

Case Report

A 10-year-old Boy with Giant Choledochal Cyst: A Case Report

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Abstract: Choledochal duct cyst is a rare congenital anomaly in the form of cystic dilatation of both intrahepatic and extrahepatic bile ducts. The clinical symptoms of choledochal cysts are generally due to bile stasis, stone formation, recurrent superinfection, and inflammation. This case depicts a 10-year-old boy presented with a chief complaint of an enlarged abdomen that was rapidly growing, sub-febrile fever and yellowing of the sclera. A choledochal duct cyst was shown in the abdominal ultrasonography. Laboratory examination showed an increase in liver function test, hypoalbuminemia, and prolonged coagulation profile. Magnetic resonance cholangiopancreatography showed a significant cystic dilatation of the common bile duct extending to the common hepatic duct. Surgery was performed twice, first to drain the cyst and second to perform complete excision and anastomoses to the jejunum, as fluid continued to refill the cyst.

Keywords: giant choledochal cyst, bile stasis

Introduction

Choledochal duct cyst is a rare congenital anomaly in the form of cystic dilatation of both intrahepatic and extrahepatic bile ducts. It often occurs in children under ten years of age and is four times more common in women than men. The pathogenesis of common bile duct cysts is unknown, but some studies conclude that it is caused by the irritation of the reflux pancreatic enzymes to the bile duct wall.^{1,2,3} Another well-known cause is the Abnormal Pancreatic-biliary Junction (APBJ), which generates the abnormal connection between the bile duct system and the pancreas.⁴ As there is no sphincter to prevent the reflux of pancreatic juice to the biliary duct, any abnormality such as APBJ or other cause that could increase the pressure of pancreatic secretory will cause the reflux.^{2,4}

The clinical symptoms of choledochal cysts are generally due to bile stasis, stone formation, recurrent superinfection, and inflammation. Obstruction and infection in all choledochal cysts, especially those involving the intrahepatic tract, will cause secondary hepatic cirrhosis in 40-50% of patients. Signs of portal hypertension will

appear, such as upper gastrointestinal bleeding, splenomegaly, and pancytopenia. Therefore, the accuracy of diagnosis is needed to reduce patient mortality and morbidity.⁴

The diagnosis of choledochal duct cyst is made by combining signs and symptoms that arise from the history, physical examination, laboratory examination, and radiological examination

Case

A 10-year-old boy presented with a chief complaint of an enlarged abdomen. Three months before admission to the hospital, the patient complained of an enlarged abdomen and a palpable lump on the right side of the abdomen that was felt to be growing rapidly. These complaints were accompanied by: sub febrile fever and yellow eyes. There was no abdominal pain, no nausea and vomiting, and no complaint of defecation. Abdominal Ultrasound showed that the patient has a choledochal duct cyst. The patient was then referred to our center for further examination.

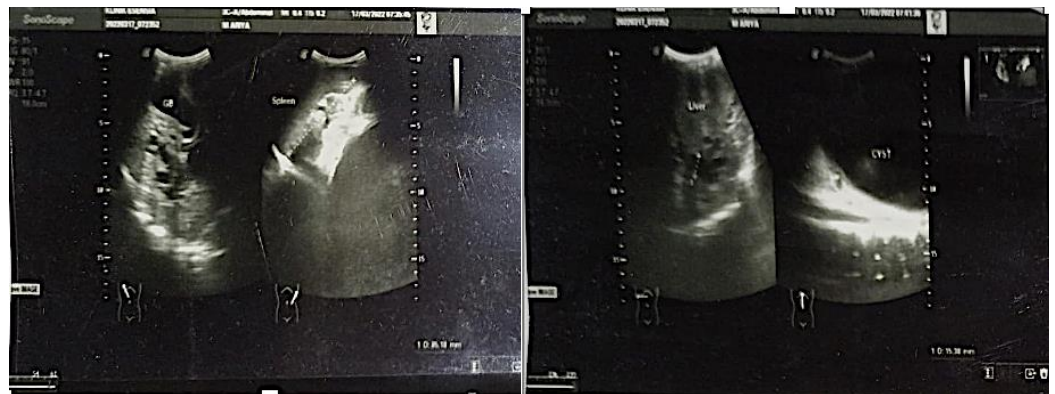


Figure 1. USG Abdomen: Large cystic lesions on the upper to lower abdomen (the size cannot be reached by the probe) with widening of the intrahepatic bile ducts, tending to be choledochal cysts (Todani classification type IVa)

The child looked thin with an enlarged, distended abdomen on the physical examination. A palpable mass was found in the right abdomen extending to the left abdomen. The largest abdominal circumference was 79 cm, and the umbilical circumference was 74 cm (pre-operative). The sclera was also icteric. Vital signs were within normal limits, while the anthropometry examination showed the impression of malnutrition with normal stature.



