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Congenital Bronchobiliary Fistula: Report of A Successfully Operated Case

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Abstract **We described a case of congenital bronchobiliary fistula who had successfully been treated in the early age and reviewed 22 previously reported cases of this anomaly. This condition is a rare cause of respiratory distress in newborn infants and should be included in the differential diagnosis of aspiration pneumonia.**

Congenital bronchobiliary fistula is a rare anomaly characterized by a fistulous tract communicating between the respiratory and the biliary systems. There was a case report published in 1889 by Chiari and cited by Ballantyne[1] in 1905, in which the right main bronchus had a double lumina but there was no communication with the liver or the biliary system being demonstrated. The first proven case of congenital bronchobiliary fistula was reported by Neuhauser et al[2] in 1952. Up to 2000, only 22 cases have been reported in the literature.[2-21] We herein presented our experience in a case of congenital bronchobiliary fistula and summarized the previously reported ones.

Case Report

A 2-day-old female infant was admitted to Queen Sirikit National Institute of Child Health with dyspnea. She was delivered elsewhere by Caesarean section due to cephalopelvic disproprotion at 38 weeks of gestation before referal. Her birth weight was 3,150 grams. Her clinical condition immediately after birth was good. On the second day of life, respiratory distress developed. She had tachypnea and mild subcostal retraction. Her chest film revealed perihilar interstitial infiltration. She gradually had respiratory difficulties. At the age of 3 days, the symptoms were worseny and she required

endotracheal intubation and mechanical ventilation. Chest film after intubation showed right lung atelectasis with patchy infiltration (Figure 1), consistent with aspiration pneumonia. She had large amount of secretion via the endotracheal tube. An esophagography was performed with a suspected diagnosis of H-type