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EDITORIAL

CHOLEDOCHAL CYST DIAGNOSTIC AND OPERATIVE CHALLENGE

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Introduction :

The variations in the morphological characteristics of the extra-hepatic biliary system are numerous. It has been stated that the extra-hepatic biliary system has more anomalies in one cubic centimeter of the space around the region of the cystic duct than any other part of the body [1,2].

The incidence of congenital anomalies of the extra-hepatic biliary system varies between 0.58% and 47.2% [3].One such rare anomaly is Choledochal cyst (CDC), also known as congenital common bile duct cyst (BDC), is a rare type of bile duct cyst of uncertain origin. The majority of cases reported are young women and children of Asian descent. In North America, its incidence is estimated to be 1/150 000[2], but it is increasing in Western adults. The most common symptoms of CDC are abdominal pain, jaundice and abdominal mass.[4]

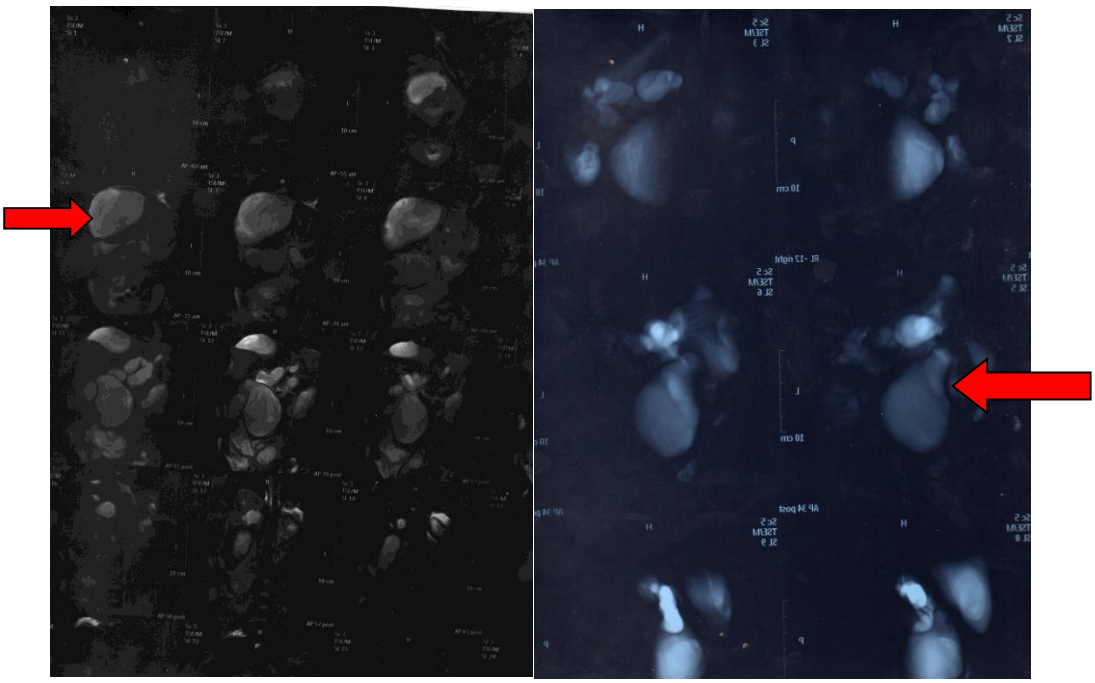
Case Report :

A female of 16years, referred for specialized surgical intervention .Two months prior to the referral time she was diagnosed as chronic calculus cholecystitis,the diagnosis depenend on the clinical features and abdominal ultrasound which revealed gall bladder stones and thick wall gall bladder. She underwent laparoscopic cholcystectomy which was converted to open cholcystectomy .The Intraoperative findings were normal gall bladder (GB), deep GB–like structure connected to the common hepatic duct. Open Cholecystectomy was done, and the Postoperative period was smooth apart of mild right hypochondrial pain . At arrival she was ill, pale ,dehydrated & jaundiced .Pulse 120/ m ,BP120/70mmHg, RR 20/min ,temperature 37.5 C .There was right subcostal scar & four small scars (laparoscopic cholcystectomy) .There was right side gurdening and tenderness .No ascities .

