CASE REPORT

Choledochal cyst in pregnancy

Koh KS, Bickle I, Mathew VV, Chong VH

Koh KS, Bickle I, Mathew VV, et al. Choledochal cyst in pregnancy. *Malays Fam Physician.* 2016;11(2 & 3);27–29.

Keywords:
Biliary disorder, pregnancy, cystic disease, cholangitis, obstructive jaundice

Authors:

Vui Heng Chong
(Corresponding author)
Department of Medicine, RIPAS Hospital, Bandar Seri Begawan, Brunei Darussalam.
E-mail; chongvuih@yahoo.co.uk/ cvhlag@gmail.com

Kai Shing Koh
Department of Surgery, RIPAS Hospital, Bandar Seri Begawan, Brunei Darussalam

Ian Bickle
Department of Radiology, RIPAS Hospital, Bandar Seri Begawan, Brunei Darussalam

Vallikad Varkey Mathew
Department of Surgery, RIPAS Hospital, Bandar Seri Begawan, Brunei Darussalam

Abstract

Introduction: Biliary diseases during pregnancy are not uncommon and are frequently due to cholelithiasis. Choledochal cyst during pregnancy is rare. The management of biliary pathologies during pregnancy poses a challenge as the window of opportunity to carry out any interventions with minimal risk is small.
Case report: We report the case of a lady who was diagnosed with a large type I choledochal cyst during the second trimester of pregnancy. Due to increasing symptoms and recurrent cholangitis, she was managed with antibiotics and surgical resection. She remained well and delivered a healthy baby boy.
Conclusion: Clinicians need to consider choledochal cyst as a differential in pregnant patients presenting with upper abdominal fullness.

Introduction

Biliary disease during pregnancy is not uncommon and is frequently due to stone disease.[1] Although reported, choledochal cyst during pregnancy is rare.[2–4] Given that radiological investigations are not frequently performed during pregnancy, diagnosis of choledochal cyst is often delayed. The management of biliary pathologies during pregnancy poses a challenge as the window of opportunity to carry out any interventions with minimal risk is small. It is limited to the second trimester when the risk of inducing miscarriage or causing premature labour is less.

Case summary

A lady, in her mid-twenties (G_1P_0), in the second trimester was referred from the peripheral clinic for the evaluation of intermittent abdominal discomfort for the past 4 weeks. This was associated with some nausea but no other abnormalities, such as fever or darkening of urine, were observed. Ultrasound scan (USS) at the peripheral clinic had detected a large cystic lesion in the right upper quadrant area. The gravid uterus was normal. Prior to this, her antenatal care had been uneventful and her pregnancy had progressed satisfactorily. She denied having any abdominal symptoms prior to her pregnancy. There was no significant personal past medical or family history. On examination, she was well and comfortable with a gravid uterus appropriate for the gestational age and fullness in the right upper quadrant and epigastrium that was mildly tender on deep palpation.

A repeat USS in the hospital confirmed the presence of a large cystic mass and it was not possible to locate the gallbladder or assess the relation to other structures. Her liver profiles showed mild cholestasis (serum ALP 234 IU, GGT 250 IU and bilirubin 24 mmol/L). A magnetic resonance cholangiopancreatography showed a large cystic dilatation (9 × 11 cm) in the right upper abdomen of biliary origin (**Figure 1A and B**). Based on these findings, type I choledochal cyst was diagnosed.

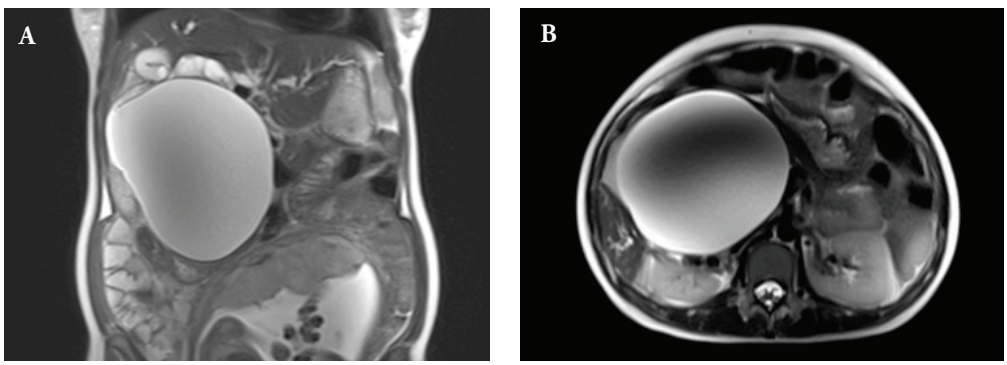

Figure 1. (A) MRI scan showing a large cystic structure in the upper right abdomen next to the gravid uterus, and (B) axial image showing the large round cystic mass; type I choledochal cyst

Management options that included endoscopic drainage (endoscopic retrograde cholangiopancreatography [ERCP] and stenting), percutaneous drainage and surgery were discussed. However, the patient's symptoms improved with medical treatment and the patient opted for conservative management. During the clinical review, the patient continued to have intermittent abdominal pain and fever. A repeat liver profiles showed worsening cholestasis with elevated bilirubin. An endoscopy (ERCP) with appropriate lead shielding was attempted, but failed due to the gravid uterus and large cyst displacing the duodenum. In view of the massive size of the cyst posing a risk to pregnancy (i.e. premature delivery, cyst rupture and also the risk of cholangitis) and after a detailed discussion with the patient and husband, it was decided to proceed with surgical resection of the cyst. She underwent surgical excision of the choledochal cyst, cholecystectomy and formation of a Roux-en-Y hepaticojejunostomy without immediate complication to the patient or pregnancy. The resected specimen showed an inflamed biliary tree but no evidence of cholangiocarcinoma. Postsurgery, her pregnancy progressed without any problem and she had a normal uncomplicated delivery of a healthy baby boy. The patient remained well in the following 2 years follow-up and went back to her country of origin.

