Radiology Case Reports

Congenital tracheobiliary fistula combined with hypoplastic common hepatic duct: Management by percutaneous transhepatic drainage

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Congenital tracheobiliary or bronchobiliary fistula is a rare developmental anomaly with a persistent communication between the biliary system and the trachea or bronchus. We report a case of a congenital tracheobiliary fistula and hypoplastic common hepatic duct associated with hypoplastic left heart syndrome in a 5-day old boy presenting with bilious endotracheal-tube secretions. The tracheobiliary fistula was treated by surgical resection. Subsequent cholangiography demonstrated dilated intrahepatic bile ducts and a residual fistulous tract with cystic proximal stump that were successfully decompressed by transhepatic drainage catheters before corrective biliary surgery.

Introduction

Communication between the respiratory tree and biliary system is a rare congenital anomaly often detected at an early age due to symptoms of respiratory distress or infection. Bilious lung secretions can also be present.

Since the first case of congenital bronchobiliary fistula was reported by Neuhauser et al. in 1952 (1), 31 cases have been described in the literature to date. The anomaly is usually managed by surgical ligation, resection of the fistulous tract, or hepatic lobectomy and reimplantation of the fistula into the gallbladder or bowel (2, 3). If the case is complicated by additional anomalies, such as atresia or hypoplasia of the extrahepatic bile duct, interventional procedures for temporary relief of symptoms before the

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surgical correction of combined anomalies may be required.

Case report

A 35-weeks-gestation male was born with a hypoplastic left heart (diagnosed prenatally) and admitted to the Pediatric Intensive Care Unit for preoperative surgical correction of the heart problem. Postnatal echocardiogram on the next day revealed a complex cardiac anatomy that included left ventricular hypoplasia with a large muscular ventricular septal defect, a hypoplastic transverse aortic arch, an unbalanced atrioventricular canal with a tresia of left atrioventricular valve, an atrial septal defect, and a large patent ductus arteriosus.

On the second day of admission, the baby was intubated due to respiratory distress and thick brownish secretions from the endotracheal tube. Chest radiography was normal, and laboratory tests were significant for hyperbilirubinemia. After bronchoscopy, a large opening was seen at the carina containing bilious secretions. The nasogastric tube aspiration was normal and did not show any bilious secretions. A temporary clinical diagnosis of congenital fistula between the trachea and the biliary system was made.

At the age of 5 days, the baby underwent a right thoracotomy. The anomalous communication from the carina to the medial liver dome was identified by direct visualization. The bronchoscope was able to be advanced into the anomalous communication, which was confirmed as a congenital fistula. The fistula was subsequently opened just

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below the carina, and the airway was closed with interrupted sutures. The distal portion of the fistula was also explored to below the diaphragm and resected. At the liver dome level, the residual portion of the fistula was closed in a similar fashion.

Histological examination of the resected specimen of fistula showed submucosal glands and fibrous soft tissue with fragments of benign cartilage (Fig. 1).



Figure 1. Photomicrograph of the specimen obtained from the proximal portion of the fistula shows a cartilage (arrows) associated with fibrous soft tissue and submucosal glands (arrowheads).

Followup CT scan of the abdomen and pelvis, including inferior portion of the chest (parameters: 1.2 mm collimation, 120 kVp, 47 mAs; CT dose index 3.19 mGy; 6 ml of Omnipaque 300 mgI/ml), was performed one month after the surgery. A fluid-filled tubular structure was seen, extending from the cystic structure at the mediastinal base through the esophageal hiatus and into the liver (Fig. 2). This finding was consistent with a residual fistulous tract with cystic dilatation of the proximal stump in the mediastinal base.

A Tc-99m hepatobiliary scintigraph was obtained approximately two months after the surgery for evaluation of prolonged hyperbilirubinemia. This demonstrated homogeneous uptake of radiotracer in the liver parenchyma without visualization of the biliary trees or small bowel, leading to a suspicion of biliary atresia.

A week later, the patient underwent surgical gastrostomy tube placement for a long-term enteric access. An intraoperative cholangiogram performed at the time of the surgery revealed a hypoplastic common hepatic duct and patent common bile duct.

A week after the operative cholangiogram, the patient was referred to Interventional Radiology for further evaluation of the biliary system and placement of a drainage catheter. Because the patient was stable, it was decided to



Figure 2. CT images of the lower chest, abdomen, and pelvis. A. Axial image just above the diaphragm demonstrates a fluid-filled oval structure (arrow) in front of the esophagus. This structure is consistent with cystic dilatation of the proximal stump after surgery. B. At the level of the liver dome, a fluid-filled residual fistulous tract (arrow) extends into to the left hepatic lobe. C. Sagittal reformation image demonstrates cystic structure (black arrow) and residual fistulous tract (white arrow) extending from the mediastinal base into the liver.

perform percutaneous biliary drainage catheter placement as an elective interventional procedure. A percutaneous transhepatic cholangiogram demonstrated bilateral intrahepatic biliary dilatation and nonfilling of the common hepatic duct. There was also a markedly dilated residual tract of fistula connecting the left intrahepatic bile duct to the cystic dilatation of the proximal stump in the base of the mediastinum (Fig. 3A). However, no persistent communication was identified between the residual fistulous tract and the trachea.



