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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Portal hypertension secondary to choledochal cyst

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Choledochal cyst, a congenital condition in infants and children, is characterized by dilatation of the bile ducts, which manifests as jaundice, abdominal mass and abdominal pain. Choledochal cysts can lead to complications such as cirrhosis and portal hypertension. Therefore, early detection is necessary. A case of a two-year-six-month-old boy with type I choledochal cyst who underwent choledochal cyst excision at six months of age was reported. Two years later, the patient presented with esophageal variceal bleeding, splenomegaly, and hypersplenism. Severe stenosis of the main portal was observed on computed tomography (CT) angiography, along with abnormalities and thickening of the portal branch. Esophagoduodenoscopic examination showed grade II esophageal varices. The patient was diagnosed with portal hypertension secondary to the choledochal cyst. A late consequence of choledochal cysts is portal hypertension. The long-term risk of liver fibrosis and portal hypertension needs to be taken into consideration. Improving the prognosis of children with choledochal cysts requires timely preventive care.

Δευτεροπαθής πυλαία υπέρταση λόγω κύστης χοληδόχου πόρου

Περίληψη στο τέλος του άρθρου

Key words

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An estimated 1:13,000 Asian populations have choledochal cysts, a congenital condition marked by bile duct dilatation that manifests in pediatric patients as jaundice, abdominal lumps, and abdominal pain.[1,2] The distal biliary obstruction was common in patients with severe choledochal cysts, resulting in bile stasis and subsequent serious complications including acute cholangitis, abnormal liver function, as well as choledochal cyst perforation and coagulopathy.[1] Choledochal cysts are typically diagnosed in children. Multimodality imaging methods such as computed tomography (CT), magnetic resonance imaging (MRI), ultrasound, and MR cholangiopancreatography (MRCP) are commonly used to make the diagnosis.[3]

Portal hypertension is a recognized consequence of a delayed diagnosis of a choledochal cyst and can be associated with secondary biliary cirrhosis, extrahepatic portal venous thrombosis, or compression of the cyst over the portal vein.[4] This case report describes a child with portal hypertension after a choledochal cyst.

CASE PRESENTATION

A two-year-six-month-old boy was admitted to the emergency department due to pallor. Two weeks before admission to the hospital, the patient experienced defecation with blackish stools that recurred for the past one month (fig. 1). This week the blackish

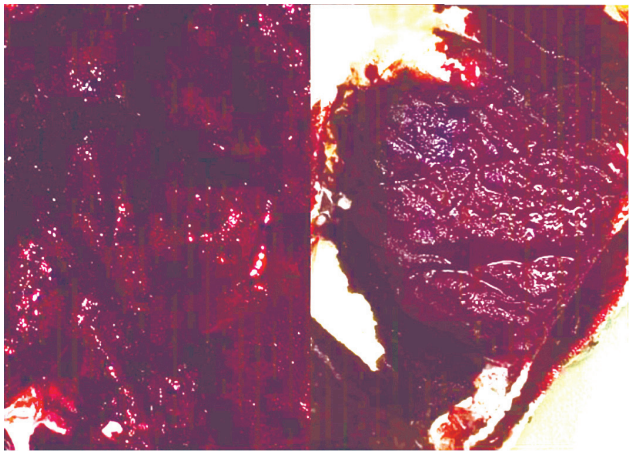

Figure 1. Blackish stool.

stools became more frequent and the patient looked paler. The patient also complained of a hardened abdomen (fig. 2). There were no complaints of fever, vomiting, diarrhea, and other.

At the age of three months, the patient was referred from a regional hospital with persistent jaundice. The patient had been jaundiced since birth followed by pale stools a week after birth, urine color like tea, enlarged abdomen, bloating, and often vomiting when drinking formula milk. The patient had complained of jaundice since one week of age but was considered physiological jaundice. Because of persistent jaundice, the patient's parents brought him to the regional hospital at two months of age and was diagnosed with Hirscprung's disease because of a distended abdomen. However, because the jaundice did not improve at the age of three months, the patient was referred for further examination. Abdominal ultrasound examination showed an anechoic lobulated lesion in the right lobe of the liver in the porta hepatis area (around the extrahepatic biliary duct) with a volume of 100 ccs which could be choledochal cyst type 1 (fig. 3).

