Case Report

Intrahepatic Biliary Cystadenoma Presenting with Obstructive Jaundice

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Biliary cystadenoma (BCA) is a rare neoplasm of the bile duct with malignant potential. We report a case of intrahepatic BCA with an unusual presentation of obstructive jaundice.Computed tomography scan of the abdomen revealed a dilated common bile duct and intrahepatic ducts with internal septa. Endoscopic retrograde cholangiography showed an oval filling defect in the bile duct causing the obstruction. At laparotomy, this proved to be a multiloculated mucinous polyp in the common bile duct, with its origin in the left intrahepatic duct, detected using intraoperative choledochoscopy. A left hemihepatectomy was performed, and histology confirmed intrahepatic mucinous BCA with mesenchymal stroma. The imaging process and surgical options for BCA are discussed. [Asian J Surg 2004;27(3):243–5]

Introduction

Biliary cystadenoma (BCA) is a rare neoplasm arising from intrahepatic or extrahaepatic bile ducts. The majority (80%) arise from the intrahepatic ducts and are usually asymptomatic. Most (68%) originate from the right intrahepatic ducts, mainly in the posterior segment. The presenting symptoms are usually right hypochondrium pain or abdominal mass. In contrast, extrahepatic BCA presents with obstructive jaundice either directly by tumour blockage or by their mucin secretion.

In this paper, we present a patient with BCA involving the left intrahepatic and left main hepatic duct presenting as obstructive jaundice. Such occurrences are rare, and we review the imaging process and the surgical strategy.

Case report

A 58-year-old Chinese lady was admitted with complaints of right hypochondrium pain, jaundice with pale stools, and tea-coloured urine for 3 days. On examination, she was febrile with tenderness in the right hypochondrium. There was hepatomegaly. Liver function tests showed elevated serum total bilirubin (283 µmol/L), alkaline phosphatase (467 U/L), alanine transaminase (688 U/L), and aspartate transaminase (385 U/L). Tumour markers were normal except for an elevated serum CA19-9 of 1,188 U/mL.

Abdominal computed tomography (CT) scan showed dilatation of intrahepatic ducts and the proximal common bile duct (CBD), with abrupt transition to normal calibre (Figures 1 and 2). The impression was either a CBD stricture or cholangiocarcinoma. Endoscopic retrograde cholangiography showed an oval filling defect in the mid-CBD with dilated intrahepatic ducts, particularly on the left side (Figure 3A). A plastic biliary stent was deployed to decompress the system.

The patient underwent laparotomy, which revealed a polypoidal mass at the confluence, causing the obstruction. This mass, with an intact stalk, was extracted through a choledochotomy (Figure 3B). Check choledochoscopy, however, revealed cystic disease of the intrahepatic segments of the left bile duct. The mass originated in the left intrahepatic ducts and had prolapsed into the common hepatic duct (Figure 3C).

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Neoplasms of bile duct origin. Symptoms are usually related to their mass effect, but biliary obstruction with intrahepatic BCA is rare. There have only been three reports of luminal bile duct obstruction causing jaundice from intrahepatic BCA, two of which were by protruding polypoidal masses and one from intracystic haemorrhage. Intraluminal mucin secretion by the tumour can cause ductal dilatation.

BCA has two forms: the more common mucinous and the rare serous type. The former is further subdivided by the presence or absence of mesenchymal stroma between an inner epithelial lining and an outer connective tissue capsule. Mesenchymal stroma occurs exclusively in young/middle-aged women, and can transform into biliary cystadenocarcinoma (BCAC). Both dysplastic epithelial changes and invasive adenocarcinoma have been found in such cases. Evolution from BCA to BCAC has been documented. BCA without mesenchymal stroma occurs in both men and women and, in the former, seems to develop into more aggressive BCAC.

In general, intrahepatic BCA appears as cystic liver masses on imaging. They are typically solitary and can pose a difficult diagnostic dilemma. However, with current understanding of the specific imaging characteristic features of the different cystic liver lesions, a preoperative diagnosis is always possible. CT scan or magnetic resonance imaging (MRI) helps to confirm the diagnosis and aids in preoperative planning. Masses are typically multilocular with internal septa (Figures 1 and 2) and mural nodules bounded by a well-defined fibrous capsule. Increased thickening and enhancement of the septa, nodules and its capsule with coarse calcifications have been cited as determinants of malignancy.

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Left hemi-hepatectomy was performed with division of the left main hepatic duct at the confluence. The choledochotomy was closed over a T-tube. The patient had an uneventful postoperative recovery.

Histology revealed mucinous cystadenoma of the left intrahepatic ducts with focal moderate atypia and compact ovarian type mesenchyme. The polypoidal mass in the CBD also showed multiple locules, lined by a single layer of mucinous glandular epithelium with focal areas of cellular mesenchyme beneath it. There was no evidence of back-to-back or cribriform glands.

At 1-year follow-up, the patient was well with no evidence of recurrent disease.

Discussion

BCA occurs most commonly in middle-aged females. They are uncommon tumours, accounting for fewer than 5% of neoplasms of bile duct origin. Symptoms are usually related to their mass effect, but biliary obstruction with intrahepatic BCA is rare. There have only been three reports of luminal bile duct obstruction causing jaundice from intrahepatic BCA, two of which were by protruding polypoidal masses and one from intracystic haemorrhage. Intraluminal mucin secretion by the tumour can cause ductal dilatation.

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MRI and its complementary multiplanar cholangiography give added value to both diagnosis and level of obstruction,
especially in the setting of obstructive jaundice. Fluid-containing BCA is usually hypointense on T1-weighted images and hyperintense on subsequent T2-weighted images. However, variable signal intensities have been reported, which correlate to the presence of haemorrhage or hyperproteinous/mucinous contents of the BCA. A hyperintense multilocular liver lesion on T1-weighted MRI may still represent a BCA or BCAC.

Endoscopic retrograde cholangiography or percutaneous transhepatic cholangiography, though invasive, are useful to delineate the level and cause of obstruction or for stenting in the presence of cholangitis. The cholangiogram may show a filling defect due to a polyp-like lesion (Figure 3), as in this case, or, rarely, mucin in the bile duct.

Preoperative diagnosis helps to strategize surgery. The bile duct could be explored through a choledochotomy or excised primarily if the BCA is solely extrahepatic. Intraoperative choledochoscopy is useful to assess the ductal system, as exemplified by this case, helping to detect the origin of the intrahepatic component, which could be missed on preoperative imaging.

In the past, treatment of BCA has included aspiration, marsupialization, internal drainage and partial excision, probably due to poor preoperative imaging and the lack of understanding of BCA’s premalignant potential. However, with current cumulative data, the ideal treatment should be complete excision of the tumour. Enucleation of this cystic lesion is simple and safe, provided that there is no coexistent malignancy. This can be excluded by intraoperative ultrasound and frozen section analysis. Albeit this, differentiation can still be difficult and, thus, a formal or partial hepatectomy is necessary to ensure complete clearance.

Intrahepatic BCA can present with obstructive jaundice. CT scan and MRI/cholangiography help in preoperative diagnosis. The intraoperative choledochoscope helps to evaluate intrahepatic ductal involvement, which will require liver resection for total excision of BCA.

**References**