Choledochal cysts—report of three cases and review

R. WALDRON
F.R.C.S.I.

C. F. MCCARTHY
F.R.C.P., F.R.C.P.I., M.D.

J. DRUMM
F.R.C.S.I., F.R.C.S.

B. MURPHY
F.R.C.S.I., F.R.C.S., F.A.C.S.

University Department of Surgery and Medicine, Regional Hospital, Galway, Ireland

Summary

Three cases of choledochal cyst are presented, with a review of previous publications. We re-emphasize the diagnostic dilemma involved and discuss current surgical management. A new complication is illustrated and an association with polycystic kidneys is reported.

Choledochal cysts are uncommon, but correct management ensures a symptom-free future for the patient. Recent reports support cyst excision if technically feasible.

KEY WORDS: polycystic renal disease, choledochocyst-jejunostomy, cholelithiasis.

Introduction

Choledochal cyst is an abnormal cystic dilatation of the biliary tract. This was alluded to by Vater in 1720 but Douglas in 1852 was the first accurately to describe the anomaly. Recently with improved biliary tract diagnostic methods choledochal cysts are being recognized with increasing frequency.

Case 1

A 3-year-old female was admitted to hospital with a transient episode of jaundice in 1968. One year later, at exploratory laparotomy for diagnosis of a right upper quadrant mass, a large cystic dilatation of the common bile duct was exposed and a choledochocyst-duodenostomy performed. Follow-up cholangiograms revealed dilatation of the biliary ducts, due to ascending infection, which lead to numerous hospital admissions over the ensuing years.

In July 1976 the choledochocyst-duodenostomy was taken down and a Roux-en-Y choledochocyst-jejunostomy fashioned. The patient did well post-operatively and remains symptom free 7 years later.

Case 2

A 31-year-old female was admitted to hospital in January 1977, with biliary colic and obstructive jaundice. At laparotomy, a cholecystectomy, with a negative duct exploration, was performed. Two operative cholangiograms were reported as normal. A T-tube cholangiogram was performed 10 days post-operatively and was also normal. The T-tube was then removed. Twenty-four hours later she developed ascending cholangitis. A transhepatic cholangiogram demonstrated three calculi in the common bile duct. At repeat laparotomy, a cystic-type sac was resected and the ductal calculi extracted. Histologically, this sac was composed of biliary ductules. Six years later, the patient remains well.

Case 3

A 31-year-old male, presented in February 1980, for the evaluation of three episodes of jaundice over a 10-year period. A left flank mass was noted on examination and intravenous urography confirmed the presence of bilateral renal polycystic disease. Endoscopic retrograde cholangio-pancreatography demonstrated cystic dilatation of the intra- and extra-hepatic biliary tree with a normal pancreatogram. This man declined further assessment and has been lost to follow-up.

Discussion

The incidence of choledochal cysts is unknown; large series quote an incidence of approximately one in thirteen thousand births, 60% of recorded cases being from Japan (Jones *et al.*, 1971). Females predominate in a ratio of 4:1, most presenting early in life, but no age is exempt the oldest patient recorded being a 78-year-old woman (Madding, 1961). Alonso-Lej, Rever and Pesagno (1959) postulated that the majority are congenital in origin.

Correspondence: B. Murphy.

Further support is given to this theory by the high incidence of associated congenital anomalies of the biliary tract (Komi *et al.*, 1977). Longmire, Mandiola and Gordon (1971) described four types of choledochal cyst, on a clinico-pathological basis (Fig. 1).

FIG. 1. Types of choledochal cysts (Longmire *et al.*, 1971).

Type 1: Spherical dilatation of the common bile duct—comprises the majority of cases. Case 1 belongs to this group.

Type 2: Congenital diverticulum protruding from the lateral wall of the common bile duct. This variety is rare, only 14 cases to date being documented. The second case report belongs to this type.

Type 3: Choledochoceles, a dilatation of the intra-duodenal portion of the common bile duct. Only nine cases have been reported to date (Olbourne, 1975).