CT scan of the abdomen showed a cystic lesion with well-defined regular borders measuring ±6.3×8.3×13.6 cm in the common bile duct (CBD), which was not visible on contrast enhancement,

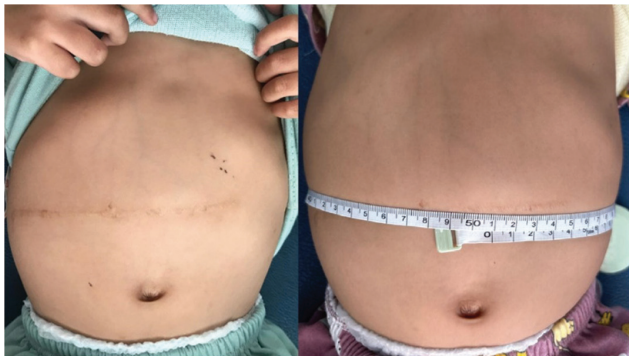

Figure 2. The abdominal examination of the patient.

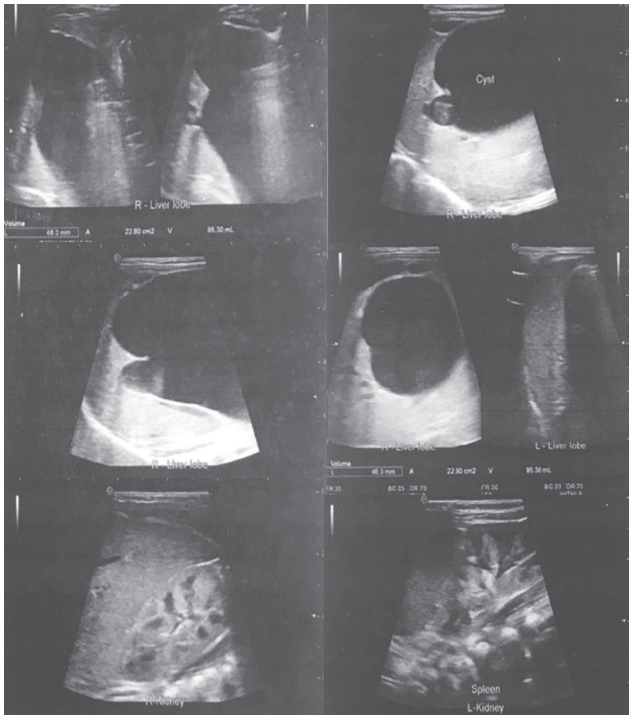

Figure 3. Abdominal ultrasound showed choledochal cyst type 1.

dilatation in the CBD ±1.4 cm, right IHBD ±0.7 cm, left ±0.6 cm, appearing to compress the surrounding bowel, displacing the right kidney posteriorly, and displacing the pancreas to the left superolateral. The liver has normal size, normal density, no visible dilatation of the intrahepatic (IHBD) or extrahepatic (EHBD) bile ducts, venous portal/hepatica normal, and no visible mass/nodules. Gallbladder was enlarged ±4.8 cm in sagittal projection, normal density, no mass/stone/cyst. Abdominal CT scan showed a cystic lesion of ±6.3×8.3×13.6 cm in the CBD with a dilated biliary tree in the proximal area and gallbladder hydrops, which suggest a type of IVa choledochal cyst (according to Todani classification) (fig. 4). Based on an abdominal ultrasound and abdominal CT scan, the patient was diagnosed with a choledochal cyst. The cholecystectomy and choledochal cyst excision with Roux-en-Y hepaticojejunostomy were performed at 6 months old. Intraoperatively, an extrahepatic cyst 20 cm×10×10 cm was found with an iatrogenic perforation of the portal vein. During excision and adhesiolysis of the cyst from the portal vein, a Lilly procedure was performed to prevent further damage. After that, the procedure continued with Roux-en-Y hepaticojejunostomy anastomosis. Intraoperative biopsy examination showed sections of cyst wall tissue without lining epithelium. The stroma was fibrous connective tissue with proliferation and dilatation of blood vessels, some congested, and a scattering of inflammatory cells, lymphocytes, and histiocytes. Tissue biopsy showed a choledochal cyst. Intraoperative liver biopsy showed liver lobules composed of hepatocytes with ballooning degeneration. There was extensive fibrosis in the portal tract with bridging necrosis in the liver lobules. A liver biopsy showed extrahepatic cholestasis with extensive fibrosis with liver cirrhosis.

