Adult hepatic fibropolycystic disease presenting as obstructive jaundice

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Abstract

Obstructive jaundice caused by compression of the common hepatic duct by a simple hepatic cyst in a 31 year old Euroid man is reported. The jaundice and duct compression resolved after percutaneous aspiration of the cyst under ultrasound direction and the patient has been well for 12 months.

Simple hepatic cysts are usually asymptomatic and are often an incidental finding at necropsy.^{1,2} The symptoms and signs described in association with hepatic cysts are abdominal distension; epigastric discomfort, nausea, and vomiting due to duodenal/gastric compression; abdominal pain, hepatomegaly, and rarely portal hypertension.^{3,4} These features are usually present in the fourth or fifth decade of life. Hepatic function is not disturbed as there is no parenchymal involvement.⁵

We report obstructive jaundice in a patient in his 40s, caused by a simple hepatic cyst which was successfully resolved by an ultrasound guided percutaneous aspiration.

Case report

A 31 year old Euroid man presented to another hospital with a three week history of painless

jaundice, severe pruritus, dark urine, and pale stools. He was on no medication and had no previous episodes of jaundice. Physical examination apart from jaundice and modest hepatomegaly was unremarkable.

An abdominal ultrasound scan showed extra- and intrahepatic biliary dilatation but no cause was identified. A laparotomy was therefore performed. This showed a non-dilated common bile duct, an empty gall bladder, and ductal dilatation at the level of the porta hepatis, but no obstructive lesion was defined. An operative liver biopsy specimen showed severe cholestasis with bile duct multiplication and widening in the portal zones and focal hepatic necrosis consistent with extrahepatic biliary obstruction.

After this the patient was transferred for further help in his management. We noted he was well, although still jaundiced (bilirubin 32 $\mu\text{mol/l}$ (normal range 3-14), alkaline phosphatase 685 U/l (normal range 30-140), aspartate aminotransferase 164 U/l (normal range 9-52)). His past medical history was unremarkable but his grandfather had died from uraemia caused by polycystic renal disease.

An endoscopic retrograde cholangiopancreatogram (ERCP) was performed. The pancreatogram was normal but the cholangiogram showed a smooth stricture 2 cm above the insertion of the cystic duct (Fig 1). The gall bladder was filled but no contrast medium passed through the stricture. An abdominal ultrasound scan showed a dilated biliary tree and a 4 cm diameter hepatic cyst adjacent to the porta hepatis. Some 30 ml of clear fluid was aspirated under ultrasound guidance. Cytology of the aspirate showed ciliated columnar cells. No other cysts in any other organ (kidneys, spleen, pancreas) were identified.

The patient's jaundice resolved and eight weeks later a repeat ERCP showed a normal cholangiogram (Fig 2), and an abdominal ultrasound scan showed no biliary dilatation and a small 1.5 cm diameter cyst adjacent to the porta hepatis.

Twelve months later the patient is well, with normal liver function tests and no ultrasound evidence of an increase in the size of the hepatic cyst.

Discussion

Jaundice caused by duct obstruction is a rare complication of solitary and polycystic liver disease,⁶ and has not previously been shown to resolve with percutaneous cyst aspiration.

Hepatic cystic disease is associated with cysts in other organs (kidney, pancreas, spleen), but this was not so in our patient.

Some 50% of patients have polycystic kid-

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Figure 1: Smooth stricture 2 cm above insertion of cystic duct is shown on endoscopic retrograde cholangiopancreatogram (ERCP).

Figure 2: Repeat ERCP showed a normal cholangiogram.

neys.^{1,2,7} The grandfather of our patient had polycystic renal disease, but our patient had normal renal function and no cysts in the kidneys.

Although only one hepatic cyst was found in our patient the number of cysts present within the liver can vary from one to many in fibropolycystic disease, and in view of the family history of polycystic renal disease we regarded this patient as being within the spectrum of this condition. Solitary non-parasitic hepatic cysts are increas-

ingly regarded as a variant of polycystic disease.⁸

The hepatic cysts arise from defective embryological development of intrahepatic bile ducts and are lined by a secretory epithelium of cuboidal or columnar cells. Ciliated columnar cells were identified from the cystic fluid in our patient. Analysis of the cystic fluid indicates that it resembles that fraction of human bile which is secreted independently of the presence of bile salts.⁹

The cysts, although congenital, are formed by functioning secretory bile duct epithelium and gradually increase in size throughout childhood and early adult life to cause symptoms in the fourth or fifth decade. In this case the cyst reached a critical size which resulted in mechanical compression of the common hepatic duct and this was relieved by aspiration.

The cysts, if symptomatic, are best managed by percutaneous aspiration under ultrasound control, although they have a tendency to recur, and introduction of a sclerosant may delay this.^{9,10} The prognosis of the condition is governed by the presence of renal involvement.

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