CASE REPORT

Giant choledochal cyst presenting during third trimester of pregnancy

Jesus Morales-Maza,¹ Jorge Humberto Rodríguez-Quintero,¹ Sonia Cortés-Vázquez,² Luis Cruz-Benítez²

¹General Surgery, Instituto Nacional de Ciencias Médicas y Nutrición "Salvador Zubirán", Mexico City, Mexico
²General Surgery, Hospital Regional de Alta Especialidad de Ixtapaluca, Ixtapaluca, Mexico

Correspondence to
 Dr Jorge Humberto Rodríguez-Quintero, huroqu90@gmail.com

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SUMMARY

Biliary cystic disease is a rare entity. Twenty-five per cent of cases are diagnosed during adulthood and only a few reports have described this condition during pregnancy, where it represents a therapeutic challenge for both obstetricians and surgeons with regard to the risks it entails for the patient and the fetus. Definitive management is surgical resection, as cysts may progress to malignancy if untreated. During pregnancy, resection is generally deferred to after delivery, especially in the context of suspected cholangitis. A 19-year-old young woman with no previous prenatal control, presented to the emergency department on her 32nd week of gestation with abdominal pain and jaundice. A giant Todani I biliary cyst was observed on imaging along with dilation of the proximal biliary tree suggesting acute cholangitis. Fetal compromise prompted immediate delivery after which percutaneous biliary drainage was performed. Following recovery, the cyst was surgically resected.

BACKGROUND

Choledochal cysts are uncommon congenital dilations of the biliary tree. They represent <1% of all benign bile duct pathology^{1,2} and are associated with several other malformations of the bile duct system. Typically, with an incompetent pancreatic duct sphincter that favours reflux of activated exocrine enzymes into the proximal biliary tree, causing cellular damage and chronic inflammation which have been shown to confer a 10% lifetime risk of progression to malignancy.³

The reported incidence of these lesions is quite variable, ranging from 1–50 000 to 1–200 000 live births. They are more frequently found in women, with a female to male ratio of 4:1 and in Asian population for unknown reasons.^{4,5}

Usual clinical presentation consists of mild right upper quadrant pain, jaundice and a growing abdominal mass, with the complete triad being present only in 10%–17% of patients. However, such findings may be inconsistent, especially in patients who remain undiagnosed for prolonged periods of time and in those with limited access to health services in low-income and middle-income countries, who may not seek medical attention until severe symptoms develop.

An even more unconventional presentation of this entity, which has been seldom described in the literature is during pregnancy, where symptoms

may remain unapproached, masked by the physiological growth of the uterus until obvious signs of complicated disease arise.⁶ It is widely known that normal pregnancy induces several modifications to the liver and biliary function such as increased bile acid secretion, with a decreased ratio of chenodeoxycholic acid to cholic acid fostering the formation of sludge and biliary stones, decreased gallbladder emptying derived from increased progesterone levels and progressive compression of the intraperitoneal contents by the growing uterus. Each one of these factors can potentially contribute to the development of obstructive complications and may explain the onset of symptoms during this period.⁷ Rupture of an enlarged cyst is the most feared complication in this scenario, and is clearly associated with poor outcomes for both the mother and the fetus. In the context of impending rupture, biliary decompression may represent a life saving manoeuvre.^{8–10}

In 1959, Alonso-Lej *et al* proposed the first classification for choledochal cysts¹¹ which was modified by Todani *et al* in 1977.¹² Currently, Todani's is still the most used classification in cystic disease of the biliary tree. It describes five classes of cysts: type I is the most common (80%–90% of the cases) and involves a fusiform dilation of the common bile duct; type II is an isolated diverticulum protruding from the common bile duct; type III is called 'choledocoele' and consists of dilation arising from the duodenal portion of the common bile duct (where the pancreatic duct meets); type IV consists of multiple dilations of the intrahepatic and extrahepatic biliary tree and type V, also known as Caroli's disease, is defined as multiple dilations of the intrahepatic ducts.

CASE PRESENTATION

We present the case of a 19-year-old primigravid woman with no relevant familiar, obstetric or medical history. Her social history was significant for being single, unemployed, having low educational status and limited access to health services. The patient presented for the first time to our hospital at her 32nd week of gestation referring no previous prenatal care and a progressive 20-day-long epigastric abdominal pain which was initially intermittent and radiated to the back and right shoulder. The pain was oppressive in character and was precipitated by eating greasy foods at the beginning but had lately become constant and

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progressively increased in intensity. Five days before attending the emergency department, the patient also presented nausea, biliary vomit, chills and mild jaundice for which she had not sought medical attention elsewhere. She finally decided to come to the hospital because she noticed decreased fetal motility.

