—Case Reports—

Choledochal Cyst Diagnosed during Pregnancy and Treated by Postpartum Laparoscopic Choledochal Cyst Excision: Case Report

Tetsuya Shimizu, Yoshiharu Nakamura, Akira Matsushita, Masato Yoshioka, Tomohiro Kanda, Yoshiaki Mizuguchi and Hiroshi Yoshida

Department of Gastrointestinal and Hepato-Biliary-Pancreatic Surgery, Nippon Medical School, Tokyo, Japan

We report a rare case of choledochal cyst with acute cholangitis that was diagnosed at 37 weeks' gestation and treated by laparoscopic choledochal resection and biliary reconstruction after delivery. A 31-year-old Japanese primigravida at 37 weeks' gestation presented with right upper quadrant pain. The patient was diagnosed as having acute cholangitis due to a type-Ia choledochal cyst, according to the Todani classification, with pancreaticobiliary maljunction. Acute cholangitis improved with conservative treatment, the fetus was delivered by Cesarean section at 38 weeks' gestation, and the patient was treated by laparoscopic choledochal cyst excision and biliary reconstruction at 47 days postpartum. Total operation time was 579 minutes and intraoperative body fluid loss was 100 mL. The patient is now healthy with normal liver function 7 years after the operation. To ensure good outcomes for the mother and fetus, treatment decisions for choledochal cyst diagnosed during pregnancy must be carefully considered. (J Nippon Med Sch 2024; 91: 417–421)

Key words: choledochal cyst, pregnancy, laparoscopic surgery

Introduction

Choledochal cyst (CC) is often overlooked and left untreated until adulthood and is sometimes diagnosed in patients presenting with abdominal pain, fever, or jaundice. Although CC is a benign condition, it is frequently associated with pancreaticobiliary maljunction and can cause cholangitis and pancreatitis. Because patients with CC have a high risk of biliary tract carcinogenesis, choledochal resection and biliary reconstruction should be performed early. We report a rare case of a CC with acute cholangitis diagnosed at 37 weeks' gestation that was treated by laparoscopic surgery after delivery.

Treatment decisions for CC diagnosed during pregnancy are critical in ensuring satisfactory outcomes for the mother and fetus. This is the first reported case of postpartum laparoscopic choledochal resection and biliary reconstruction for a patient with CC diagnosed during pregnancy.

Case Report

A 31-year-old Japanese primigravida at 37 weeks' gestation presented with frequent severe right upper quadrant pain and low-grade fever and was referred to our hospital for detailed evaluation of the abdominal pain. She reported several incidents per year of right upper quadrant pain and right back pain since childhood. Physical examination showed right upper quadrant abdominal tenderness. Routine laboratory tests revealed a mild inflammatory reaction and liver dysfunction: white blood cell count 4,800/μL, C-reactive protein 0.63 mg/dL, glutamic-oxaloacetic transaminase 39 IU/L, glutamic-pyruvic transaminase 23 IU/L, alkaline phosphatase 484 IU/L, gamma-glutamyl transpeptidase 81 IU/L, total bilirubin 0.6 mg/dL, amylase 137 IU/L. Tumor markers were within normal limits: carcinoembryonic antigen 0.5 ng/mL, carbohydrate antigen 19-9 3.9 U/mL. Ultrasonography revealed an oval dilated common bile duct (diameter 40 mm) containing a common bile stone, without intrahepatic bile duct dilation (**Fig. 1**). Ultrasonography and

Correspondence to Yoshiharu Nakamura, MD, Department of Gastrointestinal and Hepato-Biliary-Pancreatic Surgery, Nippon Medical School, 1–1–5 Sendagi, Bunkyo-ku, Tokyo 113–8603, Japan
E-mail: keishun@nms.ac.jp
https://doi.org/10.1272/jnms.JNMS.2024_91-401
Journal Website (https://www.nms.ac.jp/sh/jnms/)

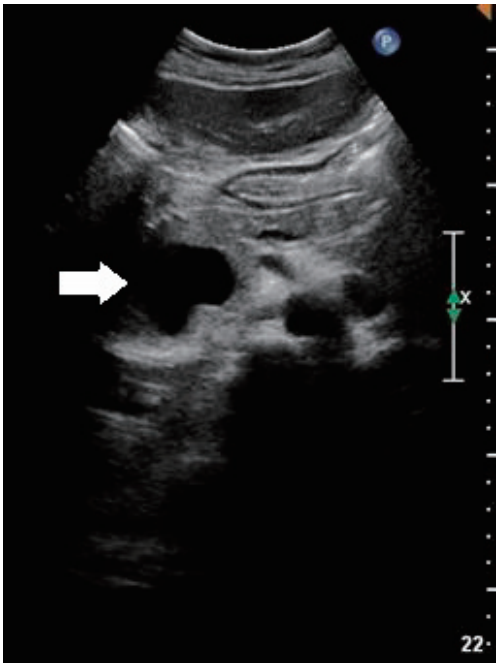

Fig. 1 Ultrasonography revealed a dilated oval common bile duct (diameter 40 mm; white arrow).

