Acute Pancreatitis as a Complication of Choledochal Cyst

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ABSTRACT

Choledochal cysts (CCs) are rare congenital disorders of the biliary tree which are associated with biliary tract and pancreatic complications. Its etiology is still unknown, but some evidences suggest that CCs are caused by infection during embryogenesis. The laboratory evaluation reveals abnormalities due to pancreatobiliary complications, such as ascending cholangitis, cholecystitis, and pancreatitis. Radiologic examination may delineate the structural abnormalities occured in CCs and confirm the diagnosis. The major mortality is caused by cholangiocarcinoma. This case discussed the diagnosis and treatment of a 20-year-old male with choledochal cyst and its complications. Management consists of therapy on complication and definitive therapy.

***Keywords**: choledochal cyst, acute pancreatitis, cholecystitis*

ABSTRAK

Kista duktus koledokus merupakan kelainan kongenital traktus biliaris yang jarang dan umumnya berhubungan dengan komplikasi pada traktus biliaris dan pankreas. Etiologinya sendiri belum diketahui secara jelas, namun beberapa bukti menunjukkan bahwa kista duktus koledokus disebabkan oleh infeksi saat proses embriogenesis. Pada hasil pemeriksaan laboratorium menunjukkan adanya kelainan yang disebabkan oleh komplikasi pankreatobiliar seperti kolangitis asendens, kolesistitis dan pankreatitis. Pemeriksaan radiologi dapat menunjukkan kelainan struktural yang terjadi pada kista duktus koledokus untuk menegaskan diagnosis. Penyebab mortalitas terbesar disebabkan oleh kolangiokarsinoma. Pada kasus ini dibahas mengenai diagnosis dan tatalaksana pasien laki-laki berusia 20 tahun dengan kista duktus koledokus dan komplikasinya. Tatalaksana yang dilakukan meliputi terapi terhadap komplikasi dan terapi definitif.

***Kata kunci**: kista duktus koledokus, pankreatitis akut, kolesistitis*

INTRODUCTION

Choledochal cysts (CCs) are congenital anomalies of the biliary tract that are manifested by cystic dilatation of the biliary tree involving extrahepatic, intrahepatic bile ducts, or both.[1] CCs are presented in infants and children; adult presentation is rare and usually associated with biliary tract and pancreatic complication. Secondary pancreatobiliary complications in adults may obscure the primary condition. Severe pancreatobiliary complications in young adults should warrant further investigation for clinical suspicion of CCs.[1,2] This case report would discuss problems in diagnosing and treating choledochal cysts and its complications in 20-year-old male.

CASE ILLUSTRATION

A 20-year-old male came to the emergency unit of Sanglah Hospital complained of continuous stabbing epigastric pain which referred to his back since one week before hospital admission. He had consumed magnesium hydroxide and simethicone and did not feel

improvement on his abdominal pain. He felt that his abdominal pain was worsening, combined with nausea and five times vomiting since three days before hospital admission. Patient also complained of fever since one day before, accompanied by yellowish eye. The urine was tea-colored yellow and had history of pale color of defecation. Since six years ago, the patient had been experiencing severe abdominal pain and jaundice for four times and had been admitted in other hospital. There was no history of chronic viral hepatitis as a risk factor. The patient also had no history of spontaneous bleeding, alcohol consumption, out of town travelling, or abdominal surgery.

On admission, the patient's axillar temperature was measured high with slight tachycardia. The conjunctivas were normal with icteric scleras. The heart and lung were normal. Bowel sound was heard with a normal frequency. Abdomen was flexible, without any signs of generalized peritonitis and with non-palpable liver and spleen. There was Murphy sign and abdominal tenderness on right upper quadrant. No shifting dullness was found, as well as no stigmata of liver cirrhosis, edema of extremity, or flapping tremor.