Figure 3A. Cholangiogram demonstrates dilated intrahepatic bile ducts (short arrow) and residual fistulous tract (arrow head) with cystic dilatation (long arrow) at the mediastinal base. Contrast extravasation is seen in the liver parenchyma (double arrows). There is a severe stricture at the level of the common hepatic duct (open arrow), corresponding to the hypoplasia of common hepatic duct. There is no communication between the cystic dilatation and the trachea or bronchus.

Attempted catheterization of the common hepatic duct failed due to severe narrowing. An external biliary drainage catheter was placed in the left intrahepatic bile duct, with the tip of the catheter positioned in the proximal portion of the left intrahepatic bile duct. The decision for the initial left biliary drainage catheter placement was made based on the operator's choice and the recent right thoracotomy incision, with dressings that were close to the right liver margin. However, the direction and position of the left biliary drainage catheter was unfavorable for sufficient drainage of the residual fistulous tract. Therefore, an additional rightsided transhepatic drainage catheter was placed through the residual fistulous tract with the pigtail formed in the cystic portion at the mediastinal base (Fig. 3B). The biliary



Figure 3B. A fluoroscopic image shows decompressed biliary tree and residual fistulous tract with cystic proximal stump in the mediastinal base. Left-sided external transhepatic drainage catheter (short arrow) is positioned in the left intrahepatic bile duct, and right-sided transhepatic drainage catheter (open arrow) is placed through the residual fistulous tract with the pigtail formed in the cystic portion (long arrow) at the mediastinal base.

tree and residual fistulous tract with cystic proximal stump were decompressed and the hyperbilirubinemia resolved.

The baby subsequently underwent resection of the common hepatic duct with creation of Roux-en-Y hepaticojejunostomy. A followup cholangiogram obtained two weeks after the surgery demonstrated a decompressed cystic portion of the proximal stump of the residual fistulous tract and biliary tree, with good emptying of contrast medium via biliary-enteric anastomosis into the jejunum. Bilateral transhepatic drainage catheters were therefore removed. No additional surgery was performed to remove the residual fistulous tract with cystic dilatation of the proximal stump in the base of the mediastinum.

The hypoplastic left-heart syndrome was managed by hybrid stage I palliation and later stage II palliation with a Norwood procedure and a Glenn bidirectional shunt. The patient recovered well following the surgery and was discharged at the age of 6 months in a stable condition.

Discussion

Congenital tracheobiliary or bronchobiliary fstula is a rare anomalous communication between the carina or the main bronchus and the biliary system. To date, only 31 cases (summarized in the table. p. 6) have been reported in the literature.

In most cases, the fistula was reported to originate either from the right main bronchus (42%) or carina (42%). In

only 16%, the fistula originated from the left main bronchus. The fistula was reported to pass through the esophageal hiatus and enter the left bile duct in 21 cases (68%). In only one case, the fistula entered into the common hepatic duct (1). In nine cases, the site of insertion in the biliary system was not specified.

The fistula was diagnosed at an early age (9 males, 18 females; mean age, 381 days; median age, 32 days; age range, 12 hours to 6 years) in most cases (87%). Only four cases were diagnosed in adults (1 male, 3 females; mean age, 38 years; age range, 22 to 65 years) (4-7). The most common presenting symptom was respiratory, including cough, dyspnea, cyanosis, bilious sputum, and pulmonary infection. However, these symptoms are nonspecific, and other pathologic conditions (including tracheoesophageal fistula, gastroesophageal reflux, gastrointestinal obstruction, and aspiration pneumonia) should be included in the differential diagnosis (8).

Bronchoscopy has been the most commonly used (52%) method for diagnosis, followed by hepatobiliary scintigraphy (Tc99m HIDA scan) and bronchography. Recently, two cases using multidetector CT with multiplanar and 3D reformations to delineate the fistula have been reported by Gunlemez et al. (9) and Chawla et al. (10), respectively. In one case, MRI was successfully used for depicting a small bronchobiliary fistula and isolated left bile duct (3). Our case was initially diagnosed only by bronchoscopy (Fig. 1) and later confirmed by surgery. After surgery, CT showed a fluid-filled residual fistulous tract and cystic dilatation in the base of the mediastinum (Fig. 2).