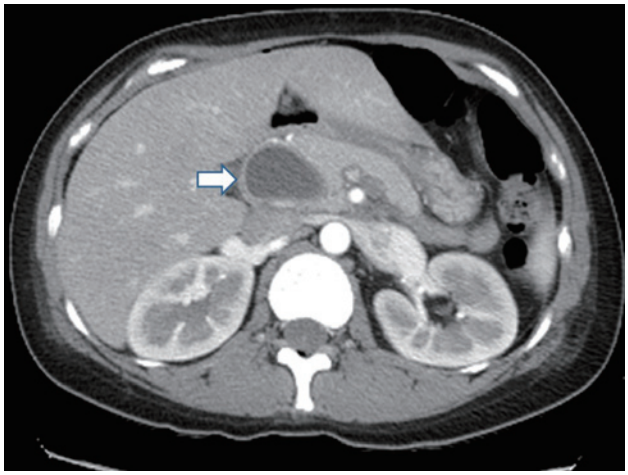

Fig. 2 Abdominal CT showed a choledochal cyst (white arrow) without intrahepatic bile duct dilation.

physical findings strongly suggested CC. Acute cholangitis assessment criteria[1] indicated Grade I (mild) severity, and symptoms were relieved by antibiotics, without intervention for biliary drainage. Because acute cholangitis improved with conservative treatment, evaluation and surgery for CC were postponed until after delivery.

Cesarean section was performed at a gestational age of 38 weeks and 6 days, and the patient delivered a healthy baby. After delivery, computed tomography (CT), endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangiopancreatography (MRCP) were performed. Abdominal CT showed a CC (diameter 40 mm) and no dilation of the intrahepatic bile duct (**Fig. 2**). MRCP revealed a dilated common bile duct containing a common bile stone, without intrahepatic bile duct dilation, and dilation of the common hepatic duct (**Fig. 3 a**). ERCP showed pancreaticobiliary maljunction (**Fig. 3 b**). In accordance with the Todani classification[2], type-Ia CC with pancreaticobiliary maljunction was diagnosed.

After the mother and child were discharged from hospital, laparoscopic CC excision was performed at 47 days after delivery because postpartum abdominal symptoms were stable. Before surgery, the nature, expected benefits, relevant risks, and adverse effects of the operation were carefully explained to the patient, and she provided her informed consent.

The patient was placed in the lithotomy position under general anesthesia, and 4 ports were inserted[3]. Through a 3-cm transumbilical incision, a wound retractor (Alexis Wound Retractor XS, Applied Medical, CA, USA) and surgical glove port with 12-mm trocars attached were inserted as an umbilical port. After incision of a hepatoduodenal ligament, the CC was identified (**Fig. 4**). The common bile duct was dissected away from the surrounding vessels and tissues in the hepatoduodenal ligament and mobilized toward the bifurcation of the common hepatic duct with a 5-mm monopolar L-hook electrode. The common hepatic duct was transected distal to the CC, after which the lower common bile duct was dissected away from the pancreas to identify the inferior end of the CC and the pancreaticobiliary junction behind the duodenum. After the narrow segment below the CC was identified and separated after distal closure with clips, the CC and gallbladder were removed. A Roux limb was created extracorporeally via the umbilical incision and returned to the abdominal cavity. After pneumoperitoneum resumed, the Roux limb was intracorporeally brought up in a retrocolic fashion. An end-to-side hepaticojejunostomy was laparoscopically established with a continuous, single-layer, full-thickness 4-0 polyglactin suture (**Fig. 5**). Two closed suction drains were left behind and above the biliary anastomosis. Total operation time was 579 minutes and intraoperative body fluid loss was 100 mL. On postoperative day 3, the amylase concentration of the intrabdominal drainage was 212,100 IU/L and CT revealed fluid collection between the porta hepatis and pancreas. The patient developed a postoperative pancreatic fistula (International Study Group of Pancreatic Fistula classification: biochemical leak), which improved with conservative treatment. The abdominal drains were removed 14 days after the opera-

