Bronchobiliary fistula due to acute cholecystitis in a suprahepatic gall bladder

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Summary: We describe a patient presenting with painless jaundice, anorexia and pruritus. The gall bladder was found to be lying above and behind a hypoplastic right lobe of liver. There was no evidence of cholangitis or biliary obstruction. The patient subsequently developed a bronchobiliary fistula with severe wheeze, cough and bile-stained sputum. Emergency percutaneous drainage of the gall bladder led to immediate cessation of bronchospasm and biloptysis, rendering the patient fit for definitive surgery.

Introduction

Expectoration of bile is pathognomonic of a bronchobiliary fistula.¹ This is rare and usually associated with intrahepatic abscess or hydatid disease.² Less common causes are congenital fistulae, trauma or surgical manoeuvres.¹ A few cases have been described in association with obstruction of the biliary tree due to calculi,³ post-operative biliary stricture,⁴ or chronic pancreatitis.⁵ Bronchobiliary and bronchopleural fistulae may also result from acute cholecystitis complicated by gall bladder perforation and subphrenic abscess.^{1,4}

We report a patient who was initially thought to have acute hepatitis followed by cholestasis and was found to have congenital abnormalities of the liver and biliary tree. He subsequently developed acute cholecystitis complicated by subphrenic abscess and bronchobiliary fistula. Immediate percutaneous gall bladder drainage was life-saving, allowing improvement of the patient's condition prior to surgery.

Case report

A 63 year-old man presented with a two week history of anorexia, pruritus, pale stools and dark urine. He had been an insulin-dependent diabetic for 10 years but was on no other medication. On examination he was markedly jaundiced with scratch marks. A firm smooth non-tender liver was palpable 12 cm below the

costal margin, and the spleen was just palpable. He had a serum bilirubin of 173 $\mu\text{mol/l}$ (normal < 17 $\mu\text{mol/l}$), an alkaline phosphatase of 827 IU/l (normal < 130 IU/l) and aspartate transaminase (AST) of 2085 IU/l (normal < 40 IU/l). Serum albumin was normal but the prothrombin time was prolonged at 21 seconds, correcting to 14 seconds after vitamin K. Autoantibodies and serological markers for viral hepatitis were absent.

Ultrasound failed to identify the extrahepatic biliary tree or gall bladder, but the intrahepatic bile ducts were not dilated. Liver biopsy showed abundant fibrous septa, centred around portal areas, intersecting the tissue and in places forming nodules. Bile duct proliferation, neutrophilic infiltrates and hepatocellular injury with Mallory bodies were striking features, and there was excessive copper associated protein. The overall appearances, while unusual and not diagnostic, suggested chronic biliary tract disease. Percutaneous cholangiography showed a common bile duct of normal calibre and free flow of contrast into the duodenum.

While being investigated, his serum bilirubin rose to 452 IU/l, and alkaline phosphatase remained high at 601 IU/l. The AST had fallen to 153 IU/l. This biochemical pattern suggested an acute hepatocellular injury followed by cholestasis, perhaps due to non-A non-B hepatitis. He was therefore given cholestyramine and a therapeutic trial of prednisolone 30 mg/day.⁶ His pruritus resolved rapidly and his liver function tests improved with a fall in bilirubin to 20 $\mu\text{mol/l}$, although the alkaline phosphatase remained elevated at 417 IU/l.

The prednisolone was tailed off 9 months later. Soon afterwards his jaundice and pruritus returned

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and he was readmitted for further investigation. Liver biopsy appearances were unchanged. A computerized tomographic (CT) scan without intravenous contrast (Figures 1 and 2) showed an irregular liver outline and a large lucent area in the posterior part of the right lobe which was thought to be a benign simple cyst. Endoscopic retrograde cholangiography (ERC) revealed a large gall bladder lying in an unusual position high up in the right lobe of liver (Figure 3). Repeat CT scanning immediately after ERC confirmed that the liver 'cyst' filled with contrast and was, in fact, an ectopic gall bladder.