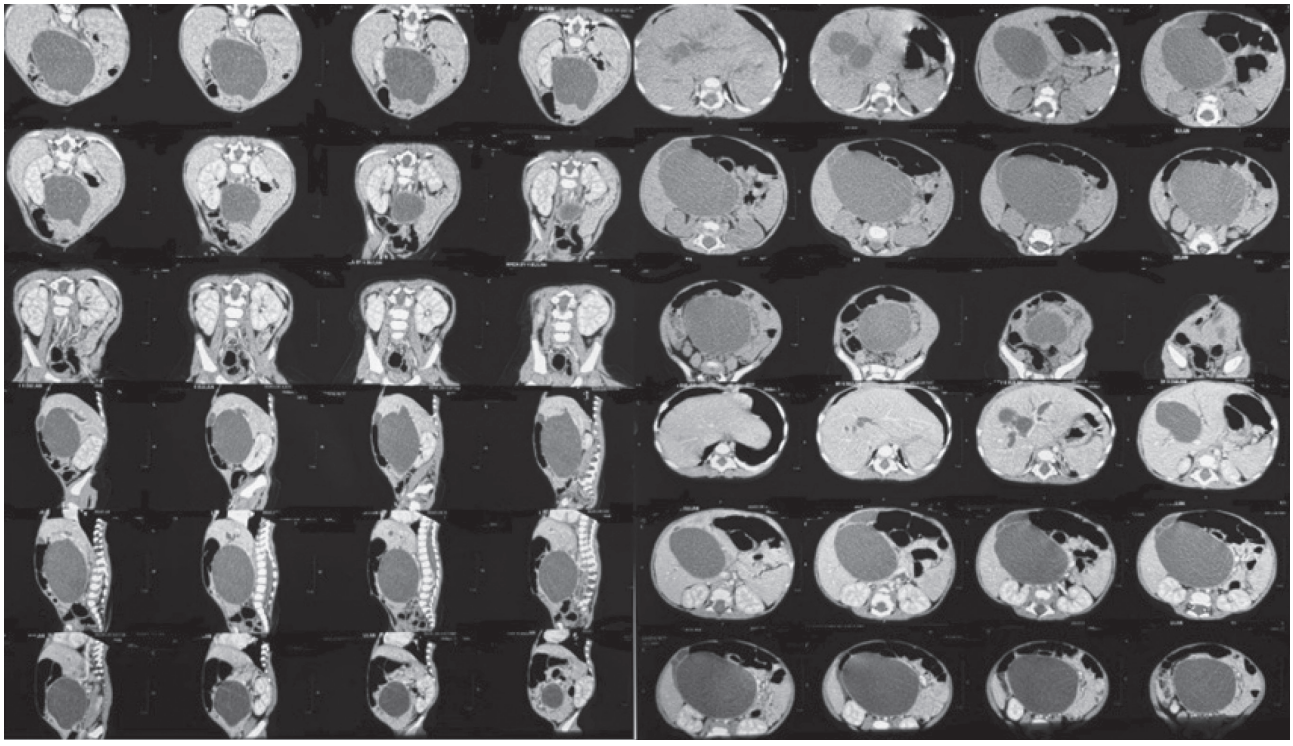

Figure 4. Abdominal computed tomography (CT) scan showed a choledochal cyst.

Two years after surgery, the abdomen appeared to be enlarged again but not jaundice. Physical examination revealed malnourishment. There were no icteric sclera and anemic conjunctiva. Abdominal examination revealed a distended abdomen, no palpable liver, but splenomegaly (Schaffner 2) (fig. 2).

