

CASE REPORT

Intraductal Papillary Mucinous Tumour of the Bile Ducts

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ABSTRACT

This report is of a rare case of intraductal papillary mucinous tumour of the bile ducts. As diffuse biliary duct dilatation can be the only radiological finding, it may be difficult to differentiate this condition from recurrent pyogenic cholangitis which is common in this region. Papillary tumour has a benign course and will benefit from early diagnosis and surgical intervention. This diagnosis should be considered in the presence of massively dilated intrahepatic ducts and endoscopic retrograde cholangiopancreatography is often diagnostic.

Key Words: Bile duct neoplasm, Papillary tumour

CASE REPORT

A 65-year-old man who was a chronic drinker and smoker was admitted to the United Christian Hospital with fever and painless jaundice. Liver function test was deranged with a slightly raised bilirubin level. Computed tomography (CT) scan revealed dilated intrahepatic (L>R) and common bile ducts. Left intrahepatic ductal stones were present with an atrophic left hepatic lobe (Figure 1). There was no intraductal filling defect or mass apparent on the CT scan. An abnormal hypodense collection with rim enhancement was present in the right lobe of the liver. The patient was diagnosed with recurrent pyogenic cholangitis complicated by abscess formation and treated accordingly. A full course of antibiotics was given and follow-up CT showed resolution of the right hepatic abscess.

Six months later, the patient was readmitted with right upper quadrant abdominal pain and jaundice. Blood test revealed a markedly raised bilirubin level of 111 to 500 $\mu\text{mol/L}$ (normal range, 5-21 $\mu\text{mol/L}$). CT scan showed further dilatation of the left intrahepatic ducts (Figure 2). Endoscopic retrograde cholangiopancreatography (ERCP) revealed a grossly dilated biliary

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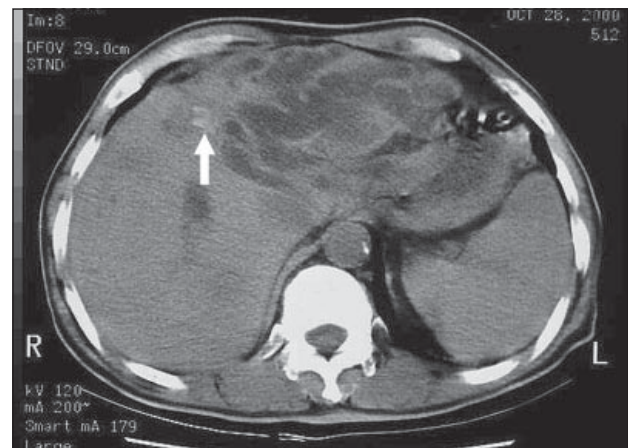


Figure 1. Pre-contrast computed tomography scan of the abdomen showing dilated intrahepatic ducts with atrophic left hepatic lobe and stones in the left intrahepatic duct (arrow).

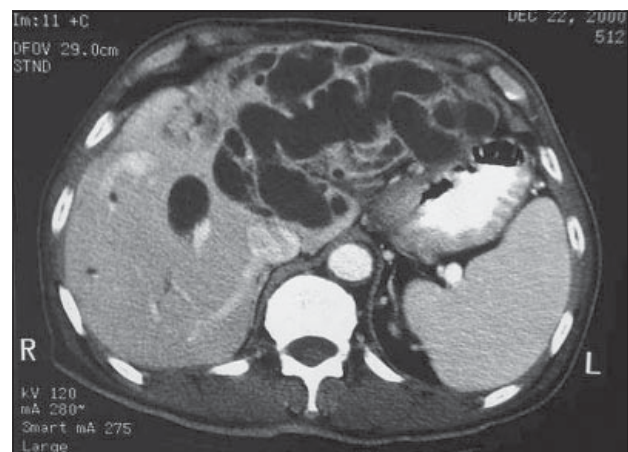


Figure 2. Contrast-enhanced computed tomography showing increasingly dilated intrahepatic ducts.

