ELSEVIER

Clinical Imaging 31 (2007) 269–272

CLINICAL IMAGING

Radiology–Pathology Conference: choledochal cyst☆

Bobbi N. Wax[a], Nicole M. Durie[b], Poonam Khullar[b], Anita P. Price[a,c], Charles V. Coren[d], Douglas S. Katz[a,*]

[a]*Department of Radiology, Winthrop-University Hospital, 259 First Street, Mineola, NY 11501, USA*
[b]*Department of Pathology, Winthrop-University Hospital, 259 First Street, Mineola, NY 11501, USA*
[c]*Department of Radiology, Memorial Sloan-Kettering Cancer Center, 1275 York Avenue, New York, NY, 10021, USA*
[d]*Department of Surgery, Winthrop-University Hospital, 259 First Street, Mineola, NY 11501, USA*

Received 10 March 2007; accepted 24 March 2007

Abstract

We report the radiologic and pathologic findings of a newborn with a choledochal cyst and review the etiology, diagnostic considerations, and management of this entity.

Keywords: Choledochal cyst; Biliary tract; Congenital anomaly; Neonatal jaundice; HIDA; Ultrasound

1. Introduction

Included in the spectrum of congenital biliary tract anomalies are choledochal cysts. Although altogether rare, type I, which is cystic dilatation of the common bile duct as classified by Todani et al. [1,2], is the most common type, accounting for 80–90% of all choledochal cysts [2]. Choledochal cysts usually present in infancy and childhood and are most frequently seen in female patients. In newborns, these anomalies may be found incidentally. The classic triad of abdominal pain, obstructive jaundice, and fever seen with a choledochal cyst occurs in less than one third of all cases; at least one component of the triad is seen in patients who are more than 1 year of age [3–5]. In this Radiologic–Pathology Conference, we review a case of a choledochal cyst, which was discovered in an asymptomatic newborn girl, as well as the corresponding imaging and pathologic findings. The diagnostic and management considerations are also discussed.

2. Case presentation

A newborn girl at our institution was subjected to a prenatal ultrasound several weeks before delivery, which revealed a cystic lesion in the upper right abdomen. Since the pregnancy was otherwise uneventful, the abnormality was followed with serial sonographic examinations until birth. The child was born via normal, spontaneous vaginal delivery. The newborn physical examination was normal; the baby was in no acute distress, and she was anicteric, without any other evidence of jaundice. There was no palpable mass or organomegaly on abdominal examination. The prenatal imaging prompted blood work, which revealed an initially normal SGPT of 42 IU/l (normal range, 27–65 IU/l) and an elevated alkaline phosphatase level of 289 IU/l (normal range, 38–112 IU/l). SGOT was not obtained secondary to a laboratory error. The total bilirubin was borderline low, at 0.1 mg/dl (normal range, 0.2–1.1 mg/dl). The albumin and total protein were also within the normal range. Amylase and lipase levels were not obtained.

Abdominal sonography, which was performed shortly after delivery (Fig. 1A and B), revealed an anechoic structure with an imperceptible wall adjacent to the gallbladder and contiguous with the common bile duct. The lesion measured 1.5 cm in greatest diameter and 2.4 cm in length. No flow was demonstrated within the structure on color and power Doppler. The liver itself was normal and had a homogenous echotexture without biliary dilatation. No gallstones were

☆ From the monthly Radiology–Pathology Conferences at Winthrop-University Hospital.

* Corresponding author. Department of Radiology, Winthrop-University Hospital, 259 First Street, Mineola, NY 11501, USA. Tel.: +1 516 663 3800; fax: + 1 516 663 8172.
E-mail address: dkatz@winthrop.org (D.S. Katz).

0899-7071/07/$ – see front matter © 2007 Elsevier Inc. All rights reserved.
doi:10.1016/j.clinimag.2007.03.005

