Congenital choledochal cyst

A case report

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Summary

A congenital choledochal cyst presented as an upper abdominal mass in a non-jaundiced woman; at laparotomy total cystectomy was performed and biliary enteric continuity re-established by a hepatico-jejunostomy Roux-en-Y anastomosis.

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Common causes of upper abdominal cystic lesions include hydatid disease of the liver and kidney and pseudocysts of the pancreas. Although rare, choledochal cysts should be considered in the differential diagnosis of an upper abdominal mass in a young patient.

The clinical features and treatment of a young woman presenting with cystic dilatation of the common bile duct are reported.

Case report

A 27-year-old coloured woman was admitted to Tygerberg Hospital with a triad of upper abdominal pain, weight loss and a palpable abdominal mass in the right hypochondrium. The pain was dull in nature, situated over the liver and radiated through to the back. Periodic vomiting and asthenia were prominent symptoms. She was never jaundiced.

Physical examination revealed a 10 x 10 cm non-tender mass situated in the right hypochondrium in an otherwise healthy female. There were no signs of anaemia or jaundice. Special investigations revealed normal haematological values, moderately elevated liver enzyme and alkaline phosphatase levels in the presence of normal albumin and bilirubin values. Serological tests for syphilis and the *Echinococcus* complement fixation test were negative.

Ultrasonography and computed tomography revealed a large single cystic mass in the liver region. Although the serological tests were negative, hydatid disease of the liver was considered the most likely diagnosis pre-operatively.

At laparotomy a large subhepatically situated choledochal cyst was identified. The liver and adjacent organs were normal. Without much difficulty a total cystectomy with ligation of the distal common bile duct was performed and biliary-enteric continuity was re-established by a hepaticojejunostomy Roux-en-Y anastomosis (Fig. 1). A postoperative technetium radio-isotope liver study showed prompt excretion of the isotope into the Roux loop, indicating a patent biliary-enteric anastomosis. The postoperative course was uneventful and the patient was discharged on the 10th day.

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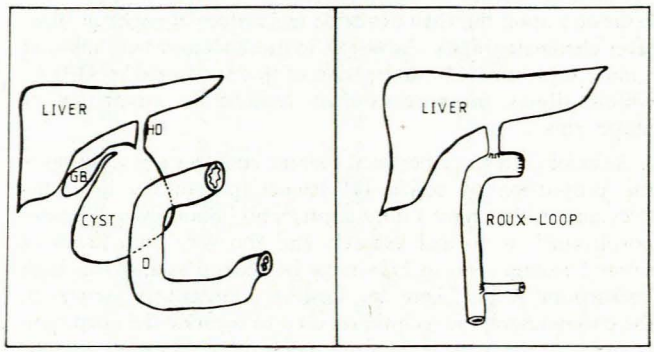

Fig. 1. Operative technique showing extent of the choledochal cyst and biliary-enteric reconstruction with a Roux-en-Y loop (GB = gallbladder; D = duodenum; HD = hepatic duct).

Discussion

The occurrence of cystic dilatation of the common bile duct in surgical practice is extremely rare.[1,2] Although the cause of the condition is unknown, one theory has indicated that the cyst is a congenital malformation which results from an abnormality of epithelial-cell proliferation during development of the bile ducts.

Choledochal cysts have been classified into four types.[1,2] Our patient fits into the type 1 choledochal cyst group, i.e. cystic dilatation of the common bile duct which begins and ends sharply and has a narrowed terminal common duct. This is by far the most frequent form.[1,2] Types 2 - 4 are rare with only 4 cases reported in the literature.

The clinical manifestations of choledochal cyst include abdominal pain, as in our patient, jaundice and a palpable right hypochondrial mass. In adults the manifestations are often intermittent with jaundice, acholic stools and fever more common in infants. Of the reported cases more than 80% have been diagnosed in patients under the age of 30 years with females outnumbering males.

