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. Untreated CCs can result in recurrent obstructive sequelae such as cholangitis, pancreatitis, hepatitis, cirrhosis, portal hypertension, and a significantly increased risk of malignancy. Currently accepted treatment includes prompt surgical resection as they are not commonly known to resolve spontaneously. 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.

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. 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...
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.[3]

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

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Conflicts of interest
There are no conflicts of interest.

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