Figure 2. Patient clinical photo pre-operative

Laboratory examination showed an increase in liver function test (LFT), hypoalbuminemia, and prolongation of the coagulation study. Magnetic resonance cholangiopancreatography (MRCP) showed a significant cystic dilatation of the common bile duct extending to the common hepatic duct (size \pm AP 16.7 x CC 26.4 x LL 18.3 cm), pressing the pancreas, stomach, and the surrounding bowel structures to the left lateral and accompanied by minimal sludge. Dilated right hepatic duct (\pm 1.5 cm in diameter) and left hepatic duct (\pm 1.7 cm in diameter) showed the impression of a choledochal cyst (Todani Classification type IVA), cystic duct dilation (\pm 1.3 cm diameter), and minimal ascites. The drainage of the choledochal duct cyst was performed by extracting around 3 liters of greenish-yellow fluid.

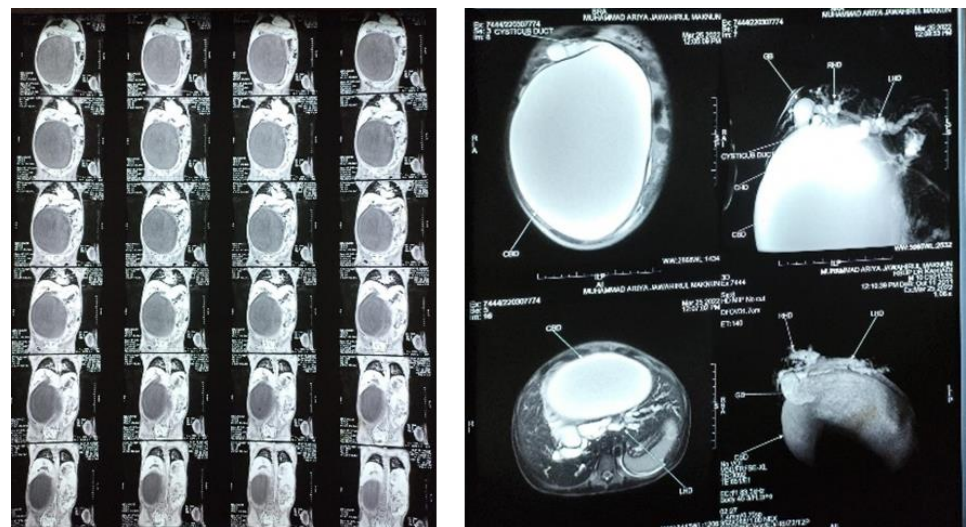


Figure 3. Magnetic resonance cholangiopancreatography (MRCP).

On the fifth day after the drainage procedure, the stomach enlarged, accompanied by fever, nausea and vomiting. An abdominal x-ray was performed and the results showed broad ground glass opacity from the right to the left hemiabdomen accompanied by increased pressure to the left side of bowel loops. The opacity was suspected to be an intra-abdominal mass. There was no picture of ileus or pneumoperitoneum. An exploratory relaparotomy was then further planned.

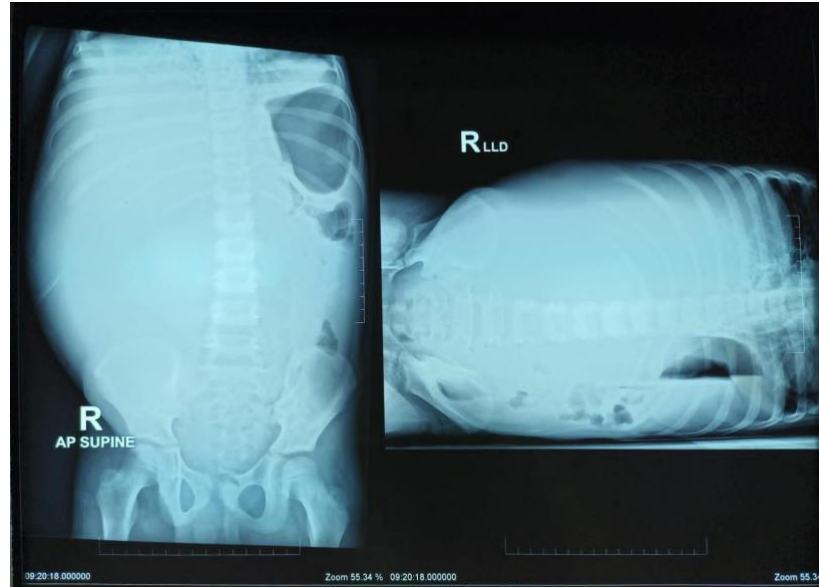


Figure 4. Abdominal x-ray after the first surgery

The results of the relaparotomy showed that the bile duct cyst was again filled with fluid for more than 2 liters; thus, a Roux en Y cystojejunostomy and open drainage were then performed.