Result of complete blood count was hemoglobin (Hb) 14.1 g/dL, hematocrite (Ht) 43.6%, leukocytes 13,300/mm^3, and thrombocytes 324,700/mm^3. In addition, result of differential count was dominant for neutrophils. The morphology of erythrocyte, leukocyte and thrombocyte was normal. From electrolyte examination, it was found that sodium 137 mEq/dL, potassium 3.5 mEq/L. Other results were serum aspartate transaminase (AST) 139.7 u/L, alanine aminotransferase (ALT) 159.8 u/L, ureum 18 mg/dL, creatinine 0.8 mg/dL, random blood glucose 93 mg/dL. Result of bilirubin was 4.76 mg/dL with dominance of direct bilirubin 3.71 mg/dL. Alkaline phosphatase was 217 U/L with gamma-glutamyl transpeptidase (γGT) 762 U/L. Amylase level was also increased (2,744 U/L). The laboratory showed no evidence of hepatitis A or B infection.

An abdominal ultrasound result indicated dilatation of intrahepatic and extrahepatic bile duct, the gallbladder was distended with a thickened gallbladder wall and contained gallbladder sludge (Figure 1). An abdominal computed tomography scan (CT-scan) showed intrahepatic and extrahepatic choledochal cyst with focal stricture in distal common bile duct without stone, space occupying lesion (SOL), or nodule. The gallbladder diameter was 10.4 cm with a thickened gallbladder wall and double layer sign. The pancreatic duct was dilated with diameter of 4.6 mm. It was connected to cystic mass which continued to choledochal duct (Figure 2).

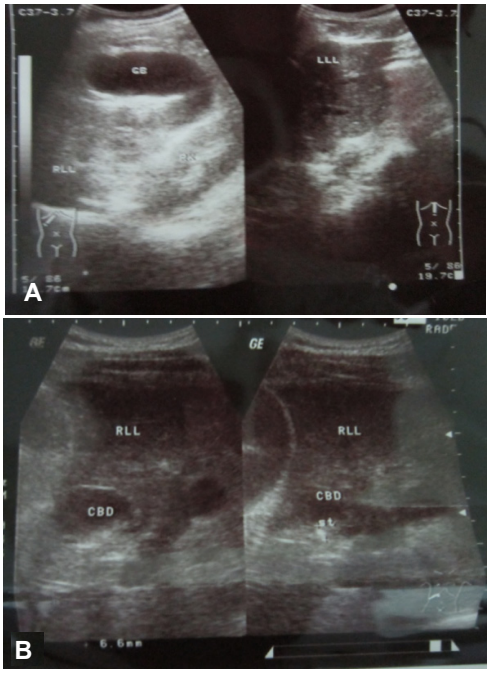
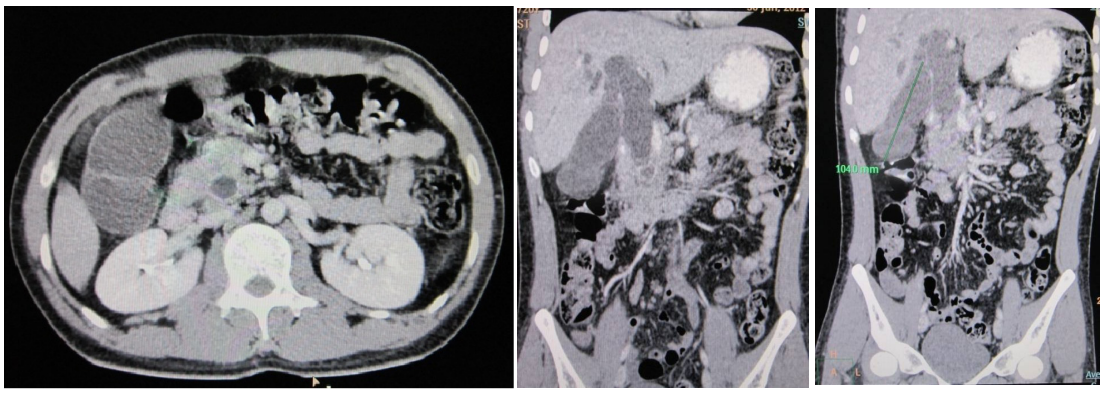


Figure 1. Abdominal ultrasound showed dilatation of intrahepatic bile duct and a distended gallbladder contained gallbladder sludge (A) and dilatation of extrahepatic bile duct (B)



A **B** **C**

Figure 2. Abdominal scans show dilated pancreatic duct (diameter 4.6 mm) with focal stricture (A and B) and cystic structure in distal portion of choledochal duct (diameter 12.6 mm) and enlarged gallbladder (long axis 104 mm) (C)