The patient was a first child, born spontaneously with a gestational age of 37 weeks, birth weight of 2,700 grams, length of 47 cm, and crying immediately. The patient was breastfed from birth until the age of two. The last immunization was measles at nine months of age. There was no similar disease in the family.

Echocardiography showed a small (0.3 cm) secundum atrial septal defect (ASD) with an ejection fraction of 85%. Abdominal ultrasound evaluation showed the liver measuring anteroposterior diameter ±8.8 cm, sharp angle, partially irregular edges, heterogeneous intensity echo texture parenchyma, no visible dilation of IHBD/EHBD, portal vein diameter ±0.6 cm (normal <0.8 cm) with portal vein velocity 16.7 cm/s (normal >20 cm/s), v. hepatica appeared normal, no visible nodules/cysts/masses. Gallbladder had normal size, no wall thickening, no stone/nodule/sludge. Lien was enlarged measuring ±11.7 cm, echo intensity of parenchyma appeared normal, and there was no mass/cyst, while v. lienalis was ±0.26 cm. The pancreas has normal size, echo intensity of parenchyma appears normal, there is no visible dilation of the ductus pancreatic, and no visible mass/cyst/calcification. The right and left kidneys are normal in size, the echo intensity of the cortex appears normal, the cortex sinus boundaries are clear, there is no ectasis of the pelvicalyceal system, and there are no stones/cysts/masses. No echo intensity of extraluminal free fluid in the abdominal cavum and pelvic cavum. Abdominal ultrasound showed cirrhotic hepatitis with splenomegaly and decreased portal vein velocity supporting the features of portal hypertension (fig. 5).

In the pre-contrast CT scan angiography examination, it was found that there was no mass in the abdominal cavum or pelvic cavum and no abnormal contrast enhancement. The liver was enlarged with a picture of kissing sign (+), normal density, and no visible mass/nodule. The gallbladder was of normal size, normal density and free of mass, stone, or cysts. Pancreas had normal size, normal parenchymal density, and no mass, stones, or cysts. Lien was enlarged ±10.2 cm, there was normal parenchymal density, and no mass, cysts were observed. Right and left kidneys were normal in size, and had normal parenchymal density, without pelvicalyceal system ectasis, and without mass, stone, or cyst. Bladder was filled with sufficient fluid, without wall thickening, stones, or masses. There was no extraluminal free fluid density in the abdominal cavum, and right and left pleural cavum, neither locoregional lymph node enlargement. There was no osteolytic or osteoblastic process. CT angiography examination showed severe stenosis of the main port with abnormality and thickening of the branch of the portal that gets collateral from segment IVb, V of the right lobe of the hepatic, as well as dilatation and tortuosity of v. lienalis and dilatation of the distal esophageal vein, and fundus to the gastric minor curvature. There was no calcification of the abdominal aorta, nor abnormality in the vena cava. The coeliac truncus had branches in the right and left hepatic arteries; a. lienalis; a. gastric right and left; a. renalis right and left; a.

