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Two-Dimensional Ultrasonographic Prenatal Diagnosis of Choledochal Cyst: Our Experience and Literature Review

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Abstract

Choledochal cyst is an abnormality of the biliary ducts, characterized by a dilatation of a portion of the biliary ductal system (most commonly a portion or the whole of the extrahepatic bile duct system) that presents usually at ultrasonography as a cystic mass in the right upper quadrant abdomen. Ultrasonographic prenatal diagnosis may be challenging since the detection of any intra-abdominal cyst raises the issue of differential diagnosis considering its nature as well as the organ of origin. In this paper, we report our experience with a case of a choledochal cyst strongly suspected during routine mid-trimester ultrasound scan, when a round liquid mass was found in the right upper quadrant of the abdomen; the cyst was independent of the other intra-abdominal organs but a connection was observed between the latter cyst and dilated bile ducts. The diagnosis was confirmed at birth and the infant underwent successful surgical correction of the anomaly. We review the current literature as well, discussing the current open challenges of diagnosing and managing these cases.

Keywords: Ultrasonographic; Prenatal diagnosis; Choledochal cyst; Trimester

Introduction

Choledochal cysts (CC) are defined as a “pseudo-cystic dilatation” of the extra- or intrahepatic biliary tree or both [1]. The Todani classification lists 5 types of CC: type 1, a pseudo-cystic dilatation of the extrahepatic bile duct (type 1A: fusiform, sac-like dilatation of the common bile duct; type 1-B: a diverticular dilatation of the common bile duct); type 2, a true cyst of the bile duct (lateral); type 3, a dilatation of the distal, intramural portion of the common bile duct (choledochocele); type 4, dilatation of both the extrahepatic and intrahepatic bile ducts; type 5, saccular dilatation(s) of the intrahepatic bile ducts. The overall incidence is very low with approximately one-third of reported cases been reported in

Japanese population [2]. CC type 1 is the most commonly clinical presentation (70-90%), and results in most cases from a malformation of the common junction between the bile duct and the pancreatic duct (at the level of the duodenal wall where they join to form a (usually very short) common channel) [1].

Because of the advance and widespread use of ultrasonography (US), CC is nowadays diagnosed frequently in infancy or early childhood before symptoms appear, and even can be recognized in the antenatal period with maternal US. Prenatal detection of CC is crucial since the newborn can be scheduled to a pre-emptive surgical treatment, and avoiding the occurrence of complications [3]. Nevertheless, diagnosis of CC at prenatal ultrasound is still challenging.

Our Experience

A 32-yr-old, gravida 2, para 1 woman was referred to our obstetric ultrasound facility at 22 weeks of gestation due to a suspected fetal anechoic mass at the level of the abdomen. The previous course of pregnancy had been uneventful. Maternal medical and family histories were unremarkable. US examination was performed through standard Aloka (Aloka Co., Ltd, Tokyo, Japan) equipped with a curved linear array trans-abdominal transducer (2-5 MHz) as well as with a trans-vaginal 4-8 MHz probe.

Detailed US imaging revealed a single fetus, of a size corresponding to 22 weeks of gestation, which was consistent with her estimated date of delivery. The only abnormality seen on ultrasound scan was a 15 × 16 mm anechoic cyst in the right upper quadrant abdomen (**Figure 1**).The mass was negative to Color-Doppler and was completely separate from the stomach, bowel, and gall bladder. Kidneys and adrenal glands were normal. A continuity between the gallbladder, the cyst and the dilated lower portion of the choledocus was clearly demonstrated (**Figure 1**).The latter observations confirmed with no doubt the diagnosis of a CC type I.

Serial US examinations every 2 weeks showed only a slight increase through gestation, measuring 16 × 19 cm at 35 weeks of gestation; at no moment was noticed a dilatation of the intrahepatic biliary system. At 38 weeks of gestation, a 3450 g

female infant was delivered vaginally after spontaneous labour, with an Apgar score of 8 and 10 at 1 and 5 minutes, respectively.

