Diagnosis and treatment of a choledochal cyst during pregnancy. A case Report

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Background:
Choledochal cysts are a rare pathology of the biliary tree, more commonly found in females and predominantly presented in childhood. Their occurrence in adulthood, particularly during pregnancy, is rare, often presenting with nonspecific signs and symptoms, which can lead to a delay in diagnosis.
We present the case of a 29-year-old female patient who began experiencing symptoms at 8 weeks of gestation. She was initially misdiagnosed with acute lithiasic cholecystitis. During intervention at 12 weeks of gestation, a Type IVa choledochal cyst was found. Initial management involved decompressing the biliary tract via a T-tube due to the complexity of the case. Subsequently, after two episodes of cholangitis, surgical management was decided before the resolution of the pregnancy, through resection of the choledochal cyst and Roux-en-Y hepaticojejunostomy. Currently, her clinical status is favorable; she completed her pregnancy at term via scheduled cesarean section without apparent maternal-fetal complications.

Keywords: Choledochal cysts.

Choledochal cysts are congenital dilatations of the biliary tree, first described by Vater and Ezler in 1793, with the first case attributed to Douglas in 1852. The initial classification was proposed by Alonso-Lej in 1959 and later modified by Todani in 1977, which is currently the most accepted (1,3).

The risk of developing cholangiocarcinoma in choledochal cysts increases with age and is more frequently observed in Type I (68%) and Type IV (21%) cysts (1), supporting the complete resection upon diagnosis. Various theories have been proposed to explain the pathogenesis of choledochal cysts, with the most accepted being the anomalous union of the pancreatic and biliary ducts outside the duodenal wall, forming a single channel that can reach lengths of 10 to 45 mm. This anomaly causes reflux of pancreatic secretions into the biliary tree, and since the pressure in the pancreatic duct is greater than that in the biliary duct, it leads to activation of pancreatic enzymes, alteration of biliary composition, local inflammation, and damage to the biliary epithelium, resulting in weakening and dilatation of the biliary duct wall (4).
The most commonly used classification is that proposed by Todani and colleagues, which categorizes choledochal cysts into five types:

- Type I: saccular or cystic dilation of the extrahepatic bile duct.
- Type II: choledochal diverticulum.
- Type III: dilation of the intraduodenal portion of the choledochus (choledochocele).
- Type IV A: multiple intrahepatic and extrahepatic cysts.
- Type IV B: multiple extrahepatic cysts.
- Type V: intrahepatic cysts (Caroli's disease).

In general, symptoms are vague; patients present with jaundice and nonspecific abdominal pain. The classic triad of presentation includes abdominal pain (87%), jaundice (57%), and a palpable abdominal mass in the right hypochondrium (17%), which occurs in only 20% of cases (5). Hepatic function tests typically show alterations. The most frequent complications are cholangitis and pancreatitis. Other complications include biliary cirrhosis, portal hypertension, rupture, peritonitis, vesicular or choledocholithiasis, with the most significant risk being malignancy, which increases with age (0.7% in those under 10 years, approximately 7% between 10 and 20 years, approximately 14% over 20 years, and about 50% in those over 50 years) (6).

Diagnosis is based on imaging findings, with

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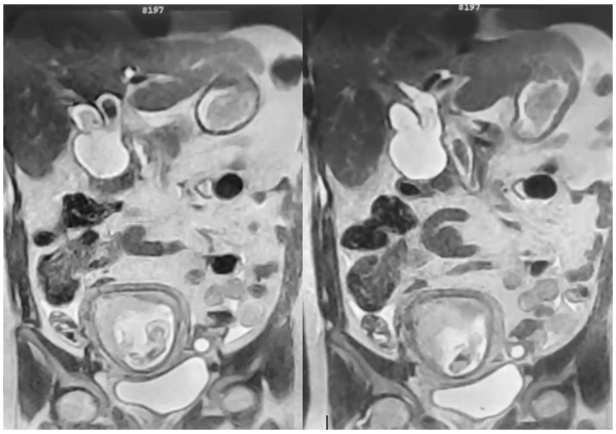

Figure 1. Magnetic resonance image of a Type IVa choledochal cyst.

ultrasound, computed tomography, cholangiography, and magnetic resonance cholangiopancreatography being the most utilized (4). Treatment involves complete cyst resection via open or minimally invasive approach, both showing acceptable results (5).