Pre-operatively, oral or intravenous cholangiography, ultrasonic echography, computed tomography and upper gastrointestinal tract contrast studies have aided in confirming the diagnosis; all have been employed with variable degrees of success. The definitive study is operative cholangiography; this is of great value in planning the surgical corrections to be undertaken.[1,2]

Surgical correction is always indicated because of the propensity of the cyst to develop complications and the unusually high incidence of malignant transformation in the cyst or adjacent bile ducts. Where possible, surgical excision is always indicated. The surgical treatment of choice for type 1 cysts is either excision combined with a biliary-intestinal anastomosis as in our case or by anastomosis of the cyst to the intestinal tract.

At present there is no unanimity of opinion regarding the surgical management of choledochal cyst. Most surgeons favour resection but if technical difficulties are encountered a drainage procedure may be achieved either by cystoduodenostomy or cystojejunostomy. In these difficult cases selection of the most

appropriate form of surgical treatment is greatly aided by the information gained by intra-operative cholangiography. The long-term results of surgical correction are good, although secondary operations are occasionally indicated to correct biliary-intestinal anastomotic strictures.

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Cavernous haemangioma of the jejunum

A case report and approach to obscure chronic gastro-intestinal bleeding

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Summary

A 31-year-old woman who had been investigated for almost lifelong iron deficiency anaemia caused by chronic gastro-intestinal blood loss was found to have a large cavernous haemangioma of the jejunum. The literature relating to the latter is reviewed and a diagnostic approach to obscure gastro-intestinal bleeding considered.

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Iron deficiency anaemia due to chronic gastro-intestinal (GI) blood loss is one of the more challenging problems in gastroenterology. Despite intensive investigations employing improved endoscopic, radiological and imaging techniques, the cause of bleeding remains undetected in up to 25% of patients.[1] The site of bleeding should, however, be sought diligently in these patients to exclude surgically remediable lesions. We report on a patient with longstanding occult GI bleeding who appears to have been cured following excision of a cavernous haemangioma of the jejunum which defied detection for many years despite numerous conventional investigations.

Case report

A 31-year-old woman was admitted to hospital for investigation of iron deficiency anaemia, first detected at the age of 4 years and initially well controlled with intermittent iron supplementation. However, in 1973 the anaemia became resistant to iron therapy and the patient subsequently required transfusion of up to 6 units of blood each year. Tests for occult blood in the stools were consistently positive during this period, but repeated upper and lower GI endoscopy, barium enema and barium meal examinations and follow-through studies and isotope scans failed to reveal the source of bleeding.

The patient's only complaint on the present admission was tiredness. There was no history of overt GI bleeding or any other symptoms related to the GI tract. Significant epistaxes were not a feature and there was no history of analgesic abuse. The family history was non-contributory. Examination showed obvious mucosal pallor, several 1 mm red blanching lesions on the hands and a small strawberry naevus on the back. Abdominal examination was negative but the stools were again positive for occult blood.

A full blood count revealed hypochromic, microcytic anaemia (haemoglobin value 8,3 g/dl). There were no stool parasites. Upper GI endoscopy and colonoscopy were negative. A technetium-99m-pertechnetate scan showed a persistent blush in the area of the uterus. Small-bowel barium enema examination (enteroclysis) demonstrated a strikingly abnormal 5 - 10 cm segment of small bowel in the pelvis surrounded by a halo of phleboliths (Fig. 1). Although the abdominal angiogram was normal, the barium contrast study and isotope scan suggested a vascular abnormality, and laparotomy was performed. A large cavernous haemangioma measuring 25 cm was present in a loop of distal jejunum lying at the pelvic brim (Fig. 2). This was resected and an end-to-end jejunal anastomosis performed. Scattered throughout the proximal jejunum were 20 - 30 dilated veins each slightly larger than a pinhead. Their significance was uncertain. A second 20 x 25 cm cavernous haemangioma in the retroperitoneal tissue over the right pelvic brim was not resected. A barium-injected radiograph of the surgical specimen revealed vascular lakes surrounded by phleboliths (Fig. 3).

Histologically the lesion was a cavernous haemangioma involving the muscularis and submucosa of the jejunum and communicating with the lumen. The patient has remained well with a stable haemoglobin value 8 months after the operation.

Discussion

This case demonstrates a number of interesting points related to obscure GI bleeding. The length of time before a diagnosis

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