Postnatal US examination confirmed the diagnosis of type I CC: the diameter of the cyst was unchanged compared to the antenatal scan, and the intrahepatic bile ducts were not dilated; also the levels of direct bilirubin, alkaline phosphatase and gamma-glutamyl-transferase were normal. For these reasons, it was proposed to observe and postpone surgery.

Serial postnatal US were performed. At 5,5 months of age, the size of the cyst was 37 × 29 mm, with no sign of any complication. At 10,5 months of age and 8.300 Kgs of weight, surgery was electively performed (**Figure 1**).

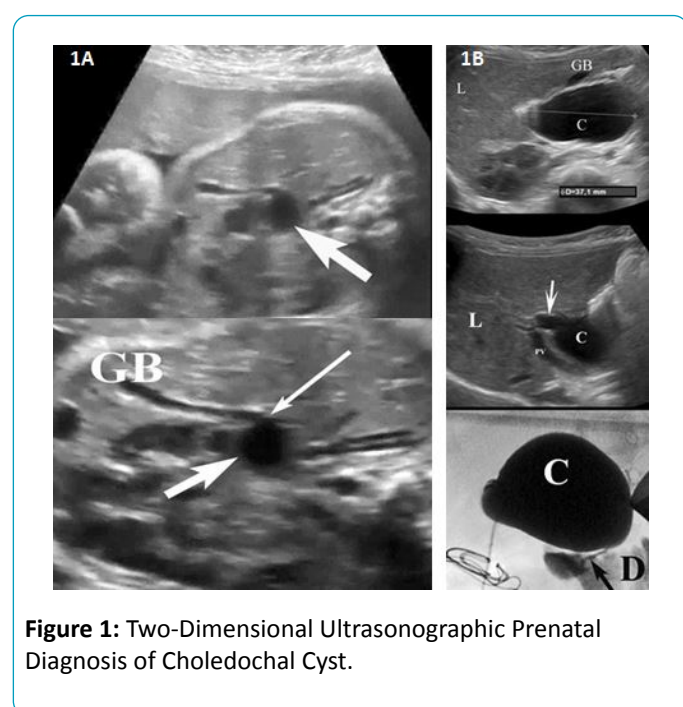


Figure 1: Two-Dimensional Ultrasonographic Prenatal Diagnosis of Choledochal Cyst.

The extrahepatic biliary system (gallbladder, cyst and choledocus) was completely resected and a Roux-en-Y hepaticojejunostomy was performed successfully. The histopathological examination confirmed the diagnosis of CC.

Discussion

The incidental US finding of an intra-abdominal cystic lesion opens the question of its nature, and the necessity of running a differential diagnosis, with regard to both the precise nature of the cyst and the organ of origin. Indeed, abdominal cystic anomalies originating from mesentery, omentum, pancreas, bile ducts, liver and ovary, and even associated to intestinal pathologies, have been reported in the literature.

Prenatal diagnosis of CC at US is rare: although this anomaly can present in any age group, most of them are diagnosed in infants and children and rarely in the fetus. In the present paper, a type I CC was diagnosed at 22-week prenatal US as based on the findings of an abdominal mass, negative to Color-Doppler, completely separated from the other intra-abdominal organs and in continuation with the gallbladder and the dilated choledochal duct. The latter observation (of a communication between the various structures) is original in that it never has been described previously in literature to our knowledge; previous reports of prenatal US diagnoses mentioned a simple, cystic mass in the upper abdomen or right upper quadrant, separated by the others intra-abdominal organs [3-7]. The US visualization of this communication between structures may be a simple, but valuable sign for addressing the correct diagnosis.

The average gestational age at time of diagnosis is 26.9 weeks, with the earliest diagnosis reported at 15 weeks (range 15 to 37 weeks) (**Table 1**) [4-9].