On arrival, the patient was found afebrile, with vital signs within normal range, a uterine fundus height measuring 28 cm from the pubic symphysis and right upper quadrant pain on palpation. Fetal heart rate was present but no fetal movements were felt during palpation. The external cervical os was closed, and there were no signs of vaginal bleeding or amniotic fluid leakage. We ordered routine labour blood work obtaining the following results.

INVESTIGATIONS

Initial complete blood count showed normal haemoglobin levels of 12.7 g/dL (normal range 12.0–15.6 g/dL) and leucocytes of $9.2 \times 10^9/L$ (normal range $4\text{--}12 \times 10^9/L$). Liver function tests presented an increased total bilirubin of 2.1 mg/dL (normal range 0.3–1 mg/dL), direct bilirubin of 1.5 mg/dL (normal range 0.3–0.18 mg/dL), indirect bilirubin of 0.6 mg/dL (normal range 0.27–0.82 mg/dL), alkaline phosphatase of 167 U/L (normal range 34–104 U/L), gamma glutamyl transferase of 56 U/L (normal range 9–38 U/L), alanine aminotransferase (ALT) of 38 U/L (normal range 7–52 U/L) and aspartate aminotransferase (AST) 43 U/L (normal range 13–39 U/L) which integrated a cholestatic pattern. The rest of the routine blood work ordered was within normal limits.

We ordered an abdominal ultrasound in which we observed a cystic lesion dependent on the common bile duct measuring approximately 14×12 cm on its longest axis. The lesion could not be precisely defined by ultrasound but we were able to notice biliary sludge and proximal dilation of the biliary tree. These findings led us to suspect the diagnosis of acute cholangitis (criteria B (total bilirubin >2.0 mg/dL) + criteria C (biliary dilation on ultrasound) according to Tokyo Guidelines 2018).¹³

Also, a biophysical profile was performed for fetal assessment obtaining a score of 4/10, consistent with fetal compromise, which prompted the decision of proceeding with delivery via caesarean section. The procedure was done without complications via a lower abdominal Pfannenstiel-Kerr incision obtaining a 2 kg healthy male infant who was admitted to the neonatal intensive care unit for surveillance.

After delivery, MR cholangiopancreatography (MRCP) was performed for better characterisation of the cyst. MRCP revealed a saccular mass dependent on the common biliary duct (measuring $14 \times 12 \times 12$ cm) causing compression over proximal biliary structures. Generalised dilation of the proximal biliary tree and biliary sludge inside the cyst were also documented in the study. The lesion was classified as a Todani type I choledochal cyst (figures 1–5).

DIFFERENTIAL DIAGNOSIS

At presentation, our main differential diagnoses were intrahepatic cholestasis of pregnancy, choledocholithiasis, acute cholangitis, acute fatty liver of pregnancy, viral hepatitis and malignancy. We were not able to rule out those differentials until performing the imaging work-up.

MRCP images suggested that obstruction was being caused by compression of the cyst over the proximal portion of the biliary tree. We concluded that such compression had already been partially relieved at the time of the study by the decreased size of the uterus after caesarean section.

Figure 1 Axial MRI showing Todani I choledochal cyst. Considerable compression on the surrounding structures due to the size of the lesion can be appreciated. Also, biliary sludge is noticed adjacent to the posterior wall of the cyst.

Also as stated, biliary sludge was documented and might have contributed to cholestasis and the development of an ascending cholangitis. Considering the complicated clinical scenario and outweighing the risks, the patient was covered with antibiotic therapy.

TREATMENT

The patient presented appropriate recovery after delivery and once we had characterised the lesion by MRCP, we proceeded

Figure 2 Coronal MRI showing Todani I choledochal cyst and proximal dilation of the intrahepatic bile ducts.

Figure 3 Sagittal MRI of the Todani I choledochal cyst. Biliary sludge is noted inside the lesion. We can also appreciate how the lesion appears to be compressing the proximal common bile duct and its surrounding structures.

to place a percutaneous biliary drain transhepatically in order to decompress the biliary tree. The patient was discharged from the hospital, and after 6 weeks we scheduled her for complete open cyst resection and gastrointestinal reconstruction with Roux-en-Y hepaticojejunostomy.

