Neonatal spontaneous bile duct perforation presenting as giant intraabdominal cyst

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A R T I C L E   I N F O
Article history:
Received 22 January 2013
Received in revised form 25 February 2013
Accepted 25 February 2013

Key words:
Neonatal
Common bile duct
Perforation
Pseudocyst
Abdominal cyst

A B S T R A C T
We present a case of neonatal spontaneous bile duct perforation in a previously healthy neonate presenting acutely at 2 weeks of age followed by abdominal distention, acholic stools and obstructive jaundice. Ultrasound and CT scan showed a giant intraabdominal cyst of unknown origin. Exploratory laparotomy revealed a tense lesser sac pseudocyst containing bile with compression and displacement of adjacent structures. The patient underwent partial excision and drainage of the cyst uneventfully with full recovery.

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Neonatal spontaneous biliary perforation (NSBP) is a rare but well documented potentially life-threatening cause of jaundice in infancy, requiring prompt diagnosis and surgical intervention. First described in 1932, fewer than 150 cases in neonates and infants have been reported to date [1–5]. We describe a very unusual presentation of NSBP in which neonate presented initially with acute presentation treated as presumed sepsis followed by development of giant lesser sac pseudocholecodochal cyst presenting as massive intraabdominal cyst of unknown origin.

1. Case report

A 16-day-old previously well and exclusively breast-fed Asian female neonate presented with persistent crying, anorexia, vomiting and lethargy. She was born by normal vaginal delivery at term with a birth weight of 2600 g following an uneventful antenatal period. Mother had group B streptococcus infection treated successfully.

At admission she was afebrile, well perfused and hydrated with normal vital signs. She underwent a full septic screen for presumed sepsis. However, all investigations including urine, blood, lumbar puncture and chest X-ray were normal. She was started on triple IV antibiotic therapy-gentamycin, amoxicillin and cefotaxime for 3 days till all single set of cultures-blood, urine and CSF were negative and baby was clinically well. She was discharged home.

At 1 month of age, her mother noted abdominal distension, diagnosed to have infantile colic and given anticoic medication. Owing to persistent distension, no response to medication and passing of pale stools; she was referred to pediatric assessment unit.

On second admission, although slightly pale she was not jaundiced. Marked abdominal distension dull on percussion was only abdominal sign. Laboratory investigations showed normal renal functions, a bilirubin of 40 mmol/L with a conjugated fraction of 29 mmol/L, C-reactive protein 92 mg/L and platelets 1,118 × 10⁹/L.

An ultrasound scan (USS) showed a large multiloculated multi-septated cyst with some internal debris extending down to the pelvis exerting mass effect on the intraabdominal structures. An abdominal computerized tomogram (CT) showed a huge cyst in the upper and left side of abdomen approximately 11 cm in transverse diameter, 13 cm in craniocaudal extent and 8 cm in anteroposterior diameter extending into the lesser sac mainly. The largest locule was in the lesser sac displacing the stomach and duodenum. There were many smaller locules in the region of porta hepatitis in very close contact with the biliary tract. The common bile duct was dilated in the region of head of pancreas, close to the stretched second part of duodenum. The bowel was displaced to the right and in lower abdomen. It was concluded to be a huge abdominal cyst.
Differentials included a duplication cyst, omental cyst or a lymphangioma (Fig. 1).

Advice was taken from the regional pediatric hepatobiliary center who felt that choledochal cyst was an unlikely pathology. A diagnostic exploratory laparotomy with therapeutic intervention was advised.

At operation, an 800 mL tense pseudocyst of the lesser sac was decompressed, which contained bile. Liver and all other visceræ appeared normal. The cyst was dissected from the mesocolon, the pancreas, splenic hilum, posterior gastric wall, superior surface of the liver and over the porta hepatitis and partially excised. Two drains were left in situ for drainage. There was dark green discoloration in the common bile duct at its junction with the cystic duct but there was no leak of bile actively and the perforation had sealed.

Histology showed that the cyst wall was not epithelialized and was likely to be a pseudocyst secondary to a spontaneous biliary perforation. Over the next few days, she recovered well from the operation and while the drains were draining between 30 and 50 mL; they were left in situ for 1 week. On serial USS the collections appeared to be resolving. The drains were removed and she was discharged home once she was feeding normally. The stools changed color to normal post-operatively.

At 6 weeks follow up, she was well and an ultrasound showed that the anterior collection had entirely resolved while the subphrenic collection was smaller than previous, now measuring only 3 × 1.9 cm.

At 3 months follow up her ultrasound was entirely normal. According to her growth chart, she was now on the 25th centile having started on the 9th centile. At 2 year follow up she is asymptomatic and thriving well.