The final diagnosis were acute cholecystitis and acute pancreatitis caused by choledochal cyst. Fluid rescucitation was given initially due to profuse vomiting. Patient was fasted to limit pancreatic secretion. Other treatment included 40 mg intravenous omeprazole twice daily, 15 mL oral sucralfate four times daily, 250 mg oral ursodeoxycholic acid (UDCA) three times daily, 2 g intravenous ceftriaxone twice times daily for 14 days, and intravenous somatostatin therapy. After treatment, patient experienced clinical improvement with decreased intensity of abdominal pain and was planned to undergo definitive treatment by digestive surgeon, including laparotomy, common bile duct (CBD) exploration, cyst excision, and biliary reconstruction. Later, patient refused to undergo surgery due to improvement of clinical symptoms. After two weeks of treatment the bilirubin level was decreased with resolution of liver function test. The patient was discharged in a good condition after improvement of pancreatobiliary infection with given therapy.

DISCUSSION

CCs are congenital dilatations of the extrahepatic and/or intrahepatic biliary tree that may result in significant morbidity, unless identified early.[1,3] The diagnosis was made on the basis of disproportional dilatation of the bile ducts after excluding the possibility of tumor, stone, or inflammation as the cause of dilatation.[4] In Western countries, these conditions are rare but not as rare as in Japan and China.[1,2,5] Reported frequency rates range from 1 case/100,000-150,000 in the West.[1,6,7]

CCs are common among females, with a female to male ratio of 3 : 1 to 4 : 1.[1,7] Most patients are diagnosed during infancy or childhood, although CCs may be discovered at any age. CCs in adults are often associated with complications. The incidence of cyst-related complications has been reported more in adults compared to children; up to 80% experience recurrent cholangitis and pancreatitis.[8-10]

CCs were classified into 5 types by Todani et al.[11] Type I is the most common CCs (80-90%) involving saccular or fusiform dilatation of a portion or entire common bile duct (CBD). Type II is an isolated diverticulum protruding from the CBD. Type III or choledochocele arise from dilatation of duodenal portion of CBD. Type IV characterized by multiple dilatations involving the extrahepatic and intrahepatic bile ducts. Type V or Caroli's disease is a cystic dilatation of intrahepatic biliary ducts.[11]

The etiologies of CCs are not exactly known and remain speculative.[1,10,12] The most widely accepted hypothesis is the long common channel theory proposed by Babbitt. The condition predisposes to reciprocal regurgitation of both ducts. The combination of pancreatic enzymes and bile acids could induce pancreatitis and inflammatory changes in the bile duct, predisposing to abnormal dilatation.[12,13]

Symptoms of CCs are usually associated to pancreatobiliary complications. CCs cause proximal bile stasis, which in turn leads to stone and sludge formation. These factors lead to ascending cholangitis and further obstruction, resulting in the classic symptoms of episodic abdominal pain, fever and obstructive jaundice. Stone and protein plug formation in the distal common bile duct and pancreatic duct causes obstruction and resultant pancreatitis.[8] Biliary stasis and presence of an anomalous pancreatobiliary duct junction in CCs were the pathomechanism that can explain pancreatobiliary complications, such as acute recurrent pancreatitis, ascending cholangitis, cholecystitis, biliary stricture, biliary cirrhosis, and cholangiocarcinoma.[12]

In this patient, the symptoms were continuous stabbing epigastric pain referred to his back, fever, nausea, profuse vomiting, and yellowish eye with history of previous multiple hospitalization due to the same complaints. Physical examination showed high axillar temperature, icteric scleras, right hypochondrium tenderness and positive Murphy sign. Laboratory results revealed elevated liver function, i.e. hyperbilirubinemia with dominance of direct bilirubin, elevated alkaline phosphatase, γ-GT, transaminase, and hyperamylasemia. The patient had ultrasonography which showed dilatation of intrahepatic and extrahepatic biliary tract, cholecystitis, and biliary sludge. Pancreas was difficult to identify due to presence of bowel gas.

