

Cholangitis and Choledocholithiasis after Repair of Duodenal Atresia: A Case Report

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Abstract- Cholelithiasis is a rare but known complication of surgery for duodenal atresia. Occurrence of choledocholithiasis as sequelae of duodenoduodenostomy is still rarer. Biliary stasis resulting from compression of common bile duct due to periductal fibrosis may predispose to gallstone formation. We are reporting a case of choledocholithiasis in a 6 year old child as a late post-operative complication of duodenoduodenostomy (for duodenal atresia in the neonatal period). To the best of our knowledge this is the first case of its kind reported in English literature. Cholecystectomy followed by choledocholithotomy was done and the patient had an uneventful recovery. Upper abdominal pain in any patient with a history of surgery for duodenal atresia in the past warrants a thorough evaluation for any biliary tract anomaly, cholecystitis, cholangitis, cholelithiasis or choledocholithiasis.

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Introduction

Cholelithiasis in children is an unusual but not an exceptional finding, and is associated with nonspecific symptoms. In contrast to adults, little is known about its epidemiology in children. The etiology may be unknown or may be related to risk factors, including hemolytic conditions, sepsis, hyperalimentation, trauma, antibiotics, immobilization, short bowel disease, inflammatory bowel disease, and cystic fibrosis etc. Cholelithiasis may occur rarely as a complication of surgery for duodenal atresia (1,2). Cholelithiasis in the first years of life is correlated with malformative, pharmacologic or iatrogenic predisposing factors. Pediatric cholelithiasis beyond the first year of life, especially in the later childhood and adolescence, can be similar to cholelithiasis of adults for epidemiology, pathogenesis, symptomatology and therapy (3).

Gallstones may migrate from gallbladder to the ductal system causing choledocholithiasis and are termed as secondary bile duct stones. Primary bile-duct stones that form within the ductal system itself are more common in East Asian countries than in the Western world. Thus pathogenesis of primary and secondary bile-duct stones is unlikely to be similar (4). We are

reporting a case of choledocholithiasis occurring after duodenoduodenostomy for duodenal atresia. To the best of our knowledge this is the first case of its kind reported in English literature (with extensive search in Pubmed revealing no such report).

Case Report

A 6 years old male child presented with pain in upper abdomen for 15 days which was more marked on right side. It was colicky in nature to begin with and later became constant. Pain was associated with high grade fever with chills and rigors along with bilious vomiting. Five days later he developed progressively increasing jaundice with passage of clay colored stools. There was a history of such episodes of colicky pain in right upper abdomen for the last 6 months which had no aggravating factor and were relieved only on taking antispasmodic medications.

The patient had been delivered preterm by caesarian section for fetal distress and polyhydramnios. At 5 days of age he had undergone exploratory laparotomy with duodenoduodenostomy for duodenal atresia type 1.

On examination the patient was found to have deep icterus.



Figure 1. USG showing choledocholithiasis with dilated common bile duct and distended gallbladder

His pulse rate was 120/min, blood pressure was 100/78 mmHg, and temperature was 38.3°C. Tenderness was present in right hypochondrium. Liver was palpable 2cm below the right costal margin with a span of 10cm. A mobile globular lump of size 5×5 cm was palpable along the lower border of liver. There was no splenomegaly or ascites. On investigation liver function tests were found to be suggestive of obstructive jaundice (total serum bilirubin was 5.5mg/dl with a direct fraction of 4.0mg/dl and indirect fraction of 1.5mg/dl, serum alkaline phosphatase was 2870 KA unit/100ml). There was no clinical or laboratory evidence of hemolytic disease. Ultrasound abdomen showed distended gallbladder with normal wall thickness and no pericholecystic fluid. Distal common bile duct had an intraluminal echogenic focus of size 14×6 mm with posterior acoustic shadow and dilatation of proximal common bile duct (internal diameter up to 17mm) (Figure 1).

