Case Report

Spontaneous Resolution of Choledochal Cyst

Yonatan Kurland, Ranjit I. Kylat, Sarah Desoky[1], Mohammad Y. Bader

Departments of Pediatrics and [1]Radiology, School of Medicine, University of Arizona, Tucson, AZ 85724, USA

Abstract

Choledochal cysts (CCs) are rare, congenital cystic dilations of the hepatobiliary tree that require surgical resection to avoid complications such as increased risk of malignancy. A 25-week gestational age male infant developed acholic stools, elevated alkaline phosphatase, and ultrasound findings consistent with CC Todani Type IVA. Surgery was deferred due to the patient's low weight. The patient's symptoms and radiographic findings subsequently resolved spontaneously. CCs have not been previously reported in extremely preterm infants. There are rare reported cases of spontaneously resolving hepatic cysts, all containing key differences from our patient. In patients in whom immediate surgery is not feasible, conservative management with close follow-up and serial ultrasound examinations would appear to be a reasonable course of action.

Keywords: *Acholic stools, Choledochal cyst, cystic dilatation, fusiform dilatation, preterm infant*

Introduction

Choledochal cysts (CCs) are rare, congenital cystic dilations of the hepatobiliary tree with an incidence of 1 in every 100,000–150,000 live births in Western countries; they are an order of magnitude more common in East Asian countries.[1] Untreated CCs can result in recurrent obstructive sequelae such as cholangitis, pancreatitis, hepatitis, cirrhosis, portal hypertension, and a significantly increased risk of malignancy.[2] Currently accepted treatment includes prompt surgical resection as they are not commonly known to resolve spontaneously.[3] We report a case of a CC in a 25-week gestational age premature infant with subsequent spontaneous resolution. CCs have not been previously reported in extremely preterm infants, and spontaneous resolution has only been documented in rare cases.[4,5]

Case Report

An 860 g male neonate was born at 25 weeks gestation to a healthy 20-year-old G1P0 mother. She had an emergent C-section at 25-weeks gestation for placental abruption following an uneventful early pregnancy. APGAR scores were 1, 3, and 5 at 1, 5, and 10 min, respectively. The infant required resuscitation followed by 14 days of mechanical ventilation as well as total parenteral nutrition (TPN) for 10 days, ending on day of life (DOL) 16. Head ultrasound screening was negative for intraventricular hemorrhage.

On DOL 26, the infant developed two isolated instances of acholic stool. Abdominal examination of all four quadrants and the costovertebral angles did not reveal distension, masses, or organomegaly. The infant had no recent history of feeding intolerance or abnormal stool patterns. Electrolytes, liver function tests, and bilirubin were within normal limits, with the exception of an elevated alkaline phosphatase (normal range: 150–420 IU/L) [Table 1]. There were no signs of bacterial infection.

Abdominal ultrasound with a high frequency linear probe showed focal regions of cystic dilation up to 6 mm of the intrahepatic ducts [Figure 1], with fusiform dilation of the proximal common bile duct (CBD) up to 3 mm [Figure 2], consistent with Todani Type IVA.[6] Early surgical intervention was not feasible due to the patient's low weight, hence the patient was followed up clinically. Acholic stools did not recur. Liver ultrasound

Address for correspondence: Dr. Ranjit I. Kylat, The University of Arizona, 1501, N Campbell Avenue, Tucson, AZ 85724, USA.
E-mail: rkylat@peds.arizona.edu

Access this article online

Quick Response Code:

Website: www.jcnonweb.com

DOI: 10.4103/jcn.JCN_108_16

How to cite this article: Kurland Y, Kylat RI, Desoky S, Bader MY. Spontaneous resolution of choledochal cyst. J Clin Neonatol 2017;6:112-5.