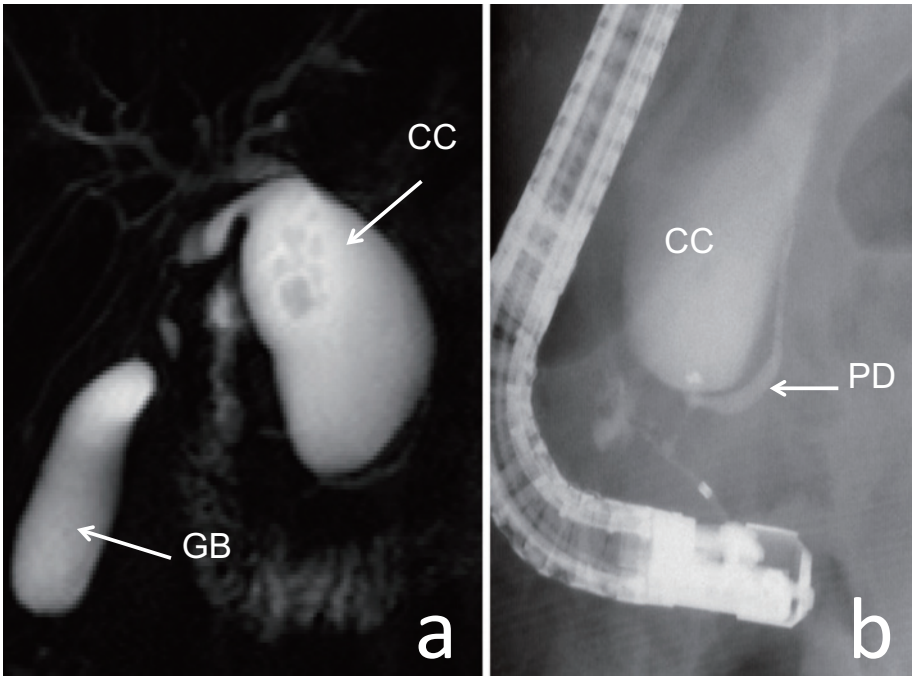

Fig. 3 (a) Magnetic resonance cholangiopancreatography (MRCP) revealed a dilated common bile duct containing a common bile stone. (CC: choledochal cyst, GB: gallbladder)
(b) Endoscopic retrograde cholangiopancreatography showed pancreaticobiliary maljunction. (CC: choledochal cyst, PD: pancreatic duct)

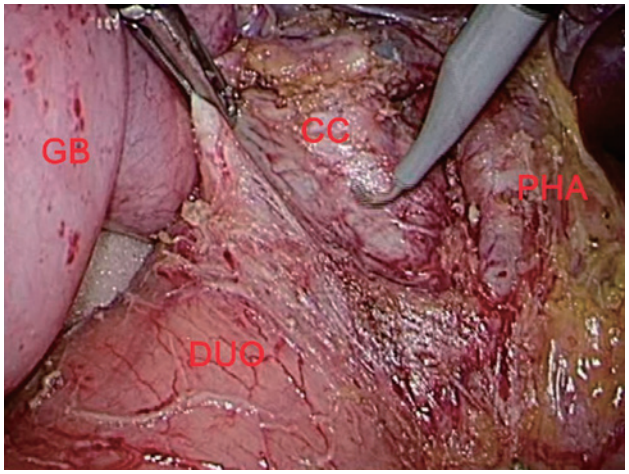

Fig. 4 The hepatoduodenal ligament was incised and a choledochal cyst was identified. (CC: choledochal cyst, GB: gallbladder, PHA: proper hepatic artery, DUO: duodenum)

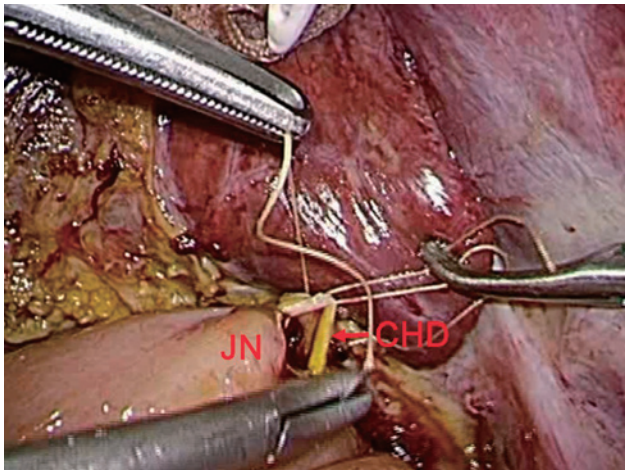

Fig. 5 An end-to-side hepaticojejunostomy was intracorporeally established. (CHD: common hepatic duct, JN: jejunum)

tion.

