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Case report

Biliary peritonitis due to choledochal cyst presenting in late pregnancy

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CASE HISTORY

A previously healthy 23-year-old primigravida at 35 weeks' gestation presented with ante-partum haemorrhage and evidence of placental abruptio on ultrasound scanning. Emergency Caesarean section was performed via a Pfannenstiel incision, and a healthy baby was delivered in good condition. The obstetrician noticed that the peritoneal cavity contained some free reactive fluid, and surgical assistance was requested. It was difficult to carry out a full investigative laparotomy through the Pfannenstiel incision, but the appendix appeared normal and the stomach, duodenum, gallbladder, and pancreas were all normal on palpation. The colon was dilated from the caecum to the splenic flexure. The appendix was removed and a caecostomy tube was inserted via the appendix stump, to deflate the colon. A peritoneal drain was inserted and the incision closed. Following this, her condition did not improve and she remained ill, with abnormal liver function tests, elevated temperature and signs of generalised peritonitis.

Four days after the Caesarean section a further laparotomy was carried out, through a right paramedian incision. On this occasion, the peritoneal cavity contained a lot of bile and the gallbladder was oedematous and inflamed, with a gangrenous and perforated cystic duct. Emergency cholecystectomy was performed and the common bile duct was explored via a supraduodenal incision. An intra-operative T-tube cholangiogram revealed a large choledochal cyst, extending from the origin of the cystic duct to the lower end of the common bile duct, with absence of flow from the common bile duct into the duodenum (Fig 1). The choledochal

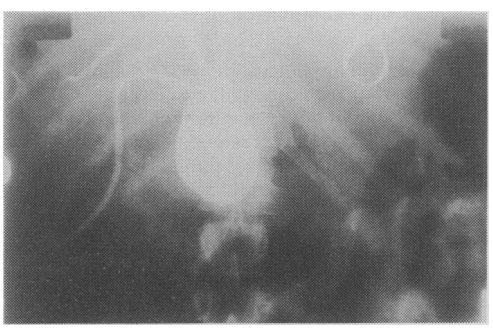

Fig 1. Intra-operative T-tube cholangiogram showing the choledochal cyst and absence of flow of contrast into the duodenum.

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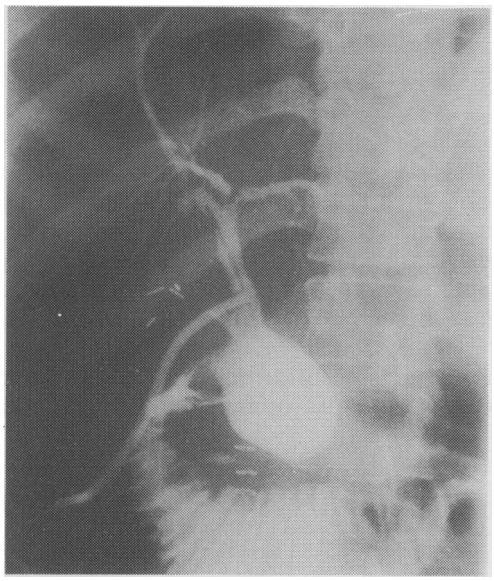

Fig 2. T-tube cholangiogram, seven days post-operatively, showing free flow through the choledochocysto-duodenostomy.

cyst was not apparent prior to the cholangiogram. In the first instance an inflamed and distended gall-bladder was found and, after its removal, an operative cholangio-gram revealed a cystic dilatation. There was no evidence of cyst formation in the liver, kidneys or pancreas. Biopsy of the choledochal cyst was not attempted. A chole-dochocysto-duodenostomy was performed and the common bile duct was drained via a T-tube.

Her post-operative recovery was uncomplicated, liver function tests returned to normal and a T-tube cholangiogram, seven days post-operatively, showed free flow through the choledochocysto-duodenostomy (Fig 2). Six months post-operatively she remained well, with normal liver function tests.

COMMENT

Choledochal cyst is a rare anomaly of the biliary tract. A male-to-female ratio of 1:4 is generally recognised[1,2] and the majority of cases occur in childhood. Twenty-five per cent of patients are diagnosed within the first year of life and a further 30 per cent before the age of 10 years.[3]

The classical triad consisting of intermittent abdominal pain, a mass in the right upper quadrant of the abdomen and jaundice, is present only in approximately 38 per cent of cases, but the majority of patients present with at least one of these clinical features. Rarely, the patient may present with biliary peritonitis due to rupture of the cyst. In most instances this apparently occurs spontaneously, but traumatic rupture has been documented, and rupture has also been reported during pregnancy or labour, in approximately 50 patients.[3] The association of this complication of rupture during pregnancy is quite significant in this condition and is presumably due to compression of the cyst as a result of increased intra-abdominal pressure in late pregnancy and labour.

The occurrence of acute cholecystitis, gangrene of the cystic duct and biliary peritonitis in this patient is unusual. The most likely explanation was obstruction of the biliary system due to increased pressure on the choledochal cyst in late pregnancy, or perhaps a result of direct compression of the cystic duct and artery by the cyst.

The pre-operative investigations in an elective patient suspected of having a choledochal cyst and not presenting as an emergency would be an ultrasound of the upper abdomen, percutaneous transhepatic cholangiography and possibly ERCP (endoscopic retrograde cholangiopancreatography). The only acceptable method of treatment of choledochal cyst is surgical. However, the choice of operation between an internal drainage on the one hand and complete excision

and reconstruction on the other remains controversial. Internal drainage is performed either into the duodenum as a choledochocysto-duodenostomy, or into the jejunum as a Roux-en-Y choledochocysto-jejunostomy, the latter being preferable.

The main advantage of internal drainage is the technical ease with which the operation may be performed, and the low operative morbidity and mortality. However, the ineffectual musculature and incomplete endothelial lining of a dilated cyst predisposes to stomal stenosis and resultant ascending cholangitis. The high morbidity and mortality previously associated with primary excision has been reduced by modern techniques, intensive care facilities and antibiotics. Excision also offers the added advantage of the elimination of the small, but real, risk of carcinoma developing in a choledochal cyst.[6] For these reasons, most recent reviews on this subject recommend excision and reconstruction with a Roux-en-Y jejunal loop in elective cases where a pre-operative diagnosis has been made.[1, 3, 7, 8, 9]

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