During the procedure, the patient was approached via an anterior oblique incision in the right upper quadrant. First, we performed mobilisation of the hepatic flexure and a wide Kocher manoeuvre to appropriately expose the posterior part of the duodenum. We proceeded to dissect the anterior aspect of the cyst until we observed narrowing of the common bile duct, marking the caudal limit of the lesion. Then, we decompressed the cyst performing a controlled puncture to allow for better dissection and tissue handling. We ligated and separated the caudal end and then displaced the cyst anteriorly to continue the dissection, finding multiple adhesences. We continued posterior dissection of the lesion, and after successfully dissecting the portal triad, the common bile duct was transected distal to the confluence of the hepatic ducts. A 60 cm limb was brought up in a retrocolic fashion in order to perform the anastomosis and re-establish gastrointestinal tract integrity. We closed the abdominal wall, leaving a Blake drain placed over the surgical field (figures 6 and 7)().

The patient was discharged uneventfully after surgery. Histopathology report dictated a fibrotic cystic wall with inflammatory infiltrate and mild goblet and squamous cell metaplasia over the epithelial lining of the cyst. No dysplastic changes or evidence of malignant disease were seen on the specimen (figure 8).

Figure 4 Three-dimensional MR cholangiopancreatography showing the Todani I choledochal cyst and proximal dilation of the biliary tree.

On postoperative day 9, the patient returned to the hospital with fever and biliary output on the drain. We documented partial dehiscence of the anastomosis and decided to introduce a percutaneous biliary drainage to control the fugue and

Figure 5 Three-dimensional reconstruction of the MR cholangiopancreatography.

Figure 6 The Todani I choledochal cyst during surgical resection.

administer a cycle of antibiotics. She was then discharged 14 days later showing full recovery and no further complications.

OUTCOME AND FOLLOW-UP

After discharge, she gradually returned to baseline function, and we continued ambulatory follow-up with periodic abdominal ultrasounds, liver function tests and tumour markers.

The patient has shown no signs of recurrence, infectious complications or development of malignancy to date.

Figure 7 Controlled puncture of the cyst before excision.

Figure 8 Histopathological image of the cyst wall showing fibrosis and inflammatory infiltrate.

DISCUSSION

Surgical approaches on biliary cystic disease have undergone significant changes over the last century. Techniques previously described included cyst marsupialisation, choledochorrhaphy and internal drainage via cystenterostomy to the duodenum or jejunum. Such approaches have been abandoned because of significant morbidity and mortality. Current treatment of choice for type I biliary cysts is complete resection and reconstruction of the gastrointestinal tract usually with a Roux-en-Y hepaticojejunostomy. This approach aims to prevent cystic recurrence and pancreatic reflux, lowering the risk of progression to malignancy in the long term.^{14 15}

Choledochal cysts represent a challenge for surgeons and are even more complex in the context of pregnancy, where they entail risks for both the mother and the fetus. There are no precise guidelines regarding the management of this condition during pregnancy as its presentation is unusual. However, several different approaches have reported successful outcomes on the literature.^{16 17}

In the context of acute cholangitis, it is currently agreed that biliary drainage (either via a endoscopic, percutaneous transhepatic or percutaneous transcystic approach) and intravenous antibiotic administration followed by deferred definitive resection and reconstruction of the gastrointestinal tract, seem to be a convenient approach.¹⁸ Emergent cyst resection during pregnancy has also been reported in the literature in the context of

Learning points

- ▶ The classic clinical presentation of choledochal cysts is jaundice, abdominal pain and abdominal tumour.
- ▶ Todani's Classification divides choledochal cysts into five types.
- ▶ The diagnosis of biliary cystic disease during pregnancy requires a high index of suspicion. Patients with limited access to healthcare may present with complicated disease.
- ▶ Definitive treatment of choledochal cysts is complete resection with Roux-en-Y hepaticojejunostomy.
- ▶ Several management approaches have been described in the literature in this context. Selecting the most appropriate will depend on the resources available and patient status at diagnosis.

cyst rupture and biliary sepsis, but is generally reserved to very specific scenarios in which other treatment options may not be feasible or available.¹⁹ Still, more evidence is needed to assess the precise long-term outcomes of each one of this strategies.

Contributors Planning of the study was performed by JMM, JHR-Q, SC-V and LC-B. JMM and JHR-Q were responsible of conducting the study and providing insight. The conception of the case was done by JMM and JHR-Q. SC-V and LC-B were responsible for acquiring the images and data. The paper was written by JMM, JHR-Q, SC-V and LC-B. Case analysis and follow-up were done by JMM, JHR-Q, SC-V and LC-B. JMM, JHR-Q, SC-V and LC-B approved the final version of the manuscript.

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