The treatment of choice for the fistula is surgical repair. In 28 cases, the fistula was repaired successfully by thoracotomy with either resection (89%) or ligation (11%). In one case, the patient died of sepsis the day after surgical resection of the fistula. Surgical repair was not performed in three patients who expired after the initial diagnosis. In our case, the fistula originated from the carina and inserted into the left intrahepatic bile duct through the esophageal hiatus. This was repaired by right thoracotomy with resection of the proximal fistulous tract.

Congenital bronchobiliary or tracheobiliary fistula is frequently associated with other abnormalities. Among 31 cases, various coexisting abnormalities have been reported in 17 (55%). Of those 17 cases, 11 cases (65%) were found to have congenital anomalies in the biliary system. If the case is complicated by combined anomaly of the biliary system, the treatment options should include fistula-enteric anastomosis, Roux-en-Y anastomosis, cholecystoduodenostomy, cholecysto-jejunostomy, and resection of the involved hepatic lobe. Hourigan et al. (3) reported a case of congenital bronchobiliary fistula connecting to the left hepatic duct that did not communicate with the common hepatic duct. The fistula was therefore dissected proximally and carried down through the esophageal hiatus to the gallbladder. Guadere et al. (2) also reported a similar case of a fistula combined with an isolated left bile duct without communication to the common hepatic duct; this was corrected by Roux-en-Y anastomosis.

In our case, the fistula was associated with hypoplasia of the common hepatic duct. Despite the initial laboratory test showing hyperbilirubinemia at the admission, the diagnosis of hypoplastic common hepatic duct was delayed due to the patient's partial response to phototherapy and the complicated postsurgical course. However, if proper additional diagnostic workup had been performed before the surgery for the tracheobiliary fistula, the hypoplastic common hepatic duct could have been managed simultaneously. Subsequently, a percutaneous, transhepatic, biliary drainage catheter was placed for temporary relief of hyperbilirubinemia. The dilated residual fistulous tract with cystic dilatation of the proximal stump was also successfully decompressed by an additional right-sided transhepatic drainage catheter that was placed through the residual fistulous tract, with the pigtail formed in the cystic portion in the base of the mediastinum before Roux-en-Y hepaticojejunostomy (Figs. 3A, 3B).

The underlying pathogenesis of congenital fistula is still not clear. However, two possible embryological mechanisms have been suggested: (a) fusion of an anomalous bronchial bud with an anomalous bile duct; (b) duplication of the upper gastrointestinal tract (1, 11). These are supported by histological evaluations of the resected fistulous tract that were described in 12 cases. They showed features of bronchial epithelium with submucosal glands, respiratory smooth muscle, and cartilage in the proximal portion of the specimen. Histological evaluation of the distal portion of the tract was performed in 8 cases that showed epithelium resembling that found in the biliary system and esophagus in 5 and 2 cases, respectively. In one case, reported by de Carvalho et al. (5), the resected specimen demonstrated normal cartilaginous rings and respiratory epithelium with areas of squamous metaplasia in the proximal portion, gastric mucosa and pancreatic tissue in the mid-portion, and biliary epithelium in the distal portion. In our case, the specimen was obtained only from the proximal portion of the fistula, and histological examination showed submucosal glands with fibrous tissue containing fragments of cartilage that corresponded to previous reported cases (Fig. 1).

In conclusion, congenital tracheobiliary or bronchobiliary fistulas are associated with additional biliary abnormalities and in this case with hypoplasia of the common hepatic duct and hypoplastic left heart syndrome. The cholangiogram demonstrated a residual distal fistulous tract with cystic dilatation of the proximal stump following surgical resection of the fistula without evidence of persistent communication to the trachea. Percutaneous transhepatic biliary drainage was useful for temporary relief of hyperbilirubinemia and for decompression of the residual mediastinal cyst before corrective biliary surgery.