Type 4: Multiple communicating cystic dilatations of the intra- and extra-hepatic biliary tree, was originally described by McWhorter (1924), attributed to Caroli, Soupault and Kossakiwski (1958), and designated as Type 4 choledochal cyst by Longmire *et al.* (1971); a total of 45 such cases have been described to date. This was the abnormality in Case 3. We have not found any report of the association of cystic dilatation of the common bile duct with polycystic kidneys previously reported in the literature.

The typical macroscopic and microscopic features of choledochal cyst have been extensively reviewed by Mahour and Lynn (1969). The cyst wall usually consists of inflammatory and fibrous tissue with no proper epithelial lining; the cyst itself usually contains infected bile, with occasionally biliary sediment and gallstones (Kune, 1965).

The preoperative diagnosis of choledochal cyst has proved to be an elusive goal in most cases, due to the rarity of the condition and the variability of the clinical manifestations. The classical triad of vague, intermittent upper abdominal pain, intermittent jaundice and a right upper abdominal mass is rarely seen. Radiological studies are the most helpful investigative procedure. Intravenous cholangiogram in the jaundiced patient may outline the cystic mass. Grey scale ultra-sound may prove to be a very effective diagnostic agent but has not as yet been used widely for this problem. Operative cholangiogram at exploratory laparotomy most often provides the definitive diagnosis.

The complications of choledochal cyst are grave and stress the importance of early corrective management. Recurrent ascending cholangitis with eventual secondary biliary cirrhosis can result and may develop postoperatively if the cyst has not been resected (as in Case 1). The risk of carcinoma, developing in the cyst wall, is markedly higher than that of the general population. A total of 63 cases have been recorded to date, an incidence of 4.7% (Iodaniti *et al.*, 1979). The overall risk of biliary tract carcinoma varies from 0.007% to 0.041%, whereas it appears to be 2% in association with choledochal cyst (Longmire, McArthur and Bastounis, 1973). We report here a further complication, namely that of stone lodgement in the neck of the cyst which may be missed at routine cholecystectomy (as in Case 2), even with preoperative cholangiograms, unless the biliary surgeon is aware of the possibility. Calculi within the cyst are an infrequent occurrence. Scharli and Bettex (1968) reviewing previous publications found only 15 such cases.

Conservative medical treatment is attended by a mortality approaching 100% (Atter and Obeid, 1955). The main controversy concerns the type of surgical management, whether internal drainage or excision is the treatment of choice. Most observers agree that cyst excision should be performed when technically feasible, taking into account the size of the cyst, adjacent inflammatory change and the general condition of the patient (Yue, 1974). McWhorter (1924) first advocated cyst excision and this more radical procedure has gained increasing popularity of late (Flanigan, 1975). After excision of the cyst continuity is re-established by either a choledochal-jejunostomy, a choledochal-duodenostomy or a choledochal-choledochostomy. Lilly (1979) has described a technique

of cyst excision whereby the serosa of the posterior wall remains intact, thus avoiding damage to the portal structure.

Internal drainage via a choledochocystenterostomy is the major alternative operation. Choledochocystgastrostomy is rarely used due to the associated high mortality (Alonso-Lej *et al.*, 1959). Choledochocystduodenostomy is a technically easy procedure, but carries a high risk of subsequent cholangitis secondary to reflux (Kirwin, 1974). Trout and Longmire (1971) demonstrated a 65% incidence of anastomotic complications during the first 5 years post-operation, 40% needing a revision operation over 5 years. Case 1 confirms these findings, her post-operative course being marred by recurrent episodes of ascending cholangitis.

Choledochocystjejunostomy is the most satisfactory of the internal drainage procedures, especially if a Roux-en-Y, when a large side-to-side anastomosis to the most dependent part of the cyst is made, is used, as advocated by Atter and Obeid (1955).

Spitz (1978) supports choledochocystjejunostomy as the primary method of treatment reserving excision for those patients who have complications. With Caroli's disease the extra-hepatic biliary cyst is treated as discussed above, and the intra-hepatic biliary cysts are only treated if complications arise, when excision or drainage may be necessary (Powell, Sawyers and Reynolds, 1981).

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