Table 1: Laboratory studies showing elevated liver function tests with elevated alkaline phosphatase

	27 days old	30 days old	3 months old
Sodium (mMol/L)	136	138	139
Potassium (mMol/L)	3.7	3.0	4.0
Chloride (mMol/L)	110	109	107
Carbon dioxide (mMol/L)	17	21	23
Blood urea nitrogen (mg/dL)	6	7	17
Creatinine (mg/dL)	0.5	0.5	0.4
Glucose (mg/dL)	55	73	128
Calcium (mg/dL)	9.0	9.2	9.1
Albumin (g/dL)	2.3	2.6	2.9
Protein (g/dL)	4.6	4.4	4.5
AST (IU/L)	28	19	22
ALT (IU/L)	11	8	12
Alkaline phosphatase (IU/L)	526*	487*	208
Bilirubin (mg/dL)			
Total	1.3	0.6	0.6
Direct		0.4	

AST – Aspartate transaminase; ALT – Alanine transaminase. *1-30 days 48-406 IU/L

was repeated 1 week later and was read by the same radiologist who reported that the previously identified cystic structure was not well seen and was filled with echogenic material. Two weeks later, the areas of cystic dilatation had fully resolved and the liver appeared normal [Figure 3].

Discussion

CCs are rare congenital dilatations of the biliary tract that commonly present in childhood.[7] They are as common as 1 in 13,000 live births in Japan but significantly rarer in Western countries with an incidence of 1 in 100,000–150,000 live births.[1] They are 4 times more common in females.[1] The etiology is not fully understood, but it has been hypothesized that anomalous pancreaticobiliary duct union causes regurgitation of pancreatic secretions into the biliary tree with subsequent weakening and dilatation of the duct.[1]

The presenting symptom varies based on the age of the patient; the "classic" triad of abdominal pain, abdominal mass, and jaundice is not very common.[7] Children will most often present with jaundice and adults with abdominal pain.[7] Asymptomatic antenatal diagnosis has been documented as early as 20-week gestational age.[8,9] Complications at the time of presentation include cholangitis, pancreatitis, portal hypertension, and elevation of serum liver enzymes.[2] Long-term complications include biliary cancer in as many as 20%–30% of patients.[2]

CCs are commonly classified based on the location of cystic abnormalities using the Todani criteria.[6] The

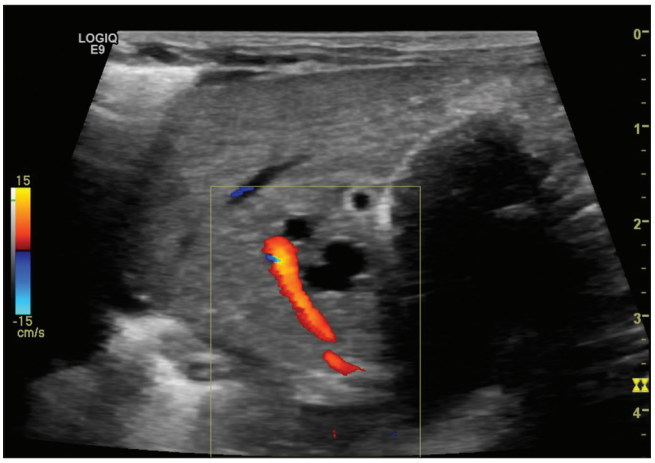

Figure 1: Abdominal ultrasound on day of life 28 demonstrating 6 mm cystic dilation of the intrahepatic ducts

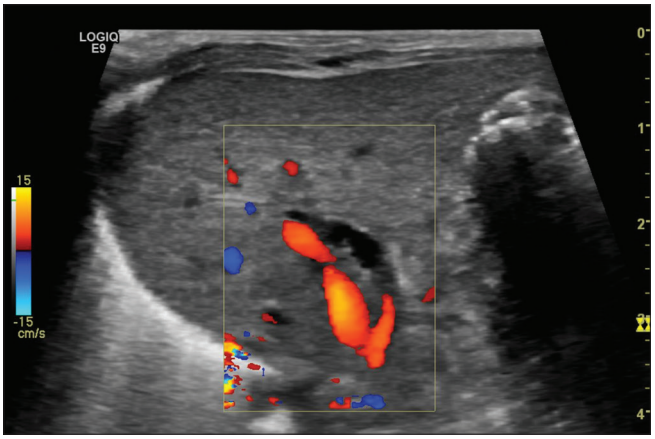

Figure 2: Abdominal ultrasound on day of life 28 demonstrating 3 mm fusiform dilation of the proximal common bile duct