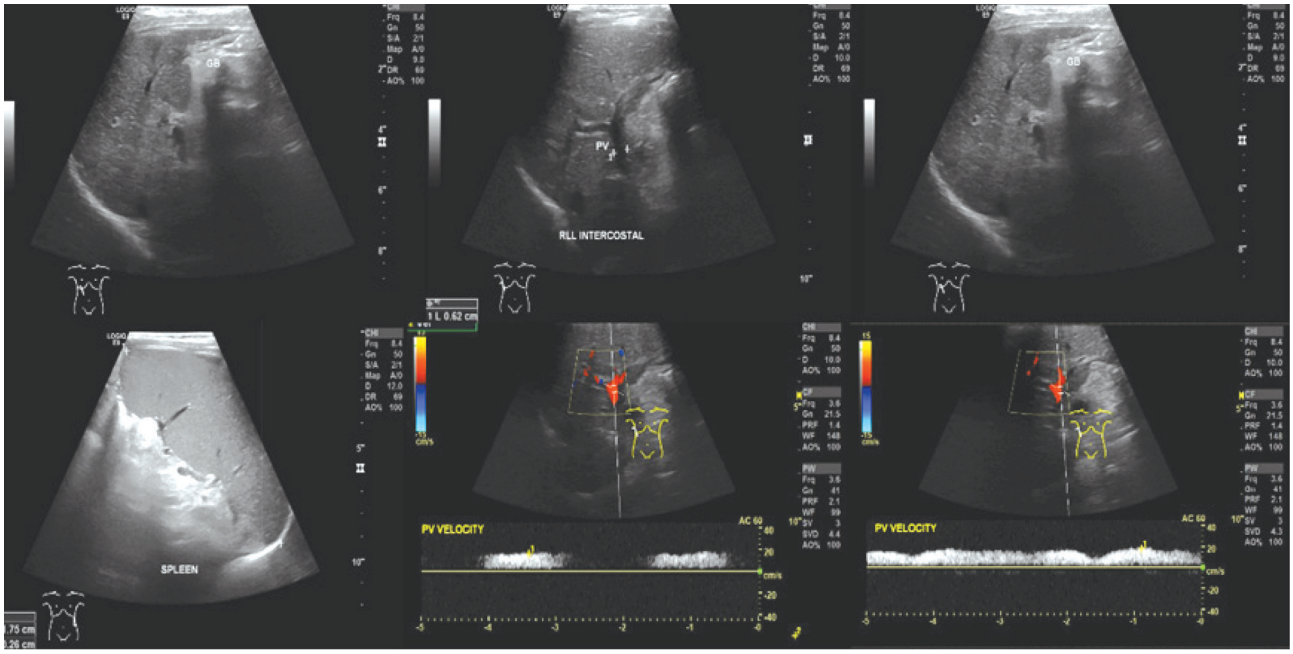

Figure 5. Abdominal ultrasound showed cirrhotic hepatitis with portal hypertension.

mesenteric superior and inferior and a. iliaca externa and interna right and left appeared normal. No arteriovenous malformation (AVM) or fistulation was observed. CT angiography showed severe stenosis of the main portal accompanied by abnormality and thickening of the branch of the portal that received collateral from segment IVb, V of the right lobe of the liver and dilatation and tortuosity of v. lienalis accompanied by esophageal varices with hepatosplenomegaly (fig. 6).

Esophagoduodenoscopic examination showed grade II esophageal varices in four lanes, red-color signs (RCS) (+), red wale markings (RWM) (+) from the middle of the esophagus. In the gastric corpus, mucosa showed erosion, antrum mucosa appeared with hyperemic liner (watermelon sign), and retroflection position appeared to have varicose veins on cardia, RCS (+). In the duodenum, DI mucosa was found, digital implant (DII) impression appeared in less dense villi arrangement. Esophagoduodenoscopic examination showed grade III esophageal varices with bleeding stigmata, gastro-oesophageal varices type 1 (GOV I) with bleeding stigmata, and mild portal hypertensive gastroduodenopathy (fig. 7).

Laboratory examination revealed hemoglobin 7.6 g/dL, hematocrit 27.0 %, white blood cells (WBC) 10.97 10^3/µL, and thrombocytes 180×10^3/µL, APTT 21.20 sec, PPT 18.0 sec, AST 66 U/L, ALT

