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CHOLEDOCHAL CYST : A DIAGNOSTIC DILEMMA

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Introduction

Choledochal cyst is a benign and rare congenital cystic dilatation of the biliary tract. It presents commonly in East Asian populations with up to 1:13 000 in Japan[1,3]. It primarily affects female infants and young children though it can present in adults. Due to its varying clinical presentations among different age groups, a delay in diagnosis can lead to severe late complications including malignant transformation, cholangitis, pancreatitis, and cholelithiasis. We report here 4 cases of choledochal cyst with unusual presentations.

Case 1

A 2-year-old boy, with a left hydrocele, presented to a private hospital with fever, vomiting, abdominal distension and lethargy for 2 days. On examination, he was found to be in septic shock, jaundiced and noted to have hepatomegaly. His urine was tea coloured. There was no history of prolonged jaundice in infancy. He was resuscitated, ventilated and put on inotropes and antibiotics. Ultrasound (US) abdomen showed a choledochal cyst (6x6cm) with ascites and a complex left hydrocele. His blood cultures grew Salmonella non typhii and the antibiotics were changed accordingly. However, his fever and abdominal pain persisted. Repeated US abdomen showed enlarging choledochal cyst (9x9cm), and was decided for percutaneous US guided cyst drainage. Elective choledochal cyst excision and Roux-en-Y hepaticojejunostomy was scheduled 6 weeks later after completion of treatment for his Salmonella infection. Intraoperatively, there was a fusiform dilatation of common bile duct (CBD) measuring 10x10cm with dilated intrahepatic duct containing a large biliary cast and debris which cultured carbapenem resistant Klebsiella pneumoniae. He was discharged well 2 weeks post operation.

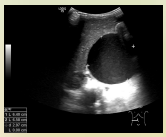

Figure 1.1: Extrahepatic choledochal cyst 6.6x6.4cm with minimal debris within.

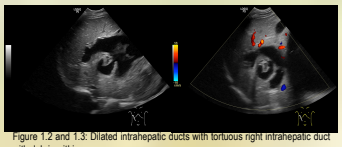

Figure 1.2 and 1.3: Dilated intrahepatic ducts with tortuous right intrahepatic duct with debris within.

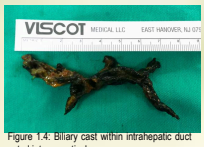

Figure 1.4: Biliary cast within intrahepatic duct noted intraoperatively.

Case 2

A 16-month-old boy came in with abdominal distension for 4 days, vomiting, diarrhoea and fever for 3 days. He had elevated liver enzymes and was treated as hepatitis A infection. US was done and reported as hepatitis with ascites. However, his condition did not improve and his stools became pale. A vague mass was felt over the right hypochondriac region. A repeated US revealed a dilated biliary tree with gallbladder stones as well as loculated ascitic fluid (6x2cm) containing debris at the subdiaphragmatic region, suggestive of a perforated biliary system. However we were unable to trace the whole CBD. A percutaneous drain was inserted at the subhepatic area. As his condition worsened, a CT abdomen was done. It was reported as dilated and fusiform CBD widest 17mm with long common channel (7x18mm) and sludge within the whole biliary tree, complicated with perforation and multiloculated collections. He was treated with antibiotics and 2 weeks after, laparotomy and cholecystostomy with a peritoneal drain insertion at the subhepatic area was done. As his condition improved, choledochal cyst excision and Roux-en-Y hepaticojejunostomy was done 8 weeks after his first operation. Intraoperatively, noted the CBD dilated measuring 3x4cm with sludge within the common channel. Postoperative, he recovered tremendously and he was allowed home a week after.

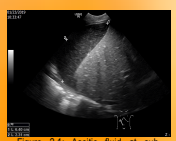

Figure 2.1: Ascitic fluid at sub diaphragmatic area measuring 6.4x7.3 cm with debris.

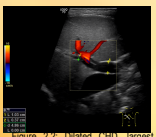

Figure 2.2: Dilated CHD, largest diameter 1cm with 4.8cm length.

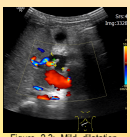

Figure 2.3: Mild dilatation of IHD measuring 4 mm and 7mm.