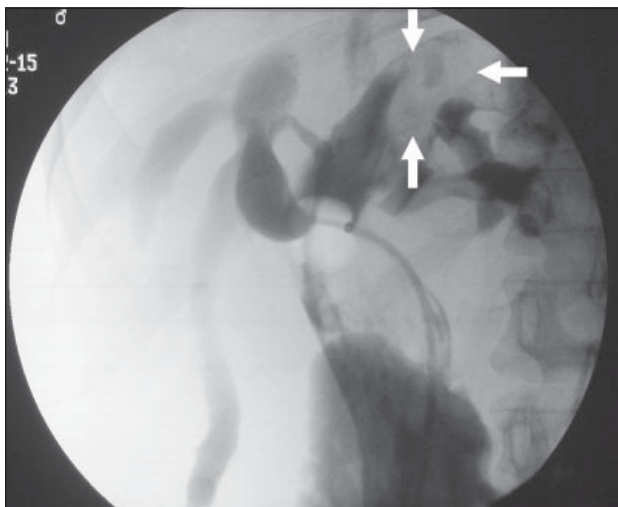


Figure 3. Endoscopic retrograde cholangiopancreatography showed a grossly dilated biliary system with multiple filling defects in the left intrahepatic duct (arrows).

system (Figure 3). The common bile duct measured 4 cm in diameter and the ductal orifice was grossly dilated. The left intrahepatic duct was distorted with stones in situ. A diagnosis of intraductal papillary mucinous tumour of the bile ducts was made based on the ERCP findings and the presence of copious amounts of mucus secretion.

Percutaneous transhepatic biliary drainage was done in both the left and right intrahepatic ducts to relieve the obstruction. The bilirubin level decreased from 500 $\mu\text{mol/L}$ to 323 $\mu\text{mol/L}$. Clinically, the patient's condition improved. He refused operation and follow-up ERCP 3 months later revealed diffuse papillomatosis along the ductal mucosal surface with copious amounts of mucus secretion in the biliary system. Eventually, the patient agreed to have an operation and a left trisectionectomy with caudate lobe resection and hepatojejunostomy were performed. Histology confirmed the diagnosis of intraductal papillary tumour with invasive carcinoma.

DISCUSSION

Papillary neoplasms of the bile ducts are rare pathology characterised by diffuse papillary proliferation of the bile duct epithelial cells. They account for 3% to 5% of cholangiocarcinomas and can arise from any portion of the intra or extrahepatic bile ducts.¹

This tumour can be histologically classified into papilloma and papillary adenocarcinoma.² Another condition with multiple neoplasms, called biliary papillomatosis, also belongs to this class of tumour and the neoplasms

usually appear as multicentric papillomas involving the intra- or extrahepatic tract.

Patients may present with abdominal pain, obstructive jaundice or biliary sepsis. The clinical symptoms and signs are caused by partial or complete biliary obstruction either by the tumour, by sloughed tumour debris, or by copious amounts of mucus secretion.

Unlike traditional ductal adenocarcinoma, papillary tumours 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. Therefore, early diagnosis and surgical resection are important and a benign course with long survival is expected.³

Most papillary tumours are peripheral in location and are nodular or papillary in shape. At imaging, the primary tumour may be depicted as an intraluminal, polypoid lesion or may not be seen when it is small (<1 cm in diameter) or spreads along a mucosal membrane. When these tumours produce excessive mucin, ultrasound and CT will show severe and diffuse dilatation of the intra- and extrahepatic ducts both proximal to and distal from the responsible tumour.^{4,5} When the tumour involves a segment or one hepatic lobe, the degree of bile duct dilatation is thus particularly severe, and the dilated bile ducts appear crowded. At ultrasound, an intraductal mass is usually echogenic. As it is confined within the bile ducts, the echogenic walls remain intact.² At CT, an intraductal papillary tumour can appear as a hypo- or iso-attenuating soft tissue mass within the dilated duct relative to the liver parenchyma, or as a segmental thickening of the bile duct wall.^{6,7} The tumour may show delayed enhancement on contrast-enhanced CT.

ERCP seems superior to CT as it shows the dilated ducts with floating filling defects. A tumour may be small and flat and the ductal wall is sometimes irregular. The mass reveals fine surface irregularities, either in the form of a velvety or serrated contour, representing the tumour's papillary surface.⁸⁻¹⁰

Endoscopic appearance of a patulous papillary orifice with copious amounts of mucus secretion is a specific finding of papillary mucinous tumour. Cytological examination by epithelial brushing and biopsies during ERCP is an effective preoperative procedure for the diagnosis of papillary tumour of the bile duct.

In conclusion, intraductal papillary tumour 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 tumour of the bile duct should be considered. ERCP is often diagnostic when a copious amount of mucus is present.

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