Discussion

Choledochal cyst is a rare congenital dilatation of the biliary system and is classified into four types: type I—fusiform dilatation of the common bile duct; type II—a diverticulum of the extrahepatic duct; type III—cystic dilation of the intraduodenal portion of the bile duct; and types IV, type IVa—intrahepatic cystic dilatations and IVb—affecting the extra and intrahepatic ducts.[5]

The exact pathogenesis of this condition is unknown. However, it is widely believed that the presence of a long common channel with an abnormal pancreaticobiliary junction results in the chronic reflux of pancreatic enzymes into the biliary tree. This results in the damage and progressive dilatation of the biliary tree.[5] However, this does not explain the various manifestations and the pathogenesis remains debated.

Choledochal cyst presenting during pregnancy is rare.[2–4,6,7] Unsuspected, the diagnosis will be delayed and this can lead to pancreatitis, ascending cholangitis, biliary cirrhosis and even cyst rupture with resultant dire foetal and maternal outcomes.[2–4,6–9] During pregnancy, the radiological imaging is often limited to USS and magnetic resonance imaging (MRI). USS features of choledochal cysts depend on the stage of the disease. Early cyst formation may resemble a dilated common ductal system as in biliary obstruction. However, as in most of the cases that present with symptoms like our case, the cystic dilatations are typically large. In some cases, continuity with the biliary system can be appreciated. However in large choledochal cysts, the appreciation of communication with the biliary tree may not be possible due to the mass compressive effects. However, any cystic dilatation in the epigastric or right upper quadrant in the location of the biliary system should heighten the suspicion of a choledechol cyst.[10] MRI is the preferred imaging modality when the diagnosis is in doubt as it is able to better demonstrate the anatomy of the biliary tree and its relations with the choledochal cyst. However, MRI is not widely available.

The management of choledochal cyst in pregnancy is challenging as there are risks to mother and foetus.[3] Given that the treatment period window is limited, an important aspect of the management is early recognition and diagnosis so that the management can be planned. Generally once diagnosed, a conservative approach is usually adopted as far as possible, and if the symptoms become progressive or cyst-related complications arise, other options need to be considered. Endoscopic intervention is the primary option to consider and radiation exposure should be minimised by limiting fluoroscopy time and the use of lead shielding. Endoscopic ultrasound guidance that avoids radiation exposure is another good and preferred option.[11] Percutaneous drainage of the choledochal cyst has been reported to be a useful alternative.[7] Surgery is usually avoided in the first trimester unless the symptoms become life threatening. Ideally, surgery should be performed in the second trimester when the surgical and anaesthetic risks are at the lowest. Due to the possibility of cyst rupture from high intra-abdominal pressure during labour, an elective caesarean section is generally recommended when the foetus is sufficiently matured and definitive surgical excision of the cyst should be planned given the increased risk of biliary tract carcinoma in choledochal cysts.

In conclusion, choledochal cyst in pregnancy is rarely encountered. Delay in appropriate management may lead to complications in both the mother and developing foetus. A conservative management may be appropriate during pregnancy; however, due to the increased risk carcinoma, excision of the cyst should be considered after pregnancy.

References

1. Mackillop L, Williamson C. Liver disease in pregnancy. *Postgrad Med J*. 2010;86(1013): 160–4. doi: 10.1136/pgmj.2009.089631.
2. 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*. 2009;200(5):588. e1–2. doi: 10.1016/j.ajog.2009.01.026.
3. Hewitt PM, Krige JE, Bornman PC, et al. Choledochal cyst in pregnancy: a therapeutic dilemma. *J Am Coll Surg*. 1995;181(3):237–40. [PMID: 7670683].
4. Nassar AH, Chakhtoura N, Martin D, et al. Choledochal cysts diagnosed in pregnancy: a case report and review of treatment options. *J Matern Fetal Med*. 2001;10(5):363–5. [PMID: 11730504].
5. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. *Can J Surg*. 2009;52(5):434–40. [PMID: 19865581].
6. Nasu K, Matsuki S, Kawano Y, et al. Choledochal cyst diagnosed and conservatively treated during pregnancy. *Am J Perinatol*. 2004;21(8):463–8. doi: 10.1055/s-2004-835963.
7. Lipton A, Carlan SJ, Teixeira A, et al. Biliary cyst in pregnancy managed by percutaneous drainage: a case report. *J Reprod Med*. 2013;58(7–8):357–60. [PMID: 23947090].
8. Joseph P, Raju RS, Vyas FL, et al. Spontaneous perforation of choledochal cyst and hyperemesis gravidarum. *Trop Gastroenterol*. 2008;29(1) :46–7. [PMID: 18564670].
9. Wig JD, Goenka MK, Chawla YK, et al. Cholangitis secondary to choledochal cyst in pregnancy and puerperium. *J Clin Gastroenterol*. 1997;25(2):489–91. [PMID: 9412960].
10. Lewis VA, Adam SZ, Nikolaidis P, et al. Imaging of choledochal cysts. *Abdom Imaging*. 2015; 40(6):1567–80. doi: 10.1007/s00261-015-0381-4. [PMID: 2 5682292].
11. Lee JJ, Lee SK, Kim SH, et al. Efficacy and safety of pancreatobiliary endoscopic procedures during pregnancy. *Gut Liver*. 2015;9(5):672–8. doi: 10.5009/gnl14217. [PMID: 2 6087783].