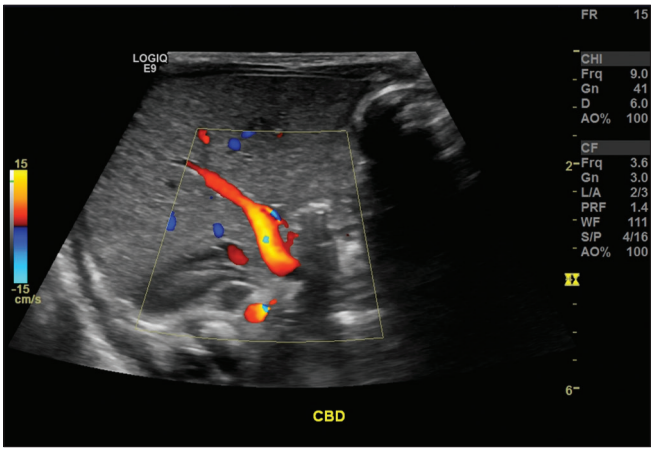

Figure 3: Abdominal ultrasound on day of life 54 showing normal liver, gall bladder, and common bile duct, without identified abnormalities

size of the CBD in full-term neonates and infants has been positively correlated with the age of the patient and has been described alternatively as 1.15 ± 0.53 mm in children of 0–12 months[10] and 0.69 ± 0.48 mm in infants of 0–3 months.[11] The sensitivity of

ultrasound to diagnose CC has only been determined in children >1 year of age and found to be 71% in dilatations >7 mm.[12] There is no data on the size of the CBD or the sensitivity of ultrasound in premature or extremely premature infants. Our patient had 6 mm of cystic dilatation of the intrahepatic ducts which is markedly greater than the normal ranges, particularly in light of his gestational age and weight (25 weeks and 860 g).

The preferred treatment is surgical resection due to the high risk of biliary cancer if left untreated.[13] The specific surgical procedure varies based on Todani classification and the extent of disease.[3] Our patient was classified as Todani Type IVA with dilatation extending from the CBD to the intrahepatic biliary tree. Surgical intervention was considered, but was determined to be infeasible due to the infant's low birthweight. Follow-up ultrasounds showed gradual resolution of the CC.

A recent randomized control trial by Diao *et al*. supports early surgical intervention even in asymptomatic infants.[14] They demonstrate a significantly increased incidence of high-grade hepatic fibrosis when surgical intervention was delayed beyond 1 month of life.

Spontaneous resolution of CCs has been very infrequently reported in the literature. Ruiz-Elizalde and Cowles[4] described a full-term 11-day-old boy whose only presenting symptom was emesis; an ultrasound showed a fusiform dilatation of the CBD to 1.4 cm. Symptoms resolved and repeat ultrasounds showed interval decreased size and subsequent resolution. In a similar case reported by Evans and Vance,[5] a 3-month-old full-term baby presented with acholic stools, conjugated hyperbilirubinemia, elevated liver enzymes, and dilatation of the entire biliary system. The authors concluded that CC was an unlikely diagnosis due to the fact that the lesion resolves spontaneously, however no alternative diagnosis was identified.

Spontaneous resolutions of noncongenital hepatic cysts have been reported in the adult literature. A 62-year-old patient with hepatitis C was found to have a 40 mm cystic lesion along the middle hepatic vein with subsequent complete resolution.[15] In another case, a 45 mm simple liver cyst transformed spontaneously into an inflammatory pseudo-tumor; the authors proposed that the lesion would have resolved spontaneously even without surgery and the pseudo-tumor may represent an intermediate stage before resolution.[16]

There are several explanations for the spontaneous resolution of our patient's CC; however, none of them are overly compelling. In a retrospective study by Lin *et al*., spontaneous resolution was seen in the majority of cases with fusiform dilatations, but in none with cystic dilatations.[17] It is possible that because our patient had both fusiform and cystic dilatations, the unknown mechanisms leading to spontaneous resolution of fusiform type dilatations played a role in resolving our patient's cystic component as well. Another possibility is that CCs can potentially resolve spontaneously during the later stages of gestation which our patient did not experience due to his birth at 25 weeks. This hypothesis is unlikely given that there are several reports of CC diagnosed antenatally prior to 25 weeks' gestation and no reports of spontaneous resolution, however the knowledge base is limited.[8,18] Finally, there have been reports of spontaneous resolution of noncystic CBD dilatations related to TPN-induced biliary sludge, however this finding was not present in our patient.[19]