Investigations revealed normal hemoglobin and Platelets, TWBCS 17000/cm^3 .Urine analysis was clear . Renal function tests were normal .Serum bilirubin 3.3mg/dl (conjugated 1.8 mg/dl) , Alkaline phosphatase 372 IU/L,liver enzymes and coagulation profile were normal. .Ultrasound showed , enlarged liver with multiple cystic areas ,connecting with dilated intrahepatic bile ducts , features of Choledochal cyst type 4 ,with biloma ?Caroli's Disease , free intraperiotneal fluid collection . MRCP revealed ,large subpherenic collection which was pushing the right lobe of the liver downward and medially, right and left hepatic ducts were dilated and the common bile duct and common hepatic duct both were dilated .Figure(1)

EDITORIAL

Figure (1): U/S scan



She was planned for LAPAROTOMY .We found afusiform type1 Choledochal cyst at the level of supraduodenal part of the common bile duct (Figure 2) about 10*12 cm and there were adhesions around the biliary system . Excision of the cyst (Figure 3) and Roux-en-y Hepaticojejenostomy was done . The histopathology was benign choledochal cyst .Uneventful post operative course. Postoperative cholangiogram was normal .

EDITORIAL

Figure(2)

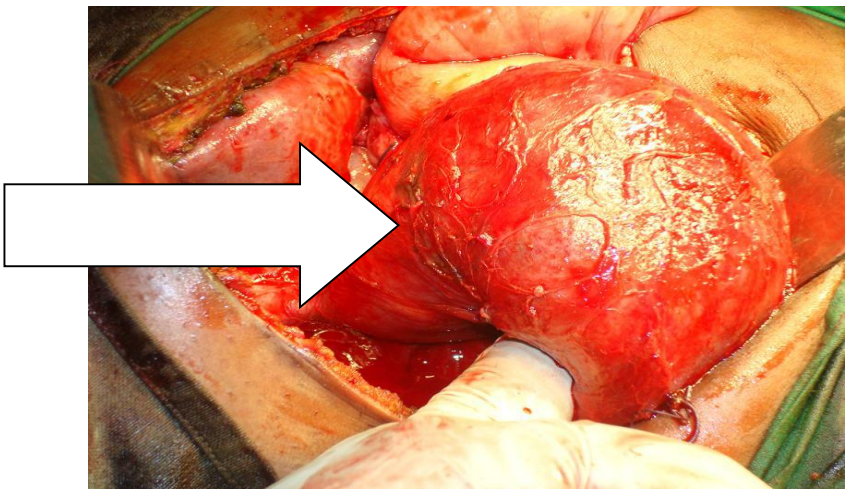
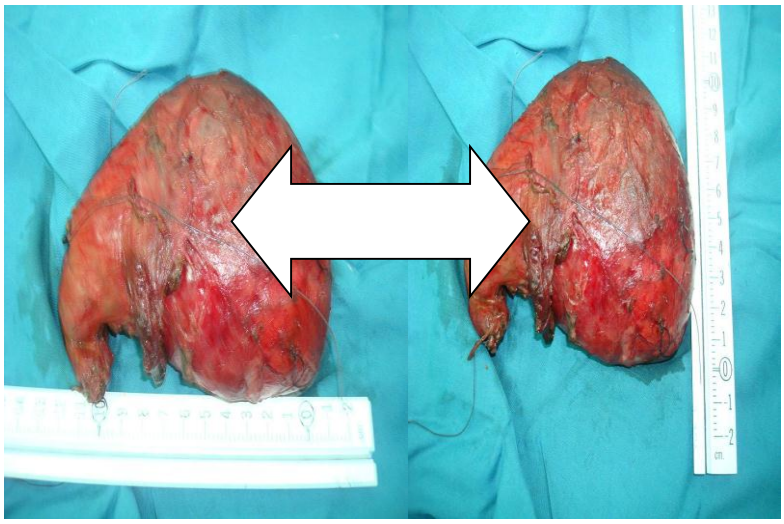


