin which can change the normal flow of bile and lead to the expansion and spread of intrahepatic and extrahepatic bile ducts. Here we are reporting a case of ipmn-b in intrahepatic bile duct which was recently treated in Chengdu Second People's hospital.

Case report

A 71 years old female patient was admitted with "upper abdominal pain for 5 days". 10 years before admission, he underwent partial resection of the left lateral hepatic lobe, cholecystectomy, common bile duct exploration and T tube drainage for

intrahepatic bile duct stones and gallbladder stones Surgery. 5 years before admission, due to common bile duct stones, abdominal cavity exploration, common bile duct exploration and T tube drainage was performed. The patient had repeated upper abdominal pain for 5 days, accompanied by chills and high fever, with a maximum body temperature of 39.0 degrees Celsius, accompanied by nausea and vomiting. An emergency CT scan in our hospital showed that the intrahepatic bile ducts were dilated and the left bile duct and common bile duct were significantly dilated with punctate calculi. She was admitted to our department for further treatment. Physical examination on admission shows her general condition was good, the skin and sclera are mild yellowish, and multiple scars from previous surgery are seen. Her other examination shows tumor indicators: CA19-9, CEA and AFP had no obvious abnormalities, her blood and biochemical examination after admission is presented in Table 1. Her abdominal ultrasound shows intra and extra hepatic bile ducts and main pancreatic duct was dilated. There

was a sedimentary stone in the lower part of the common bile duct. Her MRI+MRCP shows intrahepatic and extrahepatic bile ducts were dilated (the common bile duct of the left lobe of the hepatic bile duct was significant with a transverse diameter of about 2.4 cm; the signal in the lumen of the lower part of the common bile duct is locally reduced, which was a silt-like stone. ERCP subsequently shows the patient underwent open lower left third lobe hepatectomy, hepatobiliary lesion and T-tube drainage. During the operation, obvious expansion of common bile duct >3 cm. After puncture, golden jelly like liquid was extracted, and the upper segment was longitudinally cut for 1 cm. After choledochoscopy the opening of the large duodenal papilla was fishmouth like, and the left hepatic duct was not developed, and the common bile duct could not be filled. The contrast agent flowed into the duodenum along the wall of the duct, and the jelly like mucus was removed from the stone mesh (Figure 2). Intrahepatic papillary mucinous tumor (ipnb-m) was considered by endoscopy. The patients recovered half a month after the operation and were followed up in the first month, the third month and the sixth month after the operation. The general condition was good and no tumor recurrence was found.

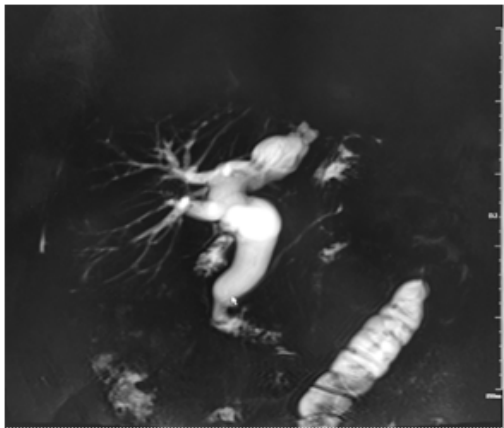


Figure 1: It is suggested that the common bile duct is dilated, the left intrahepatic bile duct is dilated, and the right anterior bile duct flows into the left hepatic duct.



Figure 2: Mucus embolus in bile duct was removed by ERCP choledochoscope with a little pus.

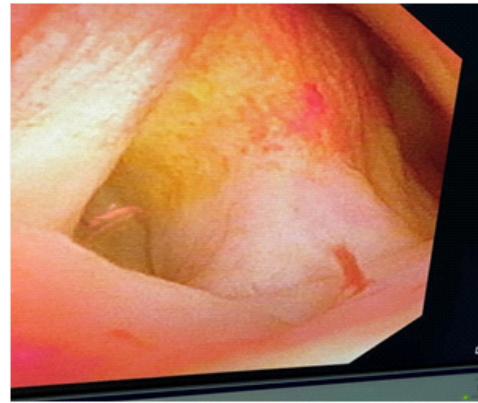


Figure 3: Intraoperative choledochoscopy showed orange peel like changes in the left hepatic bile duct (the lesion involved the left intrahepatic bile duct).



Figure 4: The bile duct was filled with jelly like fluid, and the lumen was saccular dilated, rough and leather like.

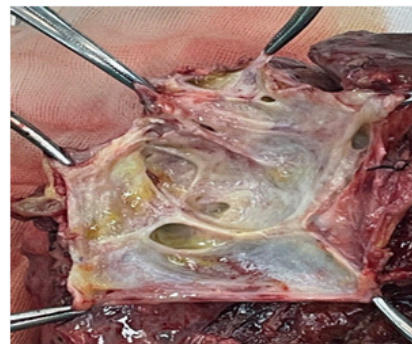


Figure 5: Histopathological examination: shows a dense papillary tumor, with a fibrovascular axis inside, and bile duct epithelial cells and goblet cells on the periphery. (X400 times)

Table 1: Changes in blood report and liver function of patients after admission.

Date	WBC (*109/L)	Total bilirubi (mmol/L)	Direct bilirubin (mmol/L)	Alanine transamination transferase (U/L)	Glutamy transferase (U/L)	Alkaline phosphatase (U/L)
2020-06-18	11.65	23.9	14.3	223	488	176
2020-06-25	3.73	35	24.2	282	273	129
2020-07-02	11.60	29.5	17.5	239	89	71
2020-07-05	10.48	19.0	11.9	213	78	67
2020-07-09	16.30	37.4	27.1	47	54	78

Postoperative pathology: Figure 5 medium grade intraductal papillary mucinous tumor of bile duct, dilatation of peripheral bile duct with medium grade intraductal neoplasia, and fatty degeneration of surrounding liver tissue. Immunohistochemistry: CK19 (+), CK7 (+), PCK (+), MUC5AC (+), MUC2 (+), ER (+), PR (+), Ki-67 (index 15%). It is consistent with intraductal papillary mucinous tumor (intermediate type).