Conclusion

Our patient is unique due to his 25-week gestational age as well as resolution of laboratory abnormalities, acholic stools, and sonographic findings. He was initially not a suitable surgical candidate due to his low weight at birth, but the subsequent clinical course ultimately rendered surgery unnecessary. The utility of early surgery has been well documented in literature and so it would be inappropriate to make any broad treatment recommendation based on this single case. It is interesting to observe that this is the second patient described in literature whose CC spontaneously resolved when immediate surgery was not clinically indicated. In patients in whom immediate surgery is not feasible and in preterm infants with CC, conservative management with close follow-up and serial ultrasound examinations would appear to be a reasonable course of action.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Soares KC, Arnaoutakis DJ, Kamel I, Rastegar N, Anders R, Maithel S, *et al.* Choledochal cysts: Presentation, clinical differentiation, and management. J Am Coll Surg 2014;219:1167-80.
2. Todani T, Toki A. Cancer arising in choledochal cyst and management. Nihon Geka Gakkai Zasshi 1996;97:594-8.
3. Ronnekleiv-Kelly SM, Soares KC, Ejaz A, Pawlik TM. Management of choledochal cysts. Curr Opin Gastroenterol 2016;32:225-31.
4. Ruiz-Elizalde AR, Cowles RA. A practical algorithm for accurate diagnosis and treatment of perinatally identified biliary ductal dilation: Three cases that underscore the importance of an individualised approach. J Matern Fetal Neonatal Med 2009;22:622-8.

5. Evans JE, Vance JC. Spontaneous resolution of extrahepatic biliary obstruction in a neonate. Aust Paediatr J 1987;23:301-3.
6. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
7. de Vries JS, de Vries S, Aronson DC, Bosman DK, Rauws EA, Bosma A, *et al.* Choledochal cysts: Age of presentation, symptoms, and late complications related to Todani's classification. J Pediatr Surg 2002;37:1568-73.
8. Tanaka H, Sasaki H, Wada M, Sato T, Kazama T, Nishi K, *et al.* Postnatal management of prenatally diagnosed biliary cystic malformation. J Pediatr Surg 2015;50:507-10.
9. Akhan O, Demirkazik FB, Ozmen MN, Ariyürek M. Choledochal cysts: Ultrasonographic findings and correlation with other imaging modalities. Abdom Imaging 1994;19:243-7.
10. Zhang Y, Wang XL, Li SX, Bai YZ, Ren WD, Xie LM, *et al.* Ultrasonographic dimensions of the common bile duct in Chinese children: Results of 343 cases. J Pediatr Surg 2013;48:1892-6.
11. Hernanz-Schulman M, Ambrosino MM, Freeman PC, Quinn CB. Common bile duct in children: Sonographic dimensions. Radiology 1995;195:193-5.
12. Lee HC, Yeung CY, Chang PY, Sheu JC, Wang NL. Dilatation of the biliary tree in children: Sonographic diagnosis and its clinical significance. J Ultrasound Med 2000;19:177-82.
13. Tanaka N, Ueno T, Takama Y, Fukuzawa M. Diagnosis and management of biliary cystic malformations in neonates. J Pediatr Surg 2010;45:2119-23.
14. Diao M, Li L, Cheng W. Timing of surgery for prenatally diagnosed asymptomatic choledochal cysts: A prospective randomized study. J Pediatr Surg 2012;47:506-12.
15. Yoshida H, Onda M, Tajiri T, Mamada Y, Taniai N, Uchida E, *et al.* Spontaneous disappearance of a hepatic cyst. J Nippon Med Sch 2001;68:58-60.
16. Blanc JF, Balabaud C, Saric J, Bioulac-Sage P. Disappearance of a biliary cyst. J Hepatol 2005;42:430.
17. Lin SF, Lee HC, Yeung CY, Jiang CB, Chan WT. Common bile duct dilatations in asymptomatic neonates: Incidence and prognosis. Gastroenterol Res Pract 2014;2014:392562.
18. Weng R, Hu W, Cai S, Guo X, Luo Q. Prenatal diagnosis and prognosis assessment of congenital choledochal cyst in 21 cases. J Obstet Gynaecol 2016;36:324-7.
19. Holgersen LO, Stolar C, Berdon WE, Hilfer C, Levy JS. Therapeutic and diagnostic implications of acquired choledochal obstruction in infancy: Spontaneous resolution in three infants. J Pediatr Surg 1990;25:1027-9.