Various therapeutic alternatives exist, with surgical treatment being the preferred choice.

In this case, we present a 29-year-old female patient diagnosed with a Type IV choledochal cyst according to the Todani classification, initially managed as acute cholecystitis due to the ambiguity of symptoms. Following diagnosis, she was treated with resection and Roux-en-Y biliodigestive diversion, achieving favorable outcomes for both mother and child, with no apparent complications at birth.

Case report

A 29-year-old female patient began experiencing colicky pain localized in the epigastrium, rated 8/10 on the visual analog scale, radiating to the right hypochondrium, accompanied by nausea without vomiting. She reported similar episodes for the past 2 years, denied tea-colored urine, clay-colored stools, and had normal skin and mucosal color. Laboratory tests revealed leukocytes at 16.05, direct bilirubin 0.7 mg/dl, indirect bilirubin 0.3 mg/dl, total bilirubin 1.0 mg/dl, amylase 29 U/L, AST 742 U/L, ALT 494 U/L, ALP 118 U/L, LDH 402 U/L, GGT 317 U/L, and a positive pregnancy immunological test. Her last menstrual period was on 25/11/2023. An ultrasound of the liver and biliary tract reported loss of gallbladder morphology, collapsed at 25 mm with anechoic content and double wall of 3 mm, adjacent collection of 3 cc measuring 22x19 mm, and a choledochus caliber of 2.1 mm, with a diagnosis of 8.2 weeks of gestation.

She showed clinical improvement with medical management and was discharged with a gastroenterology consultation due to suspicion of viral hepatitis etiology from elevated transaminases. She re-entered 3 days later with abdominal pain and increased laboratory figures. Leukocytes were 30.07, direct bilirubin 2.0 mg/dl, indirect bilirubin 0.6 mg/dl, ALT 746 U/L, AST 402 U/L, ALP 235 U/L, GGT 592 U/L. Antibiotic treatment with third-generation cephalosporins was initiated, and laparoscopic cholecystectomy was decided 30 days after the onset of cholangitis symptoms, with intraoperative findings of Type IV Parkland, tortuous cystic duct abnormality. A transcystic cholangiography was performed, observing probable dilation of the choledochus, leading to partial cholecystectomy due to technical difficulties.

Subsequently, in the late postoperative period, she persisted with oppressive epigastric pain radiating in a belt-like fashion, nausea, and vomiting of bilious content, along with leukocytosis, and bilious output through a Penrose drain, prompting reoperation for drainage of a subhepatic collection. A pinpoint leak from the common bile duct was observed, approximately dilated to 3 cm in diameter. Biliary washout was performed with the expulsion of sediment and purulent content, and an 18 Fr Kher tube was placed.

One week later, she presented dysfunction of the T-tube and bilious output through the Penrose drain, with an ultrasound reporting free fluid at segments 5 and 6 infrahepatic, communicating with the Penrose and distal right iliac fossa, measuring 113x60x67 mm with a volume over 300 cc.

ERCP was decided, assuming possible maternal-fetal risks, but it was unsuccessful due to inability to cannulate the bile duct. Finally, at 20 weeks of gestation and with a confirmed diagnosis of choledochal cyst and multiple failed interventions, the patient continued to experience biliary colic and cholangitis, prompting surgical intervention with the aim of biliodigestive diversion, identifying a Type IVa choledochal cyst. Successful Roux-en-Y hepaticojejunostomy was performed.

Resolution of the pregnancy was achieved at term via scheduled cesarean section without apparent complications for both mother and child.

Discussion

As previously mentioned, the symptoms are ambiguous. Patients may present with the classic triad of abdominal pain (87%), jaundice (57%), and a palpable abdominal mass in the right hypochondrium (17%); complications such as cholangitis and pancreatitis may also occur (6). The incidence varies, with reports of one case among 13,000 to 2 million live births, being more frequent in females (4:1) and Asians. They are primarily diagnosed during

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childhood (60%), Types I (50-85%) and IV (30-40%) cysts are the most common (6).