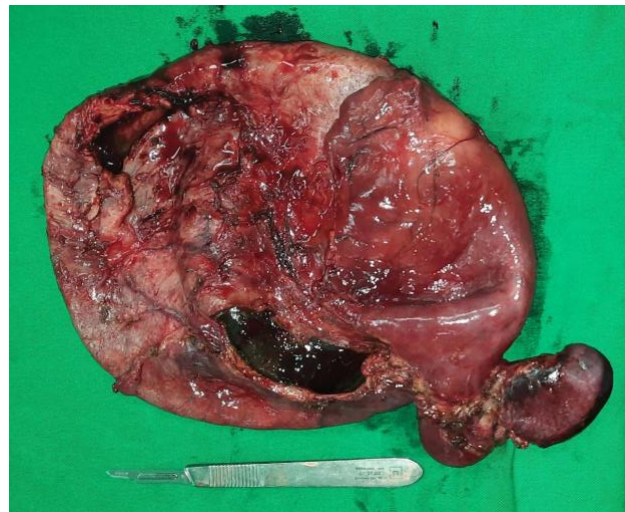


Figure 5. Choledochal duct cyst

The biopsy result revealed that the primary diagnosis of this case is choledochal duct cysts. The test also demonstrated the occurrence of chronic non-specific cholecystitis and hepatic cirrhosis with F4 degree of fibrosis. There was no sign of malignancy.

Discussion

This case described a 10-year-old boy presented with an abdominal lump that progressively enlarged within three months, accompanied by complaints of jaundice, weight loss, and increased liver function test. The lump was caused by obstruction, which damaged the hepatocytes. The hepatocytes injury could be assessed through the histopathological results, in which our patient exhibited grade 4 fibrosis. Portal hypertension often occurs in patients with choledochal cysts; however, in this particular patient, there are no signs of portal hypertension or large cysts that obstruct the biliary tract.

Ultrasonography is the initial modality used to evaluate abnormalities in the biliary tract. The diagnostic technique adequately shows cystic or fusiform structures of the hepatic portal area in choledochal cysts. A biliary CT scan, either ERCP or percutaneous retrograde cholangiography, could be performed to confirm the ultrasound examination results.^{5,6} In this patient, the ultrasound results from the previous hospital detected the impression of a choledochal cyst. To verify the ultrasound result and confirm the diagnosis, MRCP was performed. The result demonstrated a type IV-A choledochal duct cyst, and the patient was scheduled for surgery.

Recently, the surgical management of choledochal cysts has been developing. Surgical management of choledochal cysts mainly consists of drainage by cyst enterostomy. However, patients who undergo cyst drainage without cyst excision had an increased risk of carcinoma and incidence of cholangitis or pancreatitis. Thus, cyst excision is currently the prioritized method in the surgical management of choledochal cysts.⁷ The surgical management preferred in choledochal cysts cases varies depending on its type in type-IV cysts, particularly type-IVA.^{8,9}

Cholangiocarcinoma is an ominous complication of unresected choledochal cysts, occurring around 20-30% in early adulthood. The risk of cholangiocarcinoma remains high in patients who underwent drainage procedures with either partial cyst resection or no cyst resection. The risk of cholangiocarcinoma also increases in the type-I and type-IV choledochal cyst. In our case, surgery was performed twice, first to drain the cyst and second to perform complete excision and anastomoses to the jejunum, as fluid continued to refill the cyst.¹⁰ The anatomical pathology of the patient showed no signs of malignancy. However, the biopsy result suggests a grade 4 fibrosis; hence further monitoring of liver function is needed in this patient.

Conclusion

Choledochal duct cyst is a rare congenital anomaly in children. Complaints that arise vary but are often symptoms due to biliary obstruction. The diagnosis is based on history, physical examination, laboratory and radiological examination. Abdominal ultrasound can provide significant input in cases with common bile duct cysts. Surgical management of choledochal cyst is currently developing rapidly. Approximately 20-30% will develop into malignancy in early adulthood. Thus, early diagnosis and the type of cyst determine the patient's prognosis.

Abbreviation

AP	: anteroposterior
LL	: laterolateral
CC	: craniocaudal

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