The patient was discharged from hospital on the 22nd postoperative day. The CC was histologically diagnosed as non-malignant. At 7 years postoperatively she is healthy, with normal liver function; MRCP showed no stenotic hepaticojejunostomy (**Fig. 6**).

Discussion

Both CC with pancreaticobiliary maljunction and pregnancy are associated with several complications, including acute pancreatitis and cholangitis. Some authors have reported cases of CC during pregnancy in women who developed fatal cholangitis[4]. CC are frequently associated with pancreaticobiliary maljunction. The cause of CC is unknown; however, it is believed that an abnormal junc-

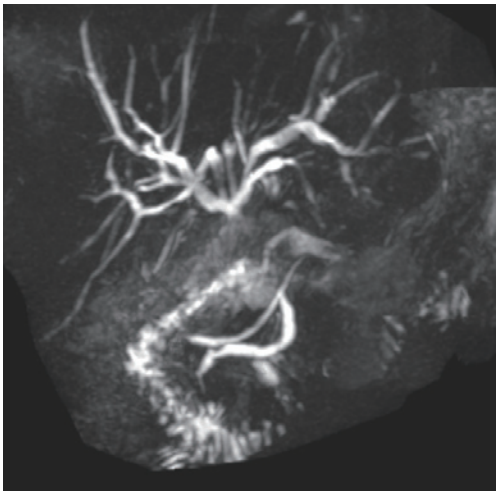

Fig. 6 At 7 years postoperatively, MRCP showed no stenotic hepaticojejunostomy.

tion of the bile duct and pancreatic duct forms a common channel that allows reflux of pancreatic juice into the bile duct, and that the action of the pancreatic enzymes on the bile duct wall causes acute cholangitis, choledochal dilatation, and biliary tract carcinogenesis[5]. Furthermore, pregnancy directly affects excretion of pancreatic juice and bile, because of the increase in intra-abdominal pressure due to enlargement of the uterus, and promotes production of biliary sludge and gallstones by biliary dyskinesia[6,7]. Although the reason for the association of CC and biliary cancer is unclear, possible causes of carcinogenesis include reflux of pancreatic enzymes into the bile duct, bile stasis, and repeated infection. Complete resection of the CC and gallbladder is recommended to decrease malignant potential.

In a patient with acute cholangitis or pancreatitis with CC during pregnancy, the treatment strategy must consider the optimal treatment for cholangitis and pancreatitis, the continuation of the pregnancy, and the timing of radical surgery for CC. Jaundice and sepsis frequently coexist with acute cholangitis, and this has a considerable effect on and can potentially disrupt pregnancy. Thus, it is vital to correctly assess the severity of acute inflammation and establish a proper treatment policy[8]. In pregnant women with acute cholangitis accompanied by CC, the first consideration is to administer antibiotics and ascertain the necessity for biliary drainage[9]. However, opinions differ on the timing of radical surgery for CC in pregnancy. When a pregnancy enters the second trimester, it is generally stable and the operation is less risky for the fetus and mother[10]. Jabbour et al.[11] reported that major hepatobiliary surgery was possible after the second trimester, and they safely performed CC excision and Roux-en Y hepaticojejunostomy under laparotomy for CC complicated with acute cholangitis at 28 weeks' gestation. However, interventions such as endoscopic biliary drainage and percutaneous transhepatic biliary drainage allow for non-surgical control for most cases of acute cholangitis. Radical surgery for CC is a complicated procedure that includes biliary reconstruction and choledochal resection, and there is a risk of postoperative complications such as bile leakage. We therefore avoid this operation during the second and third trimesters but perform it as soon as possible after delivery. The optimal timing of CC excision after delivery remains controversial; however, previous studies reported CC excision after 2 months[8,9]. Nasu et al.[9] described spontaneous rupture of the CC and bile peritonitis, which required urgent surgical management during pregnancy.

CC can spontaneously rupture during labor, because of the elevated intrabdominal pressure. Therefore, proper selection of the mode of delivery is critical for the patient and fetus[9]. To avoid rupture of a CC during labor, Cesarean section is often selected[12].