Discussion

Intrabiliary papillary myxoma is derived from bile duct epithelial cells and is supported by connective tissue from the lamina propria. It is a rare benign tumor characterized by papillary and villous spreading and growth into the duct. It is histologically described as mucinous and papillary like tumor [1]. In 2010, the World Health Organization (WHO) classified mucinous cystic tumors and intraductal papillary mucinous tumors into two different solid tumors with rare onset [2]. Rate and mucin production are their common features, and the presence of ovarian-like stroma (OLS) has been identified as the diagnosis of MCN-L.

In related studies, IPMN-B is considered to be the biliary counterpart of pancreatic papillary mucinous tumor (IPMN-P), which is similar to IPMN-P, and IPNM-B has the typical characteristics of bile ducts, which can produce a large amount of mucin, leading to bile ducts extensive expansion [3]. IPNM-B histology can be divided into four types of pancreaticobiliary duct type, intestinal type, gastric type and eosinophil type. The most common type is pancreaticobiliary duct type, which accounts for about 47%. The expression of MUC1 and MUC2 can be distinguished by immunohistochemical staining [4]. Sakamoto et al. divided IPMN-B patients into 3 types, type 1 bile duct dilatation, type 2 cyst type, and type 3 intermediate type, which are a mixture of the above two types [5]. The cases reported in this article belong to the typical intermediate type lesions. The incidence of IPMN-B is extremely low. The incidence in eastern countries such as Japan and South Korea is much higher than that in western countries. This is because endemic hepatolithiasis, cholangitis and biliary infections are more common in Asian countries. This is the occurrence of IPMN-B. The main risk factors of IPMN-B are considered to be a precancerous lesion with adenoma cancer sequence, with a cancerous rate of 64%-89% [6]. The main clinical manifestations are intermittent abdominal pain, obstructive jaundice, and fever. Intermittent jaundice is helpful for the diagnosis of IPMN-B, which may be due to its secretion of more mucin and intermittent tumor separation caused by intermittent obstruction of the bile duct. Moreover, 30% of IPNM-B patients may be accompanied by bile duct stones, which are easily misdiagnosed as simple bile duct stones and repeated visits. There is no specific laboratory test for IPMN-B, and fluctuating bilirubin can be of certain value for its diagnosis. The levels of CEA and CA242 can help distinguish

between benign and malignant IPNM-B. Imaging methods such as MRCP, CT, and ultrasound are also of limited diagnostic value. The imaging manifestations of IPMN-B are tumors in the bile ducts, diffusely dilated bile ducts and mucus in the ducts, which are the unique characteristics of IPMN-B. Approximately 68% of patients may have simultaneous expansion of the proximal and distal bile ducts [7]. This is because the upstream expansion of the bile duct may be due to the tumor's own obstruction or the stenosis caused by the tumor, and the downstream expansion is due to excessive mucin secretion. When the hepatopancreas ampulla is obstructed, it causes diffuse dilation of the intrahepatic and extrahepatic bile duct system. Therefore; we should be highly alert to this disease when we find diffuse dilatation of the common bile duct. There are also some patients who have not found tumors or nodules in the bile ducts, because MRCP and CT are difficult to detect small or flat tumors, and it is also difficult to detect mucin. The CT and MRCP signal intensity of mucin is almost the same as that of bile [8].

At this time, it needs to be combined with ERCP. The defect of the bile duct filled with contrast agent in ERCP does not match the smooth tube wall filled with contrast agent on CT, and the duodenal papilla or bile duct has jelly-like mucus in the choledochoscopy, which is of diagnostic significance, CT+MRCP examination in this case report showed obvious bile duct dilatation and special signs of ERCP, which were consistent with the description in the literature. IPMN-B has a low incidence, atypical clinical symptoms and signs, and its preoperative diagnosis is very difficult. It needs to be related to hepatic mucinous mucinous cystic tumor (MCN-L), cholangiocarcinoma, congenital biliary dilatation, and recurrent bile duct Identification of stones.

There are various treatment methods for IPMN-B, including surgical resection of lesions, liver transplantation, and palliative treatment. Due to the characteristics of invasive precancerous lesions, radical surgical resection is the preferred treatment method. The surgical effect is good, and its five-year survival rate can reach 84% [9]. For patients who have no chance of surgery or who cannot tolerate surgery, laser cauterization or ablation, chemotherapeutic perfusion, argon ion beam coagulation, combined with endoscopic placement of bile duct stents, nasobiliary ducts or PTCD can be selected. It is also useful for IPMN-B patients certain effect. In general, the prognosis of IPMN-B is better than that of cholangiocarcinoma, but the recurrence rate is higher. According to statistics, the recurrence after radical surgical resection is as high as 29.3% [9], so IPNM-B requires long-term follow-up even if the margin is negative.

Conclusion

IPMN-B is a type of benign biliary tract tumors that are clinically rare, difficult to diagnose, and have a high malignant rate. During the clinical treatment process, we need to be highly vigilant about this type of disease, reduce misdiagnosis and missed

diagnosis, and early detection and early treatment are better for such patients. Improve the quality of life and improve the survival rate.

Declarations

Ethics approval and consent to participate: The protocol was approved by the Chengdu Second People's Hospital Clinical Research Ethics Committee, and parents of all subjects provided informed consent.

Consent for publication: Not applicable.

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