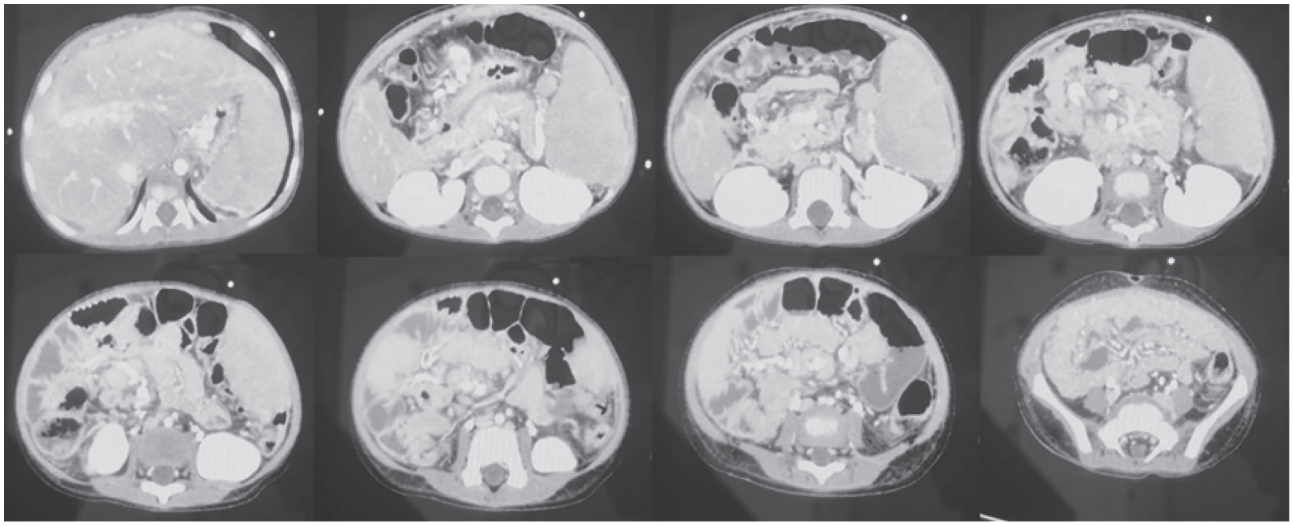

Figure 6. Computed tomography (CT) angiography showed severe stenosis of the main portal.

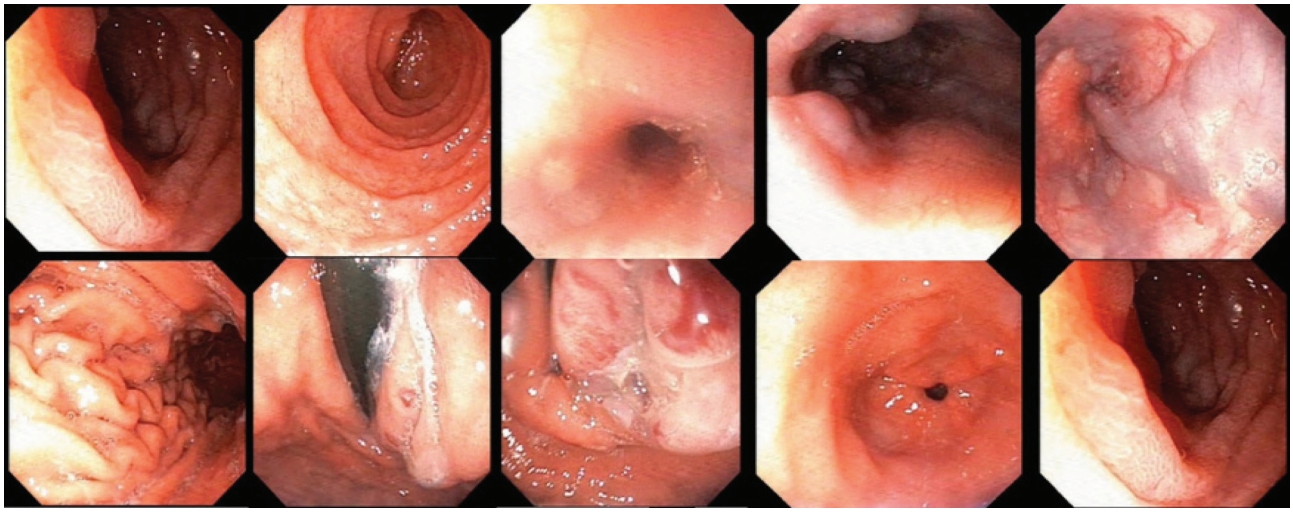

Figure 7. Esophagoduodenoscopic examination showed grade III esophageal varices.