He was readmitted 3 months later with right hypochondrial pain, increasing pruritus, cough and green sputum. Portosystemic encephalopathy and ascites had developed. He was found to have marked bronchospasm and a chest X-ray showed collapse and consolidation of the right lobe. Despite antibiotics, bronchodilators and chest physiotherapy, his bronchospasm worsened and his cough became productive of bile (up to 100 ml/day). A percutaneous drain was inserted into the gall bladder under CT control with an immediate reduction in bronchospasm and no further bile expectoration. ERC demonstrated a communication between the gall bladder and a cavity above the liver posteriorly.

At operation an abscess cavity was found in communication with the gall bladder and diaphragm. The right lower lobe of lung was adherent to the diaphragm and a fistula was identified (Figure 4). The right lobe of liver was hypoplastic and the left lobe enlarged. The abscess cavity was drained and the diaphragmatic defect sutured. The gall bladder was freed and

removed from within the right lobe of liver. An operative cholangiogram via the cystic duct showed free flow of contrast into the duodenum. Histology of the gall bladder showed acute on chronic cholecystitis.

The patient initially made a good post-operative recovery but then developed worsening ascites and renal function. He died 4 weeks post-operatively from combined hepatic and renal failure. At post-mortem the liver weighed 1300 g and showed reversal of the normal relationship between lobe size. The gall bladder bed was identified on the posterior aspect of the right lobe. The site of ligature of the cystic duct was identified, and the common bile duct and each hepatic duct appeared to conform to the normal anatomy of the porta hepatis. The vascular radicles supplying the liver also appeared normal. No evidence of congenital abnormality was seen in the rest of the body. Sections from both lobes and hila showed identical features and were characterized by portal fibrosis with nodule formation.

Discussion

This case displayed a confusing combination of clinical, radiological and histopathological features. The patient's initial presentation resembled an acute hepatitis with anorexia, painless jaundice and an extremely high aspartate transaminase level. However, the correction of the prothrombin time with vitamin K, and pruritus, implied cholestasis. Liver biopsy supported chronic biliary tract disease. However percutaneous cholangiography did not iden-

Figure 1 CT scan showing liver with an irregular outline suggesting cirrhosis, and an apparent cyst posteriorly (arrowed).

Figure 2 Line diagram illustrating the features seen on the CT scan in Figure 1. The gall bladder (GB) is lying behind the hypoplastic right lobe of liver (RL) and anterior to the diaphragm. The arrow shows the direction of the subsequent fistula between the acutely inflamed gall bladder and the right lung (RLG).

Figure 3 ERCP showing the pancreatic duct (P), the common bile duct (B) intrahepatic ducts of normal calibre, and the abnormally positioned gall bladder.

tify any primary bile duct disease or extrahepatic obstruction.

We believe the most likely explanation for the initial high AST level and subsequent hepatocellular dysfunction was recurrent cholecystitis superimposed on the congenital abnormality. A transiently high AST level (600) may be associated with extrahepatic biliary disease.⁷ The abnormal gall bladder location and the use of corticosteroids may have masked symptoms of the cholecystitis and biliary sepsis. There are four previous cases described with a hypoplastic right lobe of liver and abnormal positioning of gall bladder and cystic duct.^{8,9} Acute cholecystitis developed in two of these patients but neither was complicated by chronic cholestasis or bronchobiliary fistula.

It is of interest that bronchobiliary fistula presented

Figure 4 Diagram of the operative findings showing the fistula between the suprahepatic gall bladder (GB), subphrenic abscess cavity (SP) and right lung (RLG). The positions of the right lobe of liver (RLL), cystic duct (CD) and biliary tree (BT) are shown.

with cough and green sputum in our patient; this experience has been reported previously.¹⁰ Chest physiotherapy may have precipitated the opening of the fistula and the sudden deterioration in our patient. Emergency percutaneous drainage of the suprahepatic gall bladder under CT control stopped the flow of bile into the bronchial tree, relieved the bronchospasm, and rendered the patient fit for surgical intervention.

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