Prenatal diagnosis of a giant choledochal cyst

Yurema González Ruiz*, Paolo Bragagnini Rodriguez, Rafael Leonardo Fernández Atuán, Natalia Álvarez García, Alexander Siles Hinojosa, Natalia González Martínez-Pardo, Juan Elías Pollina

Department of Pediatric Surgery, Miguel Servet University Hospital, Paseo Isabel La Católica s/n, Zaragoza, Aragon, Spain

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A B S T R A C T

A choledochal cyst is a rare and usually benign condition that is diagnosed in 80% of cases in early infancy. Prenatal diagnosis is increasingly identifying the cysts allowing an early surgical treatment with less risk of complications. We report a case of a newborn with prenatal diagnosis of a giant right upper quadrant abdominal cyst. During surgery, an 8 cm choledochal cyst was found. A cholecistectomy, surgical excision of the cyst and formation of a Roux-en-Y hepaticojejunostomy was performed.

A choledochal cyst is a rare congenital cystic dilatation of the extra or intrahepatic biliary tract or both. It was first described by Vater and Ezler in 1723 [1]. They are primarily found in female gender in young children and are more prevalent in East Asian populations. A choledochal cyst can be identified in neonates as a palpable mass, with cholestasis or with jaundice. An early prenatal diagnosis of the choledochal cyst is essential for an earlier surgical repair which results in less complications [2].

1. Case report

We present a newborn 40 + 2 weeks gestational age with antenatal diagnosis of a giant abdominal paraduodenal cyst. In a second trimester ultrasound, a 42 × 39 mm central cyst, below the stomach was found. In a third trimester follow-up ultrasound, the cyst was 68.2 × 67 × 70.2 mm in diameter, displacing the bladder, kidneys, liver and gallbladder. The diagnostic options could be a mesenteric cyst, enteric duplication, small bowel atresia, ovarian cyst or a choledochal cyst. No other prenatal malformations were observed, the pregnancy continued uneventfully. At birth, the patient had a palpable right abdominal mass. Postnatal abdominal ultrasound showed a cystic lesion with diffuse echoes inside. A magnetic resonance cholangiography showed a 78.1 × 70.61 × 79.85 mm diameter cyst, which was in contact with the medial and inferior surface of the liver; dilatation of the central intrahepatic bile duct due to compression and the cyst seemed to continue with the gallbladder. These findings are consistent with a choledochal cyst (Fig. 1). The patient remained asymptomatic, with mild jaundice. Laboratory studies revealed a total bilirubin (TB) 4.79 mg/dl, direct bilirubin (BD) 0.6 mg/dl, alkaline phosphatase 227 U/L, GGT 122 U/L and GPT 12 U/L. The patient was operated on the 11th day of life through a laparotomy, confirming a choledochal cyst of 8 cm in diameter with thick walls adhered to the duodenum, pancreas and right paracolic gutter with the gallbladder draining into the cyst and distal common bile duct atresia (Fig. 2). The gallbladder was removed, and 150 cc of bilious content aspirated, cyst excision and Roux-Y hepaticojejunostomy was performed. The postoperative course was favorable, initiating feeds on the fifth postoperative day. He was discharged on the tenth postoperative day, with normal ultrasound follow-up and normalization of bilirubin levels (BT 0.33 mg/dL and BD 0.09 mg/dL).

2. Discussion

The choledochal cyst is a congenital cystic dilatation of the bile ducts which can be intra- or extrahepatic or both. Choledochal cysts are usually diagnosed in childhood, although in utero and adult diagnosis is also common [1]. It is a benign disease, four times more frequent in females than in males [1]. The etiology is unknown, but
in 30–70% of cases, there is an abnormal binding of bile and pancreatic ducts, allowing pancreatic reflux to the biliary tree and exposing the biliary epithelium to pancreatic enzymes, which could contribute to the cyst formation [1]. Other etiological hypothesis include, weakness of the wall of the bile duct, increased intrabiliary pressure, sphincter of Oddi dysfunction and distal common bile duct obstruction. This last one is the most likely etiology in our case, since there was an absence of cyst communication with the duodenum, leading to accumulation of bile during pregnancy and a progressive increase in cyst size up to the enormous size described.