39 U/L, total bilirubin 0.60 mg/dL, direct bilirubin 0.30 mg/dL, albumin 4.07 g/dL, BUN 14.2 mg/dL, serum creatinine 0.3 mg/dL, kalium (K) 4.30 mmoL/L, natrium (Na) 136 mmoL/L, chloride (Cl) 94.0 mmoL/L. Post packed red cells (PRC) transfusion revealed hemoglobin 12.9 g/dL, hematocrit 43.9%, WBC 4.06×10^3/µL, and thrombocytes 101×10^3/µL.

COMMENTS

An estimated 1:13,000 Asian populations have choledochal cysts, a congenital condition marked by bile duct dilatation that manifests in pediatric patients as jaundice, abdominal lumps, and abdominal pain.[1,2] The distal biliary obstruction was common in patients with severe choledochal cysts, resulting in bile stasis and subsequent serious complications including acute cholangitis, abnormal liver function, as well as choledochal cyst perforation and coagulopathy.[1] Choledochal cysts are typically diagnosed in children. Multimodality imaging methods such as CT, MRI, ultrasound, and MRCP are commonly used to make the diagnosis.[3]

Portal hypertension is a recognized consequence of a delayed diagnosis of a choledochal cyst and can be associated with secondary biliary cirrhosis, extrahepatic portal venous thrombosis, or compression of the cyst over the portal vein.[4] This case report describes a child with portal hypertension after a choledochal cyst.

The majority of choledochal cysts have been proposed to be congenital in origin. A section of the bile duct may dilate abnormally in choledochal malformations, but the bile duct can still secrete. According to the Todani classification, it is estimated that extrahepatic dilatation with cystic morphology accounts for about 80% of choledochal cysts.[5] Chodedochal cysts appear in five forms, including type I to type 5. Cystic dilatation of the bile duct is a characteristic of type I cysts, which account for 50% to 80% of choledochal cysts. Two percent of choledochal cysts are type II cysts. Between 1.4% and 4.5% of choledochal cysts are type III cysts. Between 15% and 35% of choledochal cysts are type IV cysts. Twenty percent of choledochal cysts are type V cysts, frequently referred to as Caroli's disease.[6]

The patient was jaundiced from one week of age and persisted until more than one month of age, but was considered physiological jaundice. At that time, the patient was suspected of Hirschsprung due to a distended abdomen. The persistence of jaundice until three months of age led to a referral for further examination. Choledochal cysts present with multiple complaints, mostly (>50%) with abdominal pain and jaundice, followed by episodes of pancreatitis, palpable abdominal mass, vomiting, fecal acholia, and asymptomatic. The classic triad of the disease (jaundice, abdominal pain, and palpable abdominal mass) was present in less than 5% of cases.[7] This condition usually presents in a non-specific form. Evaluating patients who show jaundice, abdominal pain, and a palpable abdominal mass requires healthcare teams to have a high index of suspicion regarding choledochal cysts. Because of the ambiguity of these symptoms and physical findings, appropriate imaging studies are essential for diagnosing choledochal cysts.[6] Choledochal cysts are diagnosed primarily by radiological imaging, and proper management requires full anatomical localization of the cyst.[8] In this case, abdominal CT revealed a ±6.3×8.3×13.6 cm cystic lesion in the CBD with a dilated biliary tree supporting the diagnosis of choledochal cyst,

while abdominal ultrasound examination revealed an anechoic lobulated lesion in the right lobe of the liver in the porta hepatis area.

Choledochal cysts with a high risk of malignant transformation, such as types I and IV, require total cyst excision, and management usually depends on cyst classification.[8] To ensure a safe anastomosis and lower the risk of postoperative complications, the bile-enteric anastomosis must be carried out in the dilated lower section of the common hepatic duct.[7] In this case, the Lilly procedure was carried out using Roux-en-Y hepaticojejunostomy anastomosis following cyst excision. After choledochal cyst excision, the five-year overall survival rate is 95.5%.[8] However, two years after excision, the patient experienced gastrointestinal bleeding such as blackish stools that required blood transfusion, splenomegaly, and signs of hypersplenism such as anemia and thrombocytopenia. A recognized consequence of a delayed choledochal cyst diagnosis is portal hypertension. Although the exact cause of portal hypertension in choledochal cysts remains to be determined, it may be associated with extrahepatic portal venous thrombosis, secondary biliary cirrhosis, or compression of the cyst over the portal vein.[4] By extrinsic compression, choledochal cysts may result in esophageal varices, portal hypertension, and partial or total portal vein blockage. In addition to the narrowing of the cholangiointestinal anastomosis or stricture because of residual cysts, improper treatment of choledochal cysts can result in complex complications including gallstones, liver cirrhosis, cancer, or inflammation of the bile duct.[8,9]