2. Discussion

The etiology of NSBP is unknown. Proposed theories have included congenital mural weakness of the common bile duct, trauma, ischemia, distal biliary obstruction and pancreatic reflux with an anomalous junction between the common bile duct and the pancreatic duct. NSBP has been suggested to be a form of acquired biliary atresia [3]. In the spectrum of biliary atresia-choledochal malformation classification, five types of true choledochal cysts are recognized in the axial biliary system [6] and a suggestion has been made to add cholecystocele as a true choledochal cyst of the paraxial biliary system [7]. Our case and various other reported cases form a group of choledochal pseudocysts and the localized varieties actually can be confused with the true choledochal cysts [8].

Various possible presentations of NSBP include generalized, regional, localized and combined lesions. Commonest presentation is biliary ascites; if it gets secondarily infected it gets converted into biliary peritonitis which is potentially lethal. The regional compartmentalization can be either in the greater or the lesser sac presenting as giant cystic lesion and it may not be possible to accurately identify them as biliary pseudocysts as they have secondary daughter localized variety of periporal or ligamentous pseudocholedochal cysts. Our current case adds to the regional mode of presentation in the form of giant false or pseudocholedochal cysts with small secondary periporal and ligamentous extensions. The localized periporal and ligamentous collections alone may get confused with choledochal cyst themselves [8].

Most often, NSBP is seen in infants aged 2–20 weeks, but it has been reported in neonates aged as young as 3 days and in children as old as 50 months [9]. The classic presentation is that of a previously healthy neonate who develops progressive ascites and abdominal distension, irritability, and fluctuating mild jaundice. Biliary peritonitis and lesser sac giant choledochal pseudocyst with acholic stools are rare presentations [8,9]. Our case was unique in which initial perforation into the lesser sac with localized inflammation and omental sealing of the foramen of Winslow probably had led to the formation of choledochal pseudocyst in the lesser sac stretching its boundaries in all directions involving periporal and ligamentous extensions. This also led to external compression of the common bile duct leading to acholic stools, which resolved following decompression of the pseudocyst.

Ultrasound and computerized tomography scans are helpful imaging tools but if the index of suspicion is very high, a technetium-99m disofenin hepatobiliary scan or MRCP may be able to show leakage into a cystic structure within the porta hepatitis, confirming the presence of a bile leak. MRCP could help distinguish anatomy more clearly. In 45%–60% of patients, the dilated pseudosac may prompt misdiagnosis of a choledochal cyst [8]. Atypical imaging features resembling a small bowel duplication cyst have also been reported on CT scan [10]. Fortunately, radionuclide hepatobiliary scan is highly sensitive and specific for SBP and is the preoperative test of choice when NSBP is suspected [10]. Leakage of the radiopharmaceutical into the peritoneum with failure of bowel filling confirms the presence of a biliary rupture and may, in some cases, even localize the site of the leak. An operative cholangiogram should be performed to delineate the location of the perforation and exclude ostial obstruction.

Abdominal paracentesis reveals dark yellow–green fluid; the gram stain, the cell count and the elevated protein content and the conjugated bilirubin helps to confirm the diagnosis and percutaneous drainage of the cyst may be established at the same time to decompress the cyst and perhaps may prove to be only therapeutic intervention required. It is useful in detecting bile within the peritoneum and it should prompt further investigations for a persistent leak and distal ductal stricture.

![Fig. 1. CT scan of the abdomen more clearly showing the extent of the cyst with its ramifications in the lesser sac boundary. GB = gall bladder, TL = triangular ligament, FL = falciform ligament, LD = lesser omentum.](image-url)
Ultrasound guided percutaneous drainage, laparoscopic drainage after examination under anesthesia and needle aspiration of the tense cyst and/or intraoperative cholangiogram to confirm the site of leak if preoperative HIDA scan has not shown the leak may all be useful alternatives and less invasive procedures. In our case, we did not do the operative cholangiogram as there was no active bile leak at exploration.

Most authors recommend a conservative approach of draining the abdomen to decompress the biliary tree; spontaneous closure is typical even with distal obstruction, once the biliary tree is decompressed. Several surgical approaches have been described, including simple drainage with or without cholecystectomy, primary repair with or without external drainage, and hepaticojejunostomy if pancreaticobiliary malformation or distal obstruction is present [1–12].

Suture repair of the bile duct or biliary reconstruction remains controversial because of the potential for stricture formation resulting from inflammation. Reported complications have included portal vein thrombosis, bile leak, and cholangitis. In a few cases, further surgery, including biliary revision and portosystemic shunting, was necessary. Early recognition and treatment yield a good prognosis, whereas lack of surgical treatment results in universal mortality.

3. Conclusion

Neonatal spontaneous bile duct perforation can be missed and the initial presentation to pediatric medical team did not consider this possibility. This presentation has not been reported earlier in the literature so far. NSBP can present as neonatal biliary ascites or peritonitis in generalized form and can present as pseudocholedochal cysts in the periperal tissues or associated ligaments around it in the localized forms. Our case presented as regional compartmentalization to lesser sac as giant pseudocholedochal cyst together with several small daughter periportal and ligamentous pseudocholedochal cysts confirmed by CT and found at exploration.

References