with the advance and widespread use of maternal ultrasonogra-
phy, congenital biliary dilatation is diagnosed more frequently
during the antenatal period.\(^1\,^3\) Detection of focal cystic dilata-
tion of the extrahepatic biliary tree on prenatal ultrasonography may be ei-
ther a choledochal cyst or cystic biliary atresia.\(^1\,^3\) The differential diagnosis
doctor biliary lesion is important to determine the appropriate time of
surgery. Definitive surgery for a choledochal cyst may be done as late as 6
months to reduce perioperative or postoperative complications; however, in
biliary atresia, this time must be less than 2 months, thus, diagnostic stud-
ies should be done as soon as possible after birth.\(^2\,^5\)

Postnatal Management of Prenatally
Diagnosed Biliary Cystic Lesions:
A Case with Cystic Biliary Atresia

Postnatal Tedavisi: Kistik Bilier Atrezili Bir Olgu

ABSTRACT We present the case of a newborn with a biliary cystic lesion found during a routine
antenatal ultrasonography. The postnatal ultrasonography and magnetic resonance cholangiogra-
phy were in favor of a choledochal cyst, while a hepatobiliary scintigraphy showed no drainage of
the tracer into the intestine. During surgery, it was found that the infant have cystic biliary atre-
sia. Accurate postnatal differentiation of prenatally detected biliary atresia with a cyst at porta from
a choledochal cyst is important but not always possible using less invasive radiologic methods such
as ultrasonography and magnetic resonance imaging. Therefore, to improve the surgical outcome,
early exploration should be performed when a clear discrimination between biliary atresia and
choledochal cyst is not possible.

Key Words: Biliary atresia; choledochal cyst; cholangiography

ÖZET Prenatal ultrasonografi takipleri esnasında tespit edilen kistik bilier lezyonlu bir yenidoğan
olgu sunuyoruz. Postnatal ultrasonografi ve manyetik rezonans kolanjiografi bulguları koledok kisti
tanımı desteklerken, hepatobiliar sintigrafide aktif maddenin intestinal sisteme geçmediği görüldü.
Ameliyat esnasında hastada kistik bilier atrezi tespit edildi. Prenatal dönemde tanınamayan kistik bilier
atrezinin koledok kistinden postnatal dönemde kesin olarak ayırmanın yapılmasi önemlidir. Ancak
bu ultrasonografi ve manyetik rezonans gibi daha az invaziv radyolojik görüntüleme yöntemleri ile
er her zaman mümkün olmamaktadır. Bu nedenle daha iyi cerrahi sonuçlar elde etmek için bilier
atrezi ile koledok kisti arasında kesin bir ayrımın yapılmasıldığı zaman erken eksplorasyon
yapılmalıdır.

Anahtar Kelimeler: Bilier atrezi; koledok kisti; kolanjiyografi

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Despite varied attempts to differentiate cystic biliary atresia from a prenatally detected choledochal cyst, no single test or imaging modality has been found to be reliable. Ultrasonography, hepatobiliary scintigraphy, and percutaneous or laparoscopic-guided cholangiography are most-frequently used diagnostic studies. Recently, magnetic resonance cholangiography was reported as being a reliable and less invasive procedure for multiplanar evaluation of the hepatobiliary system. Delayed diagnosis of a truly obstructed biliary system can result in serious deterioration of liver function that may be irreversible; therefore, early diagnosis and treatment of prenatally diagnosed biliary cystic lesions are critical.

**CASE REPORT**

The in-vitro fertilized fetus of a 34-year-old woman had a 20 × 25 mm subhepatic cystic lesion at 19 weeks’ gestation. The mass was 23.5 × 24 mm and 25 × 27 mm at 33 and 36 weeks respectively (Figure 1). Postnatal ultrasonography showed a 29 × 24 mm subhepatic cystic lesion. The gallbladder was seen coming off the cystic mass and the intrahepatic bile ducts were dilated. A total bilirubin level of 12.3 mg/dl with a conjugated fraction of 2.8 mg/dl was detected on the sixth day of life. The patient had cholic stools during the first weeks. Hepatobiliary scintigraphy, with phenobarbital enhancement, was done on the ninth day and showed normal hepatocyte function but no excretion into the intestines during the following 24 hours. A magnetic resonance cholangiography was subsequently done on the 12th day and showed mild dilatation of the intrahepatic bile ducts and a 2.5 × 3 cm cystic mass consistent with a type-1a choledochal cyst communicating with the gallbladder (Figure 2). In the light of these findings, the patient was followed with a diagnosis of choledochal cyst.

At 2 months of age, the patient’s liver enzymes and bilirubin levels increased (aspartate aminotransferase, 146 IU/L; alanine aminotransferase, 95 IU/L; total bilirubin, 8.69 mg/dl; conjugated bilirubin, 7.49 mg/dl), and she began to pass acholic stools. Upon these findings the patient underwent exploratory surgery. Exploration revealed a small gallbladder and a cholestatic liver. A 3 cm cystic mass related to the gallbladder was seen at the portal hilus. An intraoperative cholangiography showed a blind ending cyst that was communicated only with the gallbladder (Figure 3). Diagnosis of type-3d biliary atresia was made and a Kasai portoenterostomy was performed. The results of the liver biopsy showed cholestatic hepatitis and hypoplasia of the intrahepatic bile ducts. The baby had pigmented stool postoperatively and was discharged without problems 14 days after surgery. A
scintigraphy done at 1-month follow-up examination revealed normal hepatocyte function and normal passage to the intestines.

