Solitary cystic dilatation of the intrahepatic bile duct

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Abstract

A 31 year old man was hospitalised with general fatigue and epigastric pain. Abdominal ultrasonography, computed tomography, and magnetic resonance imaging showed a cystic lesion in the left lobe of the liver. Endoscopic retrograde cholangiopancreatography and percutaneous transhepatic cholangiography revealed a localised dilatation of the intrahepatic bile duct without any obstruction. However, a large mass of mucinous material was noted in the saccular intrahepatic duct and the common bile duct. There was no evidence of a choledochal cyst, anomalous pancreaticobiliary ductal union, or congenital cystic change of the kidneys. A possible diagnosis of mucinous cystic neoplasm of the intrahepatic bile duct was made and a left hepatectomy performed. Cholangiography of the resected specimen showed a non-obstructive solitary cystic dilatation of the left hepatic duct. Histologically, the dilated duct was lined with columnar biliary epithelium without any papillary proliferation and/or atypia. Neither malignancy nor hepatic fibrosis was observed; the term "solitary cystic dilatation of the intrahepatic bile duct" perhaps is more descriptive and the concept easier to understand.

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presented with neither a pertinent family history nor a personal history of blood transfusion, tattooing, or drug abuse. On physical examination, there was a slight tenderness in the epigastrium and right hypochondrium, and the liver was slightly enlarged. Results of liver function tests were: total bilirubin, 2.1 mg/l (normal value 0.2-1.0); alkaline phosphate, 127 IU/l (28-84); γ glutamyltranspeptidase 314 IU/l (4-30); aspartate aminotransferase 117 IU/l (7-20); and alanine aminotransferase 425 IU/l (7-28). Virus markers were negative for hepatitis A, B, and C. Abdominal ultrasonography, computed tomography, and magnetic resonance imaging showed a cystic lesion in the left lobe of the liver. Choledochal cysts and anomalous pancreaticobiliary ductal union were not demonstrated on endoscopic retrograde cholangiopancreatography. Percutaneous transhepatic cholangiography revealed a localised dilatation of the left intrahepatic bile duct. There was a large mass of mucinous material in the saccular intrahepatic bile ducts and the common bile duct. Under the diagnosis of a possible mucinous cystic neoplasm of the intrahepatic bile duct, we performed a left hepatectomy. A cholangiogram of the resected specimen showed a solitary cystic dilatation of the left hepatic duct (fig 1). Histologically, the

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Recently, Terada and Nakanuma described two cases of solitary cystic dilatation of the intrahepatic bile duct without choledochal cysts, anomalous pancreaticobiliary ductal union, or congenital cystic change in the kidneys.¹ To date, only 13 such cases have been reported.¹⁻⁶ Several other terms have been used in the literature with regard to localised or non-obstructive dilatation of the intrahepatic bile duct. These include the localised form of Caroli's disease,^{7,8} localised dilatation of a major intrahepatic duct,⁴ and a type V congenital bile duct cyst.⁹

We report a case of solitary cystic dilatation of the intrahepatic bile duct as well as a review of the literature, and discuss the terminology concerning this entity.

Case report

In April 1995, a 31 year old man was admitted to Kawasaki Medical School Hospital because of general fatigue and epigastric pain. He

Figure 1 Cholangiogram of the resected specimen showing a solitary cystic dilatation of the left hepatic duct.

Figure 2 The dilated left hepatic duct is lined with mucinous columnar epithelium (haematoxylin and eosin, original magnification $\times 40$).

dilated duct was lined with columnar biliary epithelium without any papillary proliferation or atypia (fig 2). There was no evidence of "mesenchymal stroma", malignancy, or hepatic fibrosis. From these clinical and pathological findings, we made a diagnosis of solitary cystic dilatation of the intrahepatic bile duct.

Discussion

The terminology concerning localised or non-obstructive dilatation of the intrahepatic bile duct is somewhat controversial. We found that at least four terms have been used in the literature for cases such as ours: localised form of Caroli's disease,⁷ localised dilatation of a major intrahepatic duct,⁴ a type V congenital bile duct cyst,⁹ and solitary cystic dilatation of the intrahepatic bile duct.¹

Caroli's disease, first described by Caroli *et al* in 1958,⁷ is a rare congenital disorder characterised by cystic dilatation of the intrahepatic bile ducts. According to Barros *et al*¹⁰ six of 46 cases (13%) of Caroli's disease were isolated forms of intrahepatic cystic dilatations. Our case may be categorised into the localised form of Caroli's disease. However, normally the localised form of Caroli's disease involves all of the left or right lobe of the liver.⁸ Therefore, our case is not a typical case and may be called a limited form of Caroli's disease.

Localised dilatation of a major intrahepatic duct was a term proposed by Longmire *et al* in 1971.⁴ Their classification is subdivided into two categories: intrahepatic and extrahepatic. The intrahepatic type is also divided into primarily parenchymal and primarily ductal, with the latter being classified into localised dilatation of a major intrahepatic duct and multiple cystic dilatations of the intrahepatic ducts.

The term type V congenital bile duct cyst was proposed by Todani *et al* in 1977.⁹ Type V is an intrahepatic bile duct cyst and it may be single or multiple (our case was single). Todani *et al* suggested that type V might be closely related to intrahepatic stones and could be categorised as a pure form of Caroli's disease without hepatic fibrosis.

Finally, solitary cystic dilatation of the intrahepatic bile duct is a term that was proposed by Terada and Nakanuma in 1987.¹ Although this is a rather descriptive term, this entity has some clinicopathological features such as female predominance and cystic dilatation involving the right or left hepatic duct near the hepatic hilus. In addition, there is no evidence of related conditions including choledochal cysts, anomalous pancreaticobiliary ductal union, or congenital cystic change in the kidney. As Terada and Nakamura pointed out, bile stasis and mucous hypersecretion are important for dilatation of the intrahepatic bile duct,¹ our case may be explained by mucous hypersecretion as a large mass of mucinous material was seen in the intrahepatic duct.

To date, the above mentioned terms have been used for the same localised or non-obstructive dilated lesion of the intrahepatic bile duct. We prefer to use the last term—solitary cystic dilatation of the intrahepatic bile duct—because it is descriptive and the concept is easy to understand. Furthermore, it has clinicopathological features. Although this lesion is very rare and under-recognised, it should be considered by both pathologists and clinicians.

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