Benign biliary papillomatosis in a patient with a choledochal cyst presenting as haemobilia: a case report

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ABSTRACT

Biliary papillomatosis is a rare condition usually detected on imaging or postoperative histopathology. It may be asymptomatic or present with features of cholangitis. We report the management of a patient presenting with haemobilia.

KEYWORDS

Benign biliary papillomatosis – Choledochal cyst – Hemobilia

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We report a rare case of benign biliary papillomatosis (BP) in a choledochal cyst. Papillomatosis arising from the intrahepatic portion of a choledochal cyst presenting with haemobilia has not been reported before.

Case history

A 57-year-old woman presented initially with periumbilical pain that had lasted for a week. She also complained of tiredness but did not have loss of weight or appetite. Imaging in another hospital had revealed common bile duct thickening with intrahepatic biliary radicle dilatation and biliary sludge. She was suspected of having cholangiocarcinoma and underwent endoscopic retrograde cholangiopancreatography (ERCP) with stent placement.

General and systemic examination and blood tests were non-contributory. Magnetic resonance cholangiopancreatography with computed tomography of the abdomen (MRCT) showed dilatation of the common bile duct and left hepatic duct (LHD). There were multiple hyperdense lesions in the LHD suggestive of intrahepatic stones (Figs 1 and 2).

The patient was offered cholecystectomy, cholangioscopy, extraction of the hepatolithiasis and hepaticojejunostomy. She came back for the operation four months later. She was pale with a history of two episodes of melaena. Her haemoglobin was 5.7g% and her blood picture was consistent with iron deficiency anaemia. The coagulation profile was normal. Oesophagogastroduodenoscopy, colonoscopy and review of MRCT of the abdomen did not reveal the source of bleeding. She was diagnosed with obscure gastrointestinal bleeding and on-table enteroscopy was added to the plan of operation.

Figure 1 Multiple filling defects/lesions in the left hepatic duct

Figure 2 Dilatation of the common bile duct and left hepatic duct

At operation there was a stent in the common bile duct with blood in the biliary tree. Stones were not present. There was a tumour in the LHD (frozen section: well differentiated glandular neoplasm). The common bile duct was dilated, consistent with choledochal cyst type IVA. The patient underwent cholecystectomy, excision of the choledochal cyst, left hepatectomy, hepaticojejunostomy with a radiological access loop. Histopathology revealed benign mucin producing BP. Malignant changes were not seen in the rest of the choledochal cyst or gallbladder. She is well on follow up four years after surgery.

Discussion

Haemobilia was first described in 1654 by Francis Glisson¹ and the term was coined by Sandblom.² The triad of upper abdominal pain, upper gastrointestinal haemorrhage and jaundice is present only in around 30% of patients. Haemorrhage can be acute and massive or chronic. Treatment depends on the cause of bleeding. Common causes include trauma, iatrogenesis, neoplasm, rupture of hepatic aneurysm, hepatic abscess, choledocholithiasis, ductal parasite and coagulopathy. Bleeding from tumours causes haemobilia in about 6% of cases.

BP ('laputa tumor') was first described by Chappet in 1894.⁵ About 200 cases have been reported. It is seen in an older population with a male-to-female ratio of 2:1. It is associated with *Clonorchis sinensis* infestation, primary sclerosing cholangitis, Caroli disease and dysplasia of the gallbladder.^{4,5} Symptoms are due to biliary obstruction (tumour emboli, mucus plugs and/or lithiasis), cholangitis and haemobilia.⁶ Obstruction may cause ductal dilatation; this may be lobar, segmental, generalised or cystic in appearance depending on the location of the tumour.

Ultrasonography shows features of biliary obstruction, non-shadowing biliary masses, sludge and choledocholithiasis.⁷ Computed tomography (CT) findings are non-specific (intrahepatic masses and duct dilatation). Mucobilia, multiple filling defects and supple stenoses are seen on ERCP. The extent of the lesions is better outlined by magnetic resonance imaging than by CT but transhepatic cholangiography or cholangioscopy may be the best investigation to assess the extent of involvement. It can involve the intra- or extrahepatic ducts and can be localised or diffuse. Fine needle aspiration shows distinctive features permitting differentiation from cholangiocarcinoma. Grossly, friable papillary masses are seen. At microscopy, multiple papillary adenomas with complex gland formation and variable degrees of atypia are seen.

BP has been subdivided into five classes according to the degree of cytological and structural atypia. Atypia and dysplastic changes are found frequently. BP may be a precancerous lesion induced by an abnormal pancreaticobiliary

junction. Involvement of the KRAS and tumour protein 53 systems with a possible adenoma-carcinoma sequence has been proposed.^{4,8-10}

Treatment options include palliative stenting, brachytherapy with iridium-192, electrocoagulation, cholangioscopic resection and hepatic resection. Liver transplantation has been reported.^{11,12} About 50–65% of patients succumb to their disease, the prognosis being worst in diffuse disease.¹⁵

Choledochal cysts can rarely present with haemobilia secondary to cholangiocarcinoma, iatrogenesis (stenting), anomalous arteriovenous communication, cyst rupture and trauma.¹⁴ While papillomatosis has been separately reported associated with a choledochal cyst and haemobilia,^{6,14} clinical presentation with haemobilia in a patient with a choledochal cyst due to papillomatosis has not been reported.

Conclusions

Haemobilia is an important differential diagnosis of obscure gastrointestinal bleeding. BP should be borne in mind when imaging suggests intrahepatic stones. Intraoperative assessment and modification of the operative plan is essential for a good outcome.

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