**DISCUSSION**

A subhepatic cyst detected during the second or third trimester raises the suspicion of biliary atresia or choledochal cyst.\(^1\)\(^-\)\(^3\)\(^,\)\(^9\)\(^,\)\(^10\) The incidence of extrahepatic cysts in biliary atresia has been reported to range from 5% to 20%.\(^4\)\(^,\)\(^10\)\(^,\)\(^11\) Casaccia et al. claimed that echogenicity, size, and increase in size were important prenatal ultrasound findings for a correct diagnosis of different types of biliary cystic lesions.\(^9\) Muller et al. have found significantly lower levels of amniotic fluid gamma-glutamyl transferase in 2 patients with biliary atresia.\(^12\) Okada et al., in a limited number of patients, showed that fibrosis or CD56-positive biliary duct cells in liver biopsy specimens are reliable for the differential diagnosis of a choledochal cyst and prenatally diagnosed type-I cystic biliary atresia.\(^13\)\(^,\)\(^14\) Nevertheless, an accurate prenatal diagnosis has been reported in only 15% of cases.\(^1\)\(^,\)\(^4\)\(^,\)\(^9\)

Ultrasonography is usually accepted as the first postnatal diagnostic study. Dilated intrahepatic bile ducts with a well-distended gallbladder have been reported as being detected in patients with a choledochal cyst.\(^4\)\(^,\)\(^11\) Kim et al. detected dilatation of the intrahepatic bile ducts in all patients with a choledochal cyst, while this finding was detected in only 5% of patients with cystic biliary atresia. These authors also observed an abnormally small gallbladder in 89% of their patients with cystic biliary atresia, however, the gallbladder was normal or distended in all patients with a choledochal cyst.\(^4\)

Hepatobiliary scintigraphy, with phenobarbital enhancement, has a high sensitivity, but low specificity for the diagnosis of biliary atresia.\(^6\)\(^,\)\(^7\) Percutaneous, surgical, or laparoscopic-guided cholangiography provides definite differentiation of biliary atresia from a choledochal cyst.\(^7\)\(^,\)\(^11\) However, these diagnostic tools are invasive interventions and require general anesthesia.\(^7\)

Recently, several studies have shown that magnetic resonance cholangiography is a less invasive technique that can completely visualize the biliary system.\(^5\)\(^,\)\(^7\)\(^,\)\(^8\) It is not limited by hepatic function or bilirubin levels.\(^8\) Han et al. and Jaw et al. report that visualization of the entire extrahepatic bile ducts excludes biliary atresia with 96% and 100% accuracy, respectively.\(^7\)\(^,\)\(^8\) A few articles reported that periportal hyposignal resolving after gadolinium injection or periportal thickening are signs of periportal fibrosis in biliary atresia on magnetic resonance cholangiography.\(^7\)\(^,\)\(^15\) However, these findings are also reported in patients with other causes of cholestatic jaundice.\(^7\)\(^,\)\(^15\) An atrophic gallbladder on magnetic resonance cholangiography indicates biliary atresia.\(^5\)\(^,\)\(^7\)\(^,\)\(^8\)

There are 34 cases with the diagnosis of antenatally detected cystic biliary atresia from 1980 to present in English literature. Four patients out of 34 with antenatally detected cyst were operated with the correct preoperative diagnosis of biliary atresia.\(^16\)\(^-\)\(^19\) The largest series with 12 patients was published by Caponcelli et al.\(^20\) They notified that the diagnoses of all patients except one were made at laparotomy and half of the patients were operated after 36 days of life. Saito et al reported seven cases with the diagnosis of biliary cystic malformation of which six were biliary atresia.
and all but one were operated after 60 days of life. Brunero et al. reported two patients with biliary atresia one of which was followed with the diagnosis of choledochal cyst and operated at 111 days. Hasegawa et al. found that postnatal ultrasound, computerized tomography or magnetic resonance imaging showed similar findings to the prenatal ultrasound and nuclear medicine scan showed no excretion from liver and they could not make differential diagnosis of cystic biliary atresia from choledochal cyst in three patients. De Matos et al. operated two patients with antenatally detected biliary cysts associated with no excretion to the intestine on hepatobiliary scintigraphy. They diagnosed biliary atresia through operative cholangiography. Mackenzie et al. reported three cases with antenatally detected biliary cysts. The diagnoses were cystic biliary atresia in two and choledochal cyst in 1 patient. Preoperative tests were consistent with choledochal cyst but biliary atresia was diagnosed through operative cholangiography. Okada et al. could not differentiate choledochal cyst from type-1 biliary atresia on the basis of magnetic resonance cholangiography in two cases with antenatally diagnosed biliary cyst. They confirmed the diagnosis with intraoperative cholangiography.

We present an additional infant with a large subhepatic cyst of unchanging size on prenatal ultrasonography. Choledochal cyst was the most likely diagnosis by ultrasonography and magnetic resonance cholangiography. Hepatobiliary scintigraphy did not show excretion into the intestines. Results of the imaging studies were not conclusive and caused some delay in doing the surgery. Differentiation between type-1a and type-3d cystic biliary atresia is based on the patency or obstruction of the common bile duct at the hilum. According to the intraoperative cholangiography, the presented case is classified as type 3d.

In conclusion, when a subhepatic cystic lesion is detected via fetal ultrasonography, cystic biliary atresia (either type 1a or type 3d) should be part of the differential diagnosis. However, prenatal and postnatal diagnoses remain difficult with respect to differentiating cystic biliary atresia from a choledochal cyst. Ultrasonography, hepatobiliary scintigraphy, and magnetic resonance cholangiography findings must be interpreted with each other and with clinical and laboratory findings to diagnose cystic biliary atresia. As a result early exploration should be performed in cases in which a clear discrimination between biliary atresia or choledochal cyst is not possible.

**REFERENCES**