Edil et al, reported that in 2008 series, adults were more likely to exhibit abdominal pain (97% versus 63%, $p < 0.001$), and less likely to experience jaundice (25% vs. 71%, $p = 0.001$).[14] In older patients, epigastric pain may result from pancreatitis. Intermittent jaundice and fever may result from recurrent episode of cholangitis. The classic triad of abdominal pain, jaundice, and a palpable abdominal mass is observed in less than 20% of patients.[1,14]

In this patient, the working diagnosis of acute cholecystitis and acute pancreatitis were made. The Virchow's triad was positive and fulfilled the diagnostic criteria for acute cholecystitis based

on Tokyo Guidelines. The working diagnosis of acute pancreatitis in this patient was based on complaints of sharp abdominal pain which radiated to the back and high levels of amylase. The patient had suffered same complaints before which made him got admitted to the hospital for several times in the past six years. From the history, we conclude that the patient had suffered from recurrent biliary infection. Because the patient lives in the tropical zone, infection as a precipitating factor cannot be ruled out.[15,16]

The patient had undergone an abdominal multi-sliced computed tomography (MSCT) to identify the etiology of pancreatobiliary disease. MSCT ruled out tumors and stones and concluded that the patient had intrahepatic and extrahepatic choledochal cyst with suspicion of focal stricture in distal CBD and cholecystitis. Lee et al, showed that MSCT can differentiate CCs from other cause of the biliary tract dilatation. Exclusion of a tumor, stone, or inflammation as the cause of the dilatation in biliary ducts may be a characteristic feature of CC.[4] The occurrence of pancreatobiliary complications were expected to be caused by biliary stasis. Biliary stasis caused biliary sludge which led to obstruction of the choledochal and pancreatic duct. Obstruction and bacterial colonization would cause recurrent biliary tract infection and recurrent acute pancreatitis which later formed stricture that worsen preformed obstruction.[4,8]

Endoscopic retrograde cholangiopancreatography (ERCP) is regarded as the gold standard for the diagnosis of CCs and associated anomalies. However, the ERCP has inherent morbidity because of its invasiveness.[4,17] ERCP can detect anomalous pancreaticobiliary duct junction which usually occurred concomitant with CCs.[4,10] Another imaging technique for evaluation of CCs is magnetic resonance cholangiopancreatography (MRCP). MRCP appears to offer diagnostic information that is equivalent to that of ERCP for assessment of choledochal cysts in adults.[4] In this patient, ERCP and MRCP was not done due to limited health care facilities.

Based on further assessment on cholecystitis and pancreatitis, the cause of both was concluded to be choledochal cyst from the findings in ultrasonography and MSCT. This was in accordance to references that stated CCs could cause acute cholecystitis and pancreatitis.[8,10]

Management consists of conservative management on complication, followed by definitive surgery.[9,10] In this patient, the management of pancreatitis and cholecystitis has been given. For definitive therapy, the patient was planned to have cyst excision and biliary reconstruction. Such therapy was declined by the patient due to his opinion that the symptoms had improved after given medication. Singham et al, declared that the main principle in CCs management are complete cyst excision and extrahepatic biliary tract reconstruction. Cysts excision aims on decreasing biliary stasis which can reduce complications. Extrahepatic biliary tract reconstructions are achieved by hepaticojejunostomy.[8]

This patient was consulted to digestive surgeon for definitive treatment on CC. The digestive surgeon planned to do CBD exploration and cyst excision with biliary reconstruction. Saluja et al, stated that cyst excision with biliary reconstruction is necessary to prevent the recurrence of complications.[10] Cyst excision separates the biliary tree from the pancreatic duct, thus ending the mixing of pancreatic and biliary secretions which was thought to be responsible in the pathogenesis of the disease. If left in situ, the risk of cancer in the retained cyst is as high as 0.7% in the first decade of life to > 14% after the age of 20 years and occurs 15 years earlier than primary cancer.[18]

From this case, imaging modalities such as MSCT concuded that the patient has choledochal cyst, with varying complications, including acute cholecystitis and pancreatitis. However, further assessment by ERCP and MRCP was important in diagnosing CCs. Early diagnosis and prompt treatment is needed to avoid long-term complications. Choledochal cyst need to be considered as one of the differential diagnosis in the etiology of pancreatobiliary infection especially in young adult.

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