A frequent consequence of cirrhotic chronic liver disease and liver vascular occlusion in children is portal hypertension.[10,11] Although the liver biopsy at 6 months of age showed liver cirrhosis, there is currently no hepatomegaly with normal liver function tests, such as albumin and coagulation tests within normal limits.

The development of portal hypertension is primarily caused by two pathogenetic factors: elevated portal blood flow and elevated portal vascular resistance.[10] Gastrointestinal bleeding has a high morbidity rate and is perceived by patients and caregivers as a frightening event that gives the impression of impending death, even though it frequently resolves on its own in children. One of the most frequent reasons for liver transplantation is portal hypertension and its complications.[10]

Subsequently, severe stenosis of the main portal accompanied by abnormalities and thickening of the portal branches was identified on imaging. Pathological examination of the liver showed extrahepatic cholestasis with extensive fibrosis, but there was no hepatomegaly on physical examination and liver function tests were within normal limits. Therefore, the final diagnosis was portal hypertension secondary to primary portal stenosis due to a choledochal cyst. Portal hypertension may be present to some extent in patients with choledochal cysts. To take timely preventive action to avoid life-threatening portal hypertension manifestations, evaluation can be performed via portal pressure measurement and liver histological examination.[12]

In conclusion, portal hypertension may occur as a late complication of choledochal cysts. Timely preventive management is necessary to improve the outcome of children with choledochal cysts.

ΠΕΡΙΛΗΨΗ

Δευτεροπαθής πυλαία υπέρταση λόγω κύστης χοληδόχου πόρου

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Η κύστη του χοληδόχου είναι μια συγγενής πάθηση σε βρέφη και παιδιά που χαρακτηρίζεται από διάταση των χοληφόρων αγωγών, η οποία εκδηλώνεται με ίκτερο, κοιλιακή μάζα και κοιλιακό άλγος. Οι κύστεις αυτές μπορεί να οδηγήσουν σε επιπλοκές, όπως κίρρωση και πυλαία υπέρταση και είναι απαραίτητη η έγκαιρη ανίχνευση. Αναφέρεται η περίπτωση ενός αγοριού 2 ετών και 6 μηνών με χοληδόχο κύστη τύπου Ι που υποβλήθηκε σε εκτομή σε ηλικία 6 μηνών. Δύο έτη αργότερα ο ασθενής παρουσίασε αιμορραγία από κιρσούς οισοφάγου, σπληνομεγαλία και υπερσπλη-

νισμό. Στην αξονική αγγειογραφία παρατηρήθηκε σοβαρή στένωση της πυλαίας φλέβας σε συνδυασμό με ανωμαλίες και πάχυνση των κλάδων της πυλαίας. Η οισοφαγοδωδεκαδακτυλοσκοπική εξέταση ανέδειξε κιρσούς οισοφάγου βαθμού ΙΙ. Ο ασθενής διαγνώστηκε με πυλαία υπέρταση δευτερογενώς λόγω της χοληδοχοκύστης. Μια όψιμη συνέπεια των χοληδοχοκύστεων είναι η πυλαία υπέρταση και γι' αυτόν τον λόγο πρέπει να λαμβάνεται υπ' όψιν ο μακροπρόθεσμος κίνδυνος ηπατικής ίνωσης και πυλαίας υπέρτασης. Η βελτίωση της πρόγνωσης των παιδιών με χοληδοχοκύστεις απαιτεί έγκαιρη προληπτική φροντίδα.

Λέξεις ευρετηρίου: Ιατρική, Κίρρωση ήπατος, Κύστη χοληδόχου, Παιδί, Πυλαία υπέρταση

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