Biliary ductal and vascular anomalies associated with choledochal cyst

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ABSTRACT

Nineteen patients operated for choledochal cyst over a 7-year period were reviewed retrospectively. Cyst excision and Roux–en–Y hepaticojejunostomy was done in all patients but one; one patient was managed with cyst excision and an antiperistaltic hepatico-appendico-duodenostomy. About one-third of patients (6/19) had biliary ductal/vascular anomalies identified at surgery. These included anomalous right hepatic artery (n = 3), primary ductal stricture (n = 2), and aberrant right hepatic duct (n = 1). A high degree of awareness of biliary ductal and vascular anomalies is required on the part of the operating surgeon to facilitate accurate intraoperative diagnosis as well as successful reconstructive surgery.

KEY WORDS: Choledochal cyst

Biliary duct anomalies are well known; they often result in significant intraoperative complications and postoperative morbidity if unrecognised at the time of surgery. Recently, a few authors have noticed a high degree of association between biliary anomalies and choledochal cysts. A retrospective analysis of records of 19 patients operated for choledochal cysts over a period of 7 years (April 1998–March 2005) was done to determine the incidence of biliary duct and vascular anomalies in association with choledochal cysts.

MATERIALS AND METHODS

The records of 19 patients operated for choledochal cysts during 7-year period were reviewed retrospectively; two patients who were diagnosed to have forme fruste choledochal cyst have not been included in this series. There were 11 females and 8 males. Their ages ranged from 4 months to 13 years with five of them presenting in infancy, five presenting in the age group 1–5 years, another five presenting between 5 and 10 years of age and four presenting above 10 years of age. Abdominal pain was the commonest complaint and was present in 79% (15/19) of the patients; jaundice was seen in 63% (12/19) of the patients, whereas a palpable abdominal lump was noticed only in 21% (4/19) of the patients. None of the patients had the classical triad of symptoms.

Four out of the nineteen patients (21%) had significant derangement of liver functions. Two of these four patients belonged to the infantile variety. One of these was later found to have a ductal stricture proximal to a fusiform choledochal cyst. This patient had severe hepatic cirrhosis with moderate ascites and was initially thought to have biliary atresia. The other two patients had history of repeated episodes of cholangitis; both of these had hepatomegaly with dilatation of intrahepatic biliary radicals on abdominal ultrasonography. One of these children was diagnosed to have primary stricture of both right and left hepatic ducts on ERCP.

All these patients had abdominal sonography as their primary imaging. Six patients with age ranging 1.5–13 years had preoperative ERCP; the abnormalities recognised included primary ductal stricture (n = 1), type 4 choledochal cyst (n = 1), and calculi in choledochal cysts (n = 2). MRCP has been performed in two patients later in the series to confirm the diagnosis. A long pancreatico-biliary common channel was, however, not demonstrated in any of the cases; the gastroenterologist and the radiologists probably did not look specifically for this anomaly. All patients but three were managed by cyst excision and Roux–en–Y hepaticocho-jejunostomy. One patient required hilar anastomosis due to presence of primary ductal stricture of both right and left hepatic ducts. Another patient with Alanso et al. type 4-choledochal cyst required left lobectomy and right hepatico-jejuno-stomy. Another patient was managed with cyst excision
RESULTS

Associated biliary/vascular anomalies were found in close to one-third (32%) of the patients. Primary ductal stricture was seen in two patients, aberrant right hepatic duct was seen in one patient, and anomalous right hepatic artery was found in three patients. All but one of these anomalies were not known preoperatively and were diagnosed intraoperatively. One patient with anomalous right hepatic artery had excessive intraoperative haemorrhage due to injury to this vessel, which was repaired using 8-0 Gore-Tex using operating loop. The patient with aberrant right hepatic duct also sustained injury of the anomalous duct; which was diagnosed and managed intraoperatively. Postoperatively this patient had biliary leak, which responded well to percutaneous drainage.

One patient had massive blood loss of an unsuspected bleeding diathesis (due to wrong preoperative laboratory reports) intraoperatively and died immediate postoperatively of shock and DIC. Another patient, who had infantile variety choledochal cyst with proximal ductal stricture, died of bronchopneumonia 2 months after surgery. One patient needed re-exploration, revision anastomosis, flexible choledochoscopy and removal of intra-hepatic biliary sludge and calculi, 4 years after the initial surgery; since then the patient has been doing well. Other 16 patients have been doing well with no clinical or biochemical evidence of cholestasis. In all, 14 patients have had a long-term follow-up ranging from 6 months to 6 years.

DISCUSSION

Many authors agree that ‘normal biliary anatomy’ is seen in less than 50% of patients undergoing biliary tract procedures. The various anomalies mentioned in the literature include primary ductal stricture, aberrant hepatic duct, anomalous hepatic arteries, low confluence of hepatic duct and a very small bile duct.

Primary Ductal Strictures often require hilar Anastomosis, redo surgeries due to persistence of symptoms because of missed proximal stricture or anastomotic strictures, and ductoplasties. Todani et al. have described primary ductal strictures at the hilum, at the confluence of right and left hepatic ducts, and at the junction of hepatic duct with cystic duct. They have classified these strictures as simple strictures and membranous strictures. In their series, one patient had a left hepatic duct stricture, which was ignored initially. Postoperatively, the patient developed recurrent episodes of cholangitis for which the patient was investigated and re-explored 15 years after the primary surgery. He underwent excision of the atrophic left lateral segment.

Coexistence of a type 1 infantile variety of choledochal cyst with distal biliary stricture has been frequently mentioned in the literature, but association of ductal stricture proximal to choledochal cyst has not been reported hitherto. One such case was treated; he was misdiagnosed as biliary atresia preoperatively.

An aberrant hepatic duct (AHD), although rare, is a significant accompaniment to choledochal cysts. Todani et al. have suggested anastomosing these with one of the main ducts and then performing hepaticojejunoanostomy, while Narsimhan et al. believed in creating a separate biliary enteric anastomosis with the aberrant hepatic duct. Awareness of this entity is important in order to avoid intraoperative injury or accidental ligation. When identified, they should be included in the anastomosis. If the AHD is damaged or goes unrecognised at surgery, postoperative bile leak may result in biliary peritonitis, biliary fistula, or abscess formation. If the AHD is accidentally ligated and remains unrecognised, it may result in infection of the lobe and subsequent atrophy of the obstructed segments with compensatory hypertrophy of the unaffected segment. Ligation of small sized AHD may be a viable option, however, in large AHDs, reconstructive surgery is recommended by most.

Caution to identify the presence of anomalous arterial supply cannot be overemphasized, since it is the only way to avoid injury or accidental ligation – both of which can result in a major surgical disaster. The aberrant artery running in front of the common hepatic duct can be easily displaced behind the jejunal loop at the time of reconstruction.

To conclude, biliary and vascular anomalies are frequently seen in association with choledochal cysts. They have tremendous implications as they are seldom diagnosed preoperatively, cause significant intraoperative complica-
tions and require a high degree of awareness on part of the operating surgeon, to facilitate accurate intraoperative diagnosis as well as successful reconstructive surgery.

REFERENCES