Figure (3)



Discussion :

Choledochal cyst (CDC) is graded based on Todani's classification[5] and can be divided into five types, which are anomalies in the intra- or extra-hepatic bile ducts, or both. CDC can be diagnosed based on percutaneous and endoscopic ultrasound or endoscopic retrograde cholangiopancreatography. Magnetic resonance cholangiography may also contribute to its diagnosis.[6]Type IV cysts are more commonly observed in adults than in children[7], while type I cysts are more commonly observed in Asian patients

EDITORIAL

.Type III cyst, a cystic-like dilation of the terminal common bile duct, is rare[8,9].In this case the diagnosis was initially missed ,because they depended mainly on ultrasound and they neglected the clinical features (Young female, jaundice and right hypochondrial pain) which are pathognomic for Choledochal cyst.

Treatment of types I and IV cysts has undergone much change in the past years. Although Mcwhorter first described cyst excision and Hepaticojejenostomy in 1924, this surgery was initially abandoned because of multiple complications. Surgical strategies of cyst marsupialization and choledochoraphy failed because of significant mortality and morbidity.[10,11]

Subsequently, internal drainage of cysts via cystenterostomy became popular. Depending on anatomic proximity, cysts were incised and anastomosed to the duodenum or jejunum.[12]Although this operation resulted in periprocedural relief of symptoms, multiple complications resulted. Reflux of the enteric contents into the cyst and biliary tree resulted in recurrent ascending cholangitis. The site of anastomosis was also prone to stricture formation, resulting in obstruction, bile stasis, stone formation and recurrent cholangitis. Most importantly, surgeons found that leaving the cyst intact carried a significant risk of malignant transformation.[13,14] The overall success rate of internal drainage procedures is 30%, the risk of postoperative malignancy is 30%, the mortality rate is 11%, and more than half who undergo this procedure require re-operation.[15,16]Therefore, internal drainage is currently thought to be a dangerous and incomplete treatment of CDC.[15]

Instead, surgeons favour complete cyst excision and hepaticoenterostomy. If left in situ, the risk of cancer in the retained cyst is as high as 50% and occurs 15 years earlier than primary cancer. Therefore, the cyst should be excised completely from the hepatic hilum to the pancreatic duct.[17,18] The hepaticoenterostomy can either be a hepaticoduodenostomy or a Roux-en-Y Hepaticojejenostomy (RYHJ). The success rate of RYHJ has been shown to be as high as 92%. This procedure has a reported complication rate of 7%, compared with a complication rate of 42% with hepaticoduodenostomy.[19] Hepaticoduodenostomy carries with it the risk of bilious gastric reflux, gastritis and esophagitis, ulceration and malignant disease. Furthermore, Todani and colleagues reverted from advocating hepaticoduodenostomy as the procedure of choice when they discovered a patient with hilar adenocarcinoma after excision. They hypothesized that that the reflux of bile and active pancreatic enzymes from the duodenum can irritate the hilar epithelium and lead to malignant transformation.[20]Many surgeons recommend end-to-end RYHJ to avoid the formation of a long blind pouch, which can result in bile stasis, reflux, cholangitis and stone formation.[19] Authors also recommend creating a wide stoma at the hepatic hilum by extending the incisions up the lateral walls of the hepatic ducts to allow free drainage and avoid anastomotic stricture. The minimum diameter of the stoma has been suggested to be 3 cm.5 After cyst excision and hepaticoenterostomy, patients symptoms improve, intrahepatic duct dilations decompress and hepatic fibrosis and varices regress . [20]The complications of cystenterostomy and benefits of cyst excision and hepaticoenterostomy are both so substantial that surgeons now recommend revision of previous internal drainage procedures even for patients with no symptoms or complications.[21]

In conclusion diagnosis of Choledochal cyst depends on the clinical triad (Young female, jaundice and right hypochondrial pain) and then diagnosis should be confirmed by abdominal ultrasound and the best modality of surgery is total excision of the diseased bile duct with reconstruction by Roux-en-Y choledochojejunostomy .

EDITORIAL

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