In our patient, acute cholangitis with CC was diagnosed at 37 weeks' gestation and successfully treated by intravenous antibiotic therapy without biliary drainage before delivery. The fetus was delivered by Cesarean section at 38 weeks' gestation, and the patient was treated by laparoscopic CC excision and biliary reconstruction at 47 days postpartum. Before, during, and after delivery, treatment decisions for CC should be carefully considered, to ensure optimal outcomes for the mother and fetus. Although CC are rare during pregnancy, obstetricians and surgeons should be familiar with the condition to ensure prompt diagnosis and adequate definitive management[9].

Although postpartum CC resection and biliary reconstruction has been reported[12–14], a search of PubMed reveals that postpartum laparoscopic CC resection has not been reported.

Conflict of Interest: None declared.

References

1. Kiriyama S, Takada T, Strasberg SM, et al. New diagnostic criteria and severity assessment of acute cholangitis in revised Tokyo Guidelines. J Hepatobiliary Pancreat Sci [Internet]. 2012 Sep;19(5):548–56. Available from: http://www.ncbi.nlm.nih.gov/pubmed/22825491
2. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer

arising from choledochal cyst. Am J Surg [Internet]. 1977 Aug;134(2):263–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/889044
3. Shimizu T, Nakamura Y, Yoshioka M, Mizuguchi Y, Matsumoto S, Uchida E. Laparoscopic resection of choledochal cyst: report of a case. J Nippon Med Sch [Internet]. 2013;80(2):160–4. Available from: http://www.ncbi.nlm.nih.gov/pubmed/23657070
4. Wu DQ, Zheng LX, Wang QS, Tan WH, Hu SJ, Li PL. Choledochal cysts in pregnancy: case management and literature review. World J Gastroenterol [Internet]. 2004 Oct;10(20):3065–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/15378796
5. Kawarada Y, Das BC, Tabata M, Isaji S. Surgical treatment of type IV choledochal cysts. J Hepatobiliary Pancreat Surg. 2009;16(5):684–7.
6. Ko CW, Beresford SA, Schulte SJ, Matsumoto AM, Lee SP. Incidence, natural history, and risk factors for biliary sludge and stones during pregnancy. Hepatology [Internet]. 2005 Feb;41(2):359–65. Available from: http://www.ncbi.nlm.nih.gov/pubmed/15660385
7. Li HP, Huang YJ, Chen X. Acute pancreatitis in pregnancy: a 6-year single center clinical experience. Chin Med J (Engl) [Internet]. 2011 Sep;124(17):2771–5. Available from: http://www.ncbi.nlm.nih.gov/pubmed/22040440
8. Wig JD, Goenka MK, Chawla YK, Nagi B. Cholangitis secondary to choledochal cyst in pregnancy and puerperium. J Clin Gastroenterol [Internet]. 1997 Sep;25(2):489–91. Available from: http://www.ncbi.nlm.nih.gov/pubmed/9412960
9. Nasu K, Matsuki S, Kawano Y, Miyakawa I, Nakashima K, Anai H. Choledochal cyst diagnosed and conservatively treated during pregnancy. Am J Perinatol [Internet]. 2004 Nov;21(8):463–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/15580542
10. Conway WC, Campos GM, Gagandeep S. Choledochal cyst during pregnancy: the patient's first pregnancy was complicated by a congenital anomaly. Am J Obstet Gynecol [Internet]. 2009 May;200(5):588.e1–2. Available from: http://www.ncbi.nlm.nih.gov/pubmed/19375577
11. Jabbour N, Brenner M, Gagandeep S, et al. Major hepatobiliary surgery during pregnancy: safety and timing. Am Surg [Internet]. 2005 Apr;71(4):354–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/15943413
12. Martinez-Ordaz JL, Morales-Camacho MY, Centellas-Hinojosa S, Roman-Ramirez E, Romero-Hernandez T, de la Fuente-Lira M. [Choledochal cyst during pregnancy. Report of 3 cases and a literature review]. Cir Cir [Internet]. 2016 Mar-Apr;84(2):144–53. Available from: https://www.ncbi.nlm.nih.gov/pubmed/26477375. Spanish.
13. Furuhashi S, Takamori H, Nakahara O, et al. Choledochal cyst during pregnancy: case report and literature review of treatment. Clin J Gastroenterol [Internet]. 2013 Aug;6(4):326–8. Available from: https://www.ncbi.nlm.nih.gov/pubmed/26181738
14. Jia B, Tan L, Jin Z, Liu Y. Duel-stage treatment for biliary cysts with cholangitis during pregnancy. Pak J Med Sci [Internet]. 2017 Mar-Apr;33(2):483–7. Available from: https://www.ncbi.nlm.nih.gov/pubmed/28523061

(Received, October 21, 2022)
(Accepted, March 16, 2023)