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	Case report		Patient		Fistula		Diagnostic		_
NO.	Authors	Year	Age*	Sex	Туре	Connection	method	Combined abnormality	Treatment
1	Neuhauser et al.	1952	5 m	F	BBF	RMB-CHD	Bronchography	None	None, expired
2	Enjoji et al.	1963	7 m	М	BBF	RMB-NS	Autopsy	Stenosis of common bile duct	None, expired
3	Stigol et al.	1966	14 m	F	BBF	RMB-NS	Bronchography	None	Resection
4	Weitzman et al.	1968	2 y, 9 m	F	BBF	RMB-LT	Bronchoscopy	None	Resection
5	Wagget et al.	1970	3 w	F	BBF	LMB-LT	Bronchoscopy	Isolated LBD	Resection
6	Sane et al.	1971	4 w	F	BBF	RMB-NS	Bronchoscopy	None	Resection
7	Cuadros et al.	1974	6 y	Μ	TBF	Carina-LT	Bronchoscopy	None	Resection
8	Kalayoglu et al.	1976	4 d	F	BBF	RMB-NS	Incidental finding	Esophageal atresia Tracheoesophageal fistula	Resection
9	Chan et al.	1984	4 d	F	BBF	RMB-NS	Fistulogram**	Atresia of CBD	Resection
10	Chang et al.	1985	12 h	F	BBF	RMB-NS	Bronchography	None	Resection
11	Lindahl et al.	1986	1.5 d	F	BBF	RMB-NS	Bronchoscopy	None	Resection
12	Levasseur et al.	1987	22 y	F	TBF	Carina-LT	Bronchoscopy	Choledochal cyst	Resection
13	de Carvalho et al.	1988	, 32 y	F	BBF	RMB-LT	^{99m} Tc HIDA scan	None	Resection
14	Mavunda et al.	1989	1 y	F	BBF	LMB-LT	99mTc HIDA scan	Isolated LBD	Ligation
15	Yamaguchi et al.	1990	32 y	М	BBF	RMB-LT	Bronchoscopy	None	Resection
16	Gauderer et al. [†]	1993	2 y	М	BBF	Carina-LT	Bronchoscopy	Isolated LBD	Resection
17	Tekant et al.	1994	15 d	F	TBF	Carina-NS	Bronchoscopy	Atresia of CBD	Resection
18	Ferkol et al.	1994	23 m	М	BBF	LMB-LT	Bronchoscopy	None	Resection
19	Egrari et al.	1996	3 d	F	TBF	Carina-NS	99mTc HIDA scan	None	Resection
20	Fischer et al.	1998	16 d	F	BBF	LMB-LT	Bronchography	None	Resection
21	Tommasoni et al.	2000	21 m	F	TBF	Carina-LT	Bronchoscopy	None	Resection
22	Tommasoni et al.		30 m	М	TBF	Carina-LT	Bronchoscopy	Brachycephaly, hypertelorism, facial asymmetry, pectus excavatum, arachnodactyly	Resection
23	Duong et al.	2000	3 у	F	TBF	Carina-LT	Bronchoscopy	None	Resection
24	DiFiore et al.	2002	Newborn	Μ	BBF	RMB-LT	Incidental finding at surgery	Right-sided congenital dia- phragmatic hernia	Resection
25	Hourigan et al.	2004	13 d	Μ	BBF	RMB-LT	MRI	Isolated LBD	Resection & reim- plantation
26	Aguilar et al.	2005	6 y	F	TBF	Carina-EHD	99mTc HIDA scan	None	Surgery [‡]
27	Chawla et al.	2008	Newborn	М	TBF	Carina-LT	MDCT	Hypoplasia of CBD	Resection
28	Uramoto et al.	2008	65 y	F	TBF	Carina-LT	Bronchoscopy	Lung cancer	None, expired ^{‡‡}
29	Gunlemez et al.	2009	9 d	F	BBF	LMB-LT	MDCT	Atresia of CBD	Ligation
30	Najdi et al.	2009	6 d	F	TBF	Carina-NS	Bronchoscopy	None	Resection, expired
31	Croes et al.	2010	3 d	F	TBF	Carina-LT	Bronchoscopy	Aberrant LBD	Ligation
32	Present Case	2010	5 d	М	TBF	Carina-LT	Bronchoscopy	Hypoplastic left heart syn- drome Hypoplasia of CHD	Resection

Notes:

Age* = The age at diagnosis

BBF = Bronchobiliary fistula

TBF = Tracheobiliary fistula

h = hour, d = day, w = week, m = month, and y = year

M = male, F = female

RMB = Right main bronchus, LMB = Left main bronchus

CHD = Common hepatic duct, CBD = Common bile duct, EHD = Extrahepatic bile duct, LT = Left hepatic or bile duct NS = Fistula entered into the intrahepatic bile duct but the side was not specified

Fistulogram** = Fistulogram via suction catheter of endotracheal tube

99mTc = 99mTechnetium

HIDA= Hepatobiliary iminodiacetic acid

MRI = Magnetic resonance imaging

MDCT = Multidetector computed tomography

 \dagger = Gauderer et al (16) reported three cases; however, the first and third cases were followups of previously presented cases by Wagget et al (5) and Mavunda et al (14) in 1970 and 1989, respectively.

 \dagger = Barlocco et al (31) reported two cases in 1992 that were the same cases presented by Tommasoni et al in 2000 (21).

= The type of surgery was not specified.
= The patient died of lung cancer 28 months after diagnosis of congenital BBF.