However, the importance of this case lies in the few reported instances during pregnancy, where diagnosis may be delayed due to ultrasound confusion with cholelithiasis and findings of biliary dilation, compounded by limited availability of magnetic resonance imaging in some hospitals. Moreover, in most reported cases, surgical resolution has been implemented after pregnancy resolution. In this case, it was not feasible, necessitating intervention in the second trimester due to the patient's intense symptoms.

DIAGNOSIS

Magnetic resonance cholangio-pancreatography is a non-invasive study and the method of choice for diagnosing and classifying choledochal cysts, with a sensitivity of 96-100%. It effectively detects pancreaticobiliary malunion with sensitivity of 53%-100% and specificity of 90-100%. (12)

Diagnosis during pregnancy is challenging, requiring a high degree of suspicion and auxiliary diagnostic tools. Ultrasound is the most common method, but it can be difficult during pregnancy due to distortion of normal abdominal anatomy and the presence of a gravid uterus (12,14). Studies involving ionizing radiation or contrast, such as computed tomography or endoscopic retrograde cholangiopancreatography, can be performed but with extreme caution. Cholangiography (percutaneous, endoscopic, or nuclear) helps determine the type of cyst, intra- or extrahepatic involvement, and abnormalities of the biliopancreatic junction. Magnetic resonance imaging can provide adequate visualization and relationships between the choledochal cyst (size and extent) and the biliary tree; however, it has limitations in diagnosing small cysts or ductal abnormalities (6,11,12).

Once a diagnosis is made, patients should be referred to a specialized unit, as inappropriate treatment can be catastrophic for the mother and fetus. Pregnant patients represent a special situation. The presence of a cyst in cases of very large lesions or the development of complications is associated with increased maternal-fetal morbidity and mortality (6,8). In symptomatic cases unresponsive to antibiotic treatment, decompression of the cyst via percutaneous transhepatic, endoscopic, or surgical methods should be considered (6,7,8).

Complications associated with procedures must be taken into account, particularly when choosing the treatment. In pregnant patients, management is generally conservative if asymptomatic. The reported mortality associated with surgical treatment in case series is 7% (6). Definitive treatment is surgical, so the timing of the surgical decision must consider the surgical risks for both mother and fetus and the complications of the disease itself. If possible, surgical treatment should be performed in the second trimester, as surgery carries a high risk of morbidity and mortality in pregnant patients (6,9,10). Definitive surgery should be based on the classification of the cyst and the general conditions of the patient. The preferred surgical procedure is Roux-en-Y hepaticojejunostomy due to its lower incidence of cholangitis and need for reoperation. Resection of the cyst is widely accepted for Types I, II, and IV. Type III cysts generally require surgical drainage or endoscopic treatment with sphincterotomy. For Type IV cysts, the approach remains controversial, with options including resection only of the extrahepatic cyst or total cyst resection with hepatectomy. For Type V cysts, some authors recommend hepatic resection. Resection is preferred over cyst diversion due to a higher incidence of cholangiocarcinoma in the cyst remnant (6,7,9).

Conclusion

Choledochal cysts in pregnancy are a rare condition, and the successful outcome for pregnant patients relies on timely diagnosis of the disease, the patient’s general conditions, clinical evolution during monitoring, and deciding the optimal timing for definitive treatment. Patients should be referred to specialized units for comprehensive management. In asymptomatic patients, conservative treatment is widely indicated; however, in those with cholangitis, antibiotic treatment and, if necessary, decompression of the biliary tract via percutaneous transhepatic or endoscopic retrograde cholangiopancreatography may be a temporary measure. The definitive and preferred procedure is cyst resection with biliodigestive diversion via Roux-en-Y during the second trimester of pregnancy or after resolution of the pregnancy. However, during pregnancy, it may be necessary to consider conservative treatment such as percutaneous or endoscopic drainage, always individualizing based on the specific characteristics of the clinical presentation and considering all potential maternal-fetal risks due to the high morbidity and mortality associated with the condition, as well as the likelihood of short- and long-term complications related to the cyst.

Currently, there is no algorithm defining the ideal management for cases like this when clinical manifestation occurs from the first trimester of pregnancy. There should be a multidisciplinary consensus among the specialties of gynecology, endoscopic surgery, hepatobiliary surgery, and radiology to establish appropriate and specific

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management for these clinical scenarios, ensuring the safety of both the pregnant patient and the fetus.

Conflicts of interests

No conflicts of interest relevant to this article.

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