Case 3

An 11-month-old girl came to a private hospital with high grade fever and seizures associated with reduced oral intake. Upon examination, she was septic looking but not jaundice. She was covered with antibiotics and the cause of the fever was investigated. A mass was palpable at the right hypochondrium and her stools became pale. US abdomen was reported as a large choledochal cyst measuring 7.7x10.5cm with a dilated right IHD up to 2.9cm and presence of complex fluid around the cyst and the right subhepatic fossa, suggestive of perforation. An US guided percutaneous drainage of cyst was inserted and she was treated with antibiotics. We proceeded with choledochal cyst excision and Roux-en-Y hepaticojejunostomy 6 weeks after completion of her intravenous antibiotic. She was discharged well 2 weeks after her operation once she was able to tolerate orally.

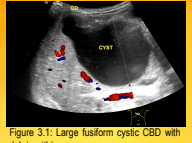

Figure 3.1: Large fusiform cystic CBD with debris within.

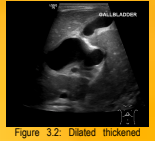

Figure 3.2: Dilated thickened wall gallbladder and cystic duct with sludge within

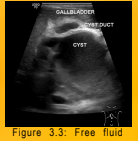

Figure 3.3: Free fluid around cyst and sub hepatic region

Case 4

An 18-month-old girl presented with a 3 day history of vomiting and abdominal pain for 1 day. Otherwise, there was no fever or obstructive symptoms. Upon examination, she was slightly jaundiced and her abdomen was distended with a tender vague mass palpable at right hypochondrium. Abdominal X-ray revealed no dilated bowel. Ultrasound was reported as elongated tubular CBD measuring 2.6x2.6x4.2cm with debris within. The cystic duct and CHD were also dilated and the gallbladder wall was diffusely thickened. There were large amount of generalised free fluid with debris and septa seen. Antibiotic was started and cholecystostomy with cyst drain and peritoneal drain insertion at right subhepatic region was done. 6 weeks after her first operation, we proceeded with choledochal cyst excision and roux-en-Y hepaticojejunostomy. Intraoperatively, perforation noted at posterior wall of cyst near junction of cystic duct and CHD. Postoperative recovery was uneventful and she was discharged 2 weeks after the operation.

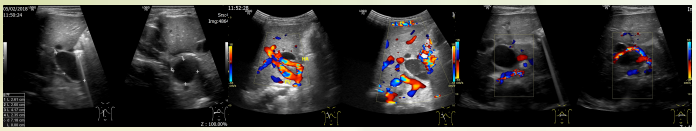

Figure 4.1: Elongated cyst at portal hepatis (choledochal cyst) with size of 2.6x2.6x4.2cm

Figure 4.2: Dilated CBD which extends until pancreatic head.

Figure 4.2: Dilated and tortuous cystic duct

Discussion

Base on these case reports, they presented to the hospital with different presentations. Due to the unusual presentations, these lead to a delay in diagnosis. However, prompt investigations were done and appropriate treatments were performed.

There is no consensus in the literature about the most common clinical presentation. The presentation also changes according to age. Though abdominal pain, jaundice and abdominal mass are described as the classic triad of choledochal cyst, it only presents around 0-33% in children[1,3,4,6]. It is found that infants were more likely to present with an abdominal mass and older children presented more with nausea, vomiting, and abdominal pain[1,3]. It is also reported that there is no different of jaundice presentations for both infants and children[3]. One study reported that only paediatric population developed rare presentations of spontaneous biliary perforation and none in adults[6].

US is the initial diagnostic modality of choice[1,2]. A properly performed high resolution US can measure the duct dilatation precisely and detect the presence of stone or sludge. Computed tomography would help in providing additional information in comparison with the US. Magnetic resonance cholangiopancreatography (MRCP) is useful in assessing the hepatobiliary and pancreatic pathology in children[2,4].

Surgical treatment is essential in reducing the risk of potential morbidity of developing malignancy and prevent other complications. The surgical procedure of choice now is choledochal cyst excision and Roux-en-Y hepaticojejunostomy[1,2,4].

Conclusion

Choledochal cyst is a rare congenital biliary tract disorder. Its symptoms vary and non specific, causing a delay in diagnosis and may lead to the progression of its complications. It requires a high degree of suspicion and correct investigation for diagnosing and treating appropriately. US is the choice of initial diagnostic modality to support the diagnosis. Definite management will be choledochal cyst excision and Roux-en-Y hepaticojejunostomy.

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Acknowledgement

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