Table 1: The earliest diagnosis reported at 15 weeks

	Ultrasonographic Prenatal Diagnosis	GA (wk) at Diagnosis	GA (wk) at Delivery	Age (wk) at Surgery	Size (mm) at Diagnosis
Benhidjeb [3]	Choledochal cyst	29	40	16	NA
Lugo-Vicente [4]	Choledochal cyst	37	40	16	30
Redkar [5]	Choledochal cyst/ Intraabdominal cyst/Duodenal atresia (8 cases)	16-26	NA	Jan-28	13-35
Schroeder [6]	Choledochal cyst	15	42	5	20
Tongprasert [7]	Choledochal cyst	29	36	8	19
Hernández Herrea [8]	Intra-abdominal cyst	N.A.	39	8	50
Cong [9]	Intra-abdominal cyst/Biliary cystic malformation (16 cases)	19-38	22-NA	04-Dec	NA
Our case	Choledochal cyst	22	38	32	37

In a series of Redkar [6] the median gestational age at antenatal detection was 20 weeks. Benhidjeb [4] suggest that

it is possible to make a presumptive prenatal diagnosis of CC as early as 15 weeks.

The differential diagnosis includes duodenal atresia, biliary atresia, enteric duplication cyst, liver cyst, pancreatic cysts, gallbladder duplication, as well as ovarian cyst in female fetuses.

Although magnetic resonance imaging (MRI) or three-dimensional (3D) US with reconstructed planes may aid the prenatal diagnosis, we believe that a careful and precise two-dimensional (2D) US should be encouraged, since it may be satisfactory enough, in most cases, to demonstrate the sign of the connection between the gall bladder and the cyst, like in our case [10-15]. Indeed, MRI as well as 3D technology, although can be very useful in many cases, especially those of unfavourable fetal position, are expensive tools, require expert operators and are not readily available in some clinical contexts.

Biliary atresia presenting prenatally with a sub-hepatic cyst, although very rare, merits a special attention as this would trigger early surgery on the contrary of the strategy proposed with most other diagnosis: the US changes of the liver parenchyma, and the rising direct bilirubin in presence of acholic stool rapidly after at birth help for a rapid positive diagnosis of biliary atresia.

CC, when undiagnosed, would typically grow in size progressively with age until clinical symptoms or complications develop: the classic triad consist of jaundice (69%), abdominal mass and abdominal pain (caused by complications as cholangitis, bile obstruction or pancreatitis). In rare cases with a much prolonged course, chronic biliary obstruction is associated with prolonged icterus; evolution towards a secondary biliary cirrhosis and portal hypertension has been reported in rare cases. On the contrary, an incidental diagnosis done early in infancy (or a prenatal diagnosis as in our case) before clinical symptoms appear, is much likely to be associated with the absence of any complication and in particular biliary obstruction or pancreatitis [16,17]. As in our case, asymptomatic anicteric infants should be scheduled for elective surgery even in absence of complications, as evolution towards complication is the rule. In neonates with no complications and in absence of signs of hepatic suffering (dilatation of the bile ducts at US, elevation of Gammaglutamyl-transferase, slight elevation of direct bilirubin) the best strategy may consists in waiting a few months and postponing surgery to when the child is 8-10 Kgs of weight: this strategy allow to do surgery in best conditions, from an anatomical, technical and anesthesiological point of view.

In cases presenting complications, surgery should be preceded by adequate management of the complication (drainage of biliary system, endoscopic shincterotomy, medical treatment of pancreatitis) [5,17].

The recommended treatment of CC is total resection of the cyst and of the extrahepatic bile ducts; resection of the cyst is important as malignant transformation of the biliary epithelium of cysts left in place has been described in young adults operated during childhood [17]. The biliary drainage is then restored by a Roux-en-Y hepaticojejunostomy reconstruction; long-term outcome of such operations is good,

except for few patients presenting later in age with biliary obstruction and/or lithiasis caused by anastomotic stricture, which respond well to conventional management (radiological or surgical or both) [18]. For that reason, these patients needs a long-term follow-up consisting of US Doppler and liver tests at regular interval (annually or bi-annually).

Conclusion

Prenatal ultrasound of choledochal cyst by 2D-US is challenging but it is important for the proper prenatal-postnatal management of the case. Currently, the improved and shared experience of operators, with an increasing number of cases reported in the literature, may be helpful for addressing such an issue.

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