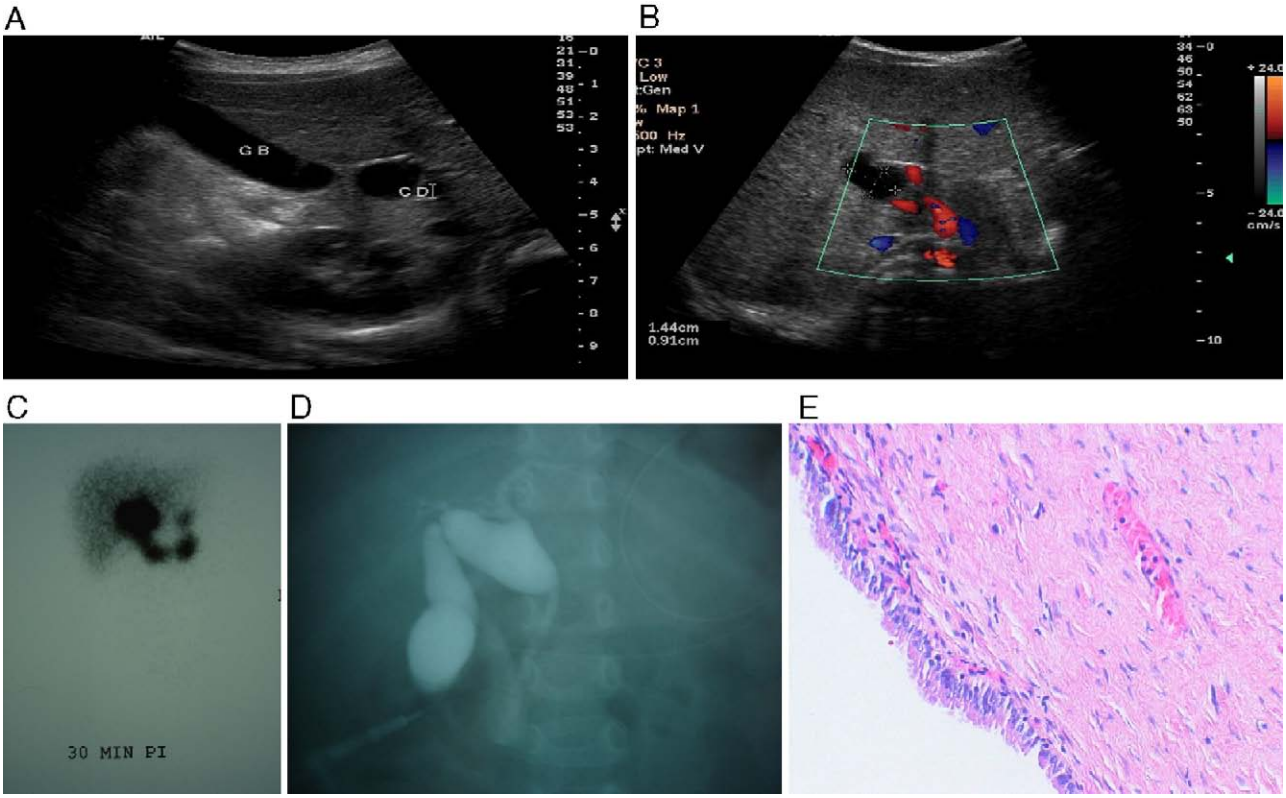

Fig. 1. A newborn girl with an abnormal prenatal ultrasound. (A) Sonographic image shows the gallbladder (GB) and the adjacent, markedly dilated common bile duct (CD) representing a type I choledochal cyst. (B) Color Doppler axial image shows absence of flow in the dilated cystic structure, further confirming that it represents the common bile duct. (C) Thirty-minute frontal image from a HIDA scan showing marked dilatation of the common bile duct without biliary obstruction. (D) Frontal image from an intraoperative cholangiogram. Marked cystic dilatation of the common bile duct is again demonstrated, with communication to the gallbladder (to the patient's right) via the cystic duct, confirming a type I choledochal cyst. (E) Representative microscopic image of the cyst wall, demonstrating a surface of columnar epithelium (H&E stain, ×200).

visualized, and there was no gallbladder wall thickening or pericholecystic fluid. A technetium-99m (Tc-99m) hepatobiliary iminodiacetic acid (HIDA) scan was also performed (Fig. 1C), which showed normal hepatic uptake and normal biliary-to-bowel transit time. There was an abnormal accumulation of radiotracer in the region of the porta hepatis, which correlated with the cystic dilatation of the common bile duct. The patient then underwent intraoperative cholangiography (Fig. 1D), which again demonstrated cystic dilatation of the common bile duct with communication to the gallbladder via the cystic duct, confirming a type I choledochal cyst. The patient had a resection of the choledochal cyst, a cholecystectomy, and a hepaticojejunostomy.

The surgical specimen submitted consisted of a 6.7×1.4 cm gallbladder with a 1.5×1 cm, collapsed, previously opened cyst attached near the cystic duct. The cyst was thin walled, with a smooth lining; its lumen was empty. No calculi were identified in the cyst or in the gallbladder. On microscopic evaluation, the cyst was lined by columnar epithelium with fibrous tissue. No atypia or inflammation was identified (Fig. 1E).

Follow-up laboratory studies showed a peak in SGPT, SGOT, alkaline phosphatase, amylase, and lipase levels on Postoperative Day 1, with a subsequent decline in the abnormal blood work. A follow-up ultrasound performed 2 months later revealed normal postoperative anatomy.

3. Discussion

Choledochal cysts are rare congenital anomalies that need recognition because they can lead to cholangitis, calculi formation, portal hypertension, and malignancy of the biliary tract [5,6]. Approximately two-thirds of choledochal cysts are discovered before the age of 10 years [1,3,4]. While choledochal cysts are frequently discovered in the neonatal period, they may alternatively be diagnosed later in life. Choledochal cysts have a predilection for females and occur most prevalently in patients of Asian descent [7]. The routine use of prenatal ultrasonography has led to more frequent prenatal diagnosis of choledochal cyst in recent years. Choledochal cysts may also be suspected in neonates and young infants who have a conjugated hyperbilirubinemia, which should be differentiated from biliary atresia as an alternative underlying diagnosis.

Ultrasound is the imaging modality of choice for the initial evaluation of neonatal jaundice. Common duct and gallbladder size and morphology play important roles in differenti-