It has been suggested that age-related differences in presentation is determined by whether there is a reflux of activated pancreatic juice [3]. It may present itself with the classic triad of abdominal pain, jaundice and right upper quadrant mass and this triad is most commonly seen in pediatric patients. However, it is usually asymptomatic or it may be accompanied by complications such as cholangitis, pancreatitis, choledocholithiasis and malignant transformation. There are distinct differences to the pattern of presentation in adults and children. Specifically, adults are more likely to present with biliary or pancreatic symptoms and abdominal pain and are more likely to have symptomatic gallstones or acute cholecystitis. Children are more likely to present with an abdominal mass and jaundice. Cyst rupture is rare and typically is seen only in neonates and infants [4,5]. In neonates, biliary fibrosis and cirrhosis can occur due to biliary tree obstruction [6].

Prenatal diagnosis is increasingly common because of improved ultrasound equipment and routine prenatal ultrasound screening. It is essential since it leads to early surgery with less risk of postnatal complications such as cholestasis, cholangitis, perforation, biliary cirrhosis, pancreatitis, portal hypertension, and liver failure [2]. Some studies reported that delayed surgery results in liver fibrosis [7]. However, the diagnosis is sometimes difficult, and it is usually made during the third trimester of pregnancy, especially in cases of large cysts, as in our case. In some instances, the biliary tract cannot be differentiated from the cyst due to proximity to other structures. Ultrasounds performed at an early stage of pregnancy can confuse a cyst on right upper quadrant with the gallbladder or the umbilical vein and differential diagnosis must be made with duodenal atresia, ovarian cyst, hydronephrosis, liver cyst, enteric duplication cyst and giant meconium pseudocyst [2,8,9]. Also, it is difficult to distinguish biliary atresia from choledochal cyst in the perinatal period. Prenatal differential diagnosis of biliary atresia and choledochal cyst has been achieved on the basis of chronological changes in cyst size, cyst patterns, or timing of expression, but no definitive index exists [7]. The observation that patients with prenatally diagnosed choledochal cysts may, in fact, have biliary atresia warrants cautious interpretation of prenatal studies with careful family counseling. Postnatal diagnostic studies should be obtained immediately after birth, with a low threshold of suspicion for early exploration [10].

The prenatal management of choledochal cysts is conservative. However, Nasu et al. describe a prenatally diagnosed symptomatic choledochal cyst in which a percutaneous transhepatic drainage was performed during pregnancy, later undergoing surgical excision of the choledochal cyst, cholecystectomy and formation of a Roux-en-Y hepaticojejunostomy postpartum [11].

There are few cases described in the literature of large choledochal cyst, Rento et al. [12] described a 15 cm diameter cyst at birth, producing abdominal distension and respiratory compromise.

The concept of treatment of extrahepatic choledochal cysts has changed in the past 20 years because of a persistent high risk of malignancy after drainage procedures [3]. Today, treatment consists of complete excision of the extrahepatic cyst because of the risk of malignant transformation [1,3,13] and a bilioenteric reconstruction through a hepaticoduodenostomy or Roux-en-Y hepaticojejunostomy, our preferred technique [13]. Simple cyst excision can be performed depending on the anatomy of the cyst, as described by Todani, and type III cysts remain an exception to these guidelines because the risk of carcinoma is considered low, so, these patients are effectively treated by endoscopic sphincterotomy [3,13].

Management of prenatally diagnosed cystic lesion of the biliary tree is controversial. Conventionally, surgery is postponed until 3–6 months of age in asymptomatic patients because of the technical difficulties of the surgery and the risk of anesthesia during the neonatal period [1,8,14]. In symptomatic patients with large cysts an early surgery is indicated to confirm the diagnosis and avoid complications such as gallstones, cholangitis, perforation, cholestasis, vomiting by compression of the stomach or intestine, and even respiratory compromise [14]. However, some authors

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**Fig. 1.** Magnetic resonance cholangiography showing a 78.1 × 70.61 × 79.85 mm diameter cyst consistent with a choledochal cyst.

**Fig. 2.** An 8 cm choledochal cyst with the gallbladder draining into the cyst.
recommend excision of choledochal cyst and restoration of bile flow before 2 weeks of life, even in asymptomatic patients [6,15–17], in order to avoid the complications previously described and prevent progressive liver damage leading to liver fibrosis and biliary cirrhosis [6].

3. Conclusion

Prenatal diagnosis of choledochal cysts is important because it can determine the evolution, decreasing the risk of complications if an early surgical repair is done. Large choledochal cysts have a higher risk of complications such as perforation and compression of other organs. So, an early treatment is indicated, regardless if the patient is asymptomatic.

References