Supportive treatment including intravenous fluids and injections cefotaxime and amikacin was started. Since facilities for endoscopic retrograde cholangiopancreatography were not available in the

hospital, the patient was subjected to exploratory laparotomy which revealed adhesions between parietes, gallbladder, extrahepatic biliary tree, duodenum and jejunum. On adhesiolysis gallbladder was found to be markedly distended with dilated proximal common bile duct and extra-hepatic ducts.

Common bile duct exploration revealed a single friable yellowish-brown stone of size 15×6 mm obstructing the distal part.

Cross section of the stone revealed alternate light and dark bands. Cholecystectomy followed by choledocholithotomy was done and bile duct was closed over a T-tube.

Per-operative cholangiogram after adhesiolysis and removal of stone showed normal passage of contrast through the T-tube into the biliary channels and duodenum with no mucosal irregularity, filling defect or stricture (Figure 2). Bile culture later revealed growth of *Escherichia coli*. Histopathological examination of gallbladder showed features of chronic cholecystitis with superimposed acute changes. T-tube was removed on tenth postoperative day and the patient had an uneventful recovery.



Figure 2. Per-operative cholangiogram after adhesiolysis and removal of stone showing normal passage of contrast through the T-tube into the biliary channels and duodenum with no mucosal irregularity, filling defect or stricture

Discussion

Duodenal atresia and stenosis is a frequent cause of congenital, intestinal obstruction (5). The condition may be associated with anomalies of biliary system (1,2,6-9). In children with duodenal atresia, associated abnormalities of the biliary tract have been implicated in the development of cholelithiasis which may occur several years after successful relief of the duodenal obstruction by some form of by-pass procedure (1,2). In a case of operated congenital duodenal atresia, choledochal cyst with intracystic stone was detected at the age of 27 years (6). However, intraoperative cholangiogram in our patient carried out after adhesiolysis and removal of stone did not reveal any biliary tract anomaly.

The stone in our patient was friable and yellowish-brown indicating that it was a primary bile duct stone probably formed secondary to biliary stasis and associated cholangitis. Such stones are brown pigment stones with an earthy texture, ovoid shape and fragile structure, with alternating light and dark brown pigmented layers on cross-section. Chemically these stones contain low levels of cholesterol, with high levels of bilirubin and calcium salts. Bile duct bile of patients

with primary choledocholithiasis was always moderately to profusely infected (10).

After major abdominal operations reductions in bowel motility, cholecystokinin release and vagal activity could all lead to gall bladder hypotonicity and biliary stasis predisposing to gall stones (11). Biliary stasis due to compression of common bile duct from fibrosis in hepato-duodenal area may also predispose to gallstone formation (2). Moreover dragging of sphincter of Oddi by postoperative adhesions results in its hypomotility leading to reflux of duodenal fluid into the biliary duct and pancreatic duct. Gallstones may form when duodenal juice was added to lithogenic bile (12). Duodenal transection has also been reported to produce significant changes in interdigestive sphincter of Oddi motility, possibly contributing to augmented duodenobiliary reflux and then lithogenesis. Myoneural continuity between the stomach and sphincter of Oddi at the proximal duodenum may play an important role in maintaining normal biliary dynamics (13).

In disorders of biliary motility and associated bacterial infection, bacteria producing slime and those containing the enzyme glucuronidase cause enzymatic hydrolysis of soluble bilirubin glucuronide to form free bilirubin which then precipitates with calcium (14).

Escherichia coli infection documented in our case has been incriminated as the most common organism in the causation of cholangitis in patients without biliary stent (15). It is hypothesized that in our patient choledocholithiasis could probably be attributed to an interplay of factors like compression of common bile duct from post-operative adhesions (resulting from surgery for duodenal atresia), cholangitis and altered biliary dynamics. Thus, upper abdominal pain in any patient with a history of surgery for duodenal atresia in the past warrants a thorough evaluation for any biliary tract anomaly, cholecystitis, cholangitis, cholelithiasis or choledocholithiasis. Endoscopic ultrasound which is an important diagnostic tool for gastrointestinal diseases in adults, has rather limited utility in children. Some authors, however, feel that it is an effective tool in the evaluation of pediatric gastrointestinal patients, mostly with pancreatobiliary disorders (16). The overall sensitivity of this procedure in detecting choledocholithiasis has been reported to range from 93 to 100% (17). The technological accuracy of magnetic resonance cholangiopancreatography (MRCP) rivals that of endoscopic evaluation (18,19). The diagnostic accuracy of MRCP in patients with choledochal cysts and stenoses has been reported to be 100% (20). Whereas its sensitivity and specificity in detecting choledocholithiasis has been reported to be 88% and 96.8%, respectively (21). A new generation of magnetic resonance imaging scanner offers a 3-dimensional magnetic resonance cholangiopancreatography with very high spatial resolution using a non-breath-hold technique and allows an excellent demonstration of all relevant pancreaticobiliary ducts and pathologic findings in children (22).

The role and value of endoscopic retrograde cholangiopancreatography (ERCP) in the pediatric age group is not well established (23). Besides being an operator-dependent and invasive procedure, it has its own complications and limitations. However, it has been reported to have a high success rate even in children provided it is performed by experienced endoscopists. The delicate delineation of the anatomy by ERCP and its therapeutic potential make it absolutely superior to other less invasive tools such as MRCP (24). Some studies, on the other hand, concluded that ERCP in the pediatric population had a much higher complication rate as compared to adults (33.3%) even in experienced hands, and advocated that the risk and benefits should be carefully reviewed before proceeding (25).

The management of symptomatic or incidentally discovered common bile duct (CBD) stones is still

controversial, the best practice being still unknown. A recent NIH statement on ERCP showed that both ERCP and laparoscopic CBD clearance were safe and reliable to clear stones (26,27). The sequential approach, combining endoscopic retrograde cholangiopancreatography (ERCP) with endoscopic sphincterotomy (ES) prior to or after cholecystectomy, was for a long time considered to be the preferable choice for choledocholithiasis and even today many gastroenterologists, endoscopists and surgeons still prefer it in clinical practice. Late complications (stone recurrence and/or cholangitis, and acute cholecystitis) have been reported in approximately 12% of patients after ES (28). Some authors suggest primary treatment of choledocholithiasis by laparoscopic approach in children. In case of residual obstruction, a postoperative ES could be performed (29). This technique had lower morbidity and mortality rates compared to preoperative ERCP/ES in the management of patients with suspected CBD stones even if the chance of CBD stones reached 100% (30).

Some others were of the view that ERCP should be the treatment of choice in children with CBD stones who were going to have or had previously undergone laparoscopic cholecystectomy (23). Some authors recommend that intraoperative cholangiography should be routinely performed in children before the use of ERCP to avoid unnecessary ERCP unless CBD stones are specifically visualized by ultrasound scan (31). The "rendezvous" (RV) procedure which combines laparoscopic cholecystectomy, intra-operative cholangiography, and endoscopic bile duct clearance carries interesting advantages over sequential treatment but is more complex to organize as it calls for synchronized collaboration between surgeon and endoscopist (32). It also solves problems at the papilla of Vater that are the main causes of retained stones and recurrence. Simple mechanical lithotripsy (ML) may be used to crush difficult bile duct stones and the usual success rate reported ranges from 51% to 100%. Factors like an oversized stone and presence of distal CBD stricture have been reported to account for failure of ML (33). Electrohydraulic lithotripsy (EHL), extracorporeal shock wave lithotripsy (ESWL), and laser therapy may also be used for the treatment of difficult bile duct stones. Frequency Doubled Double Pulse Nd:YAG (FREDDY) laser is a newly developed economical, short-pulse, double frequency, solid-state laser that causes less tissue damage (34). Thus, in the absence of any consensus guidelines on management of choledocholithiasis in children, options vary from purely

surgical to non-surgical procedures depending upon etiology, individual surgical preferences, and availability of techniques and expertise.

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Cholangitis and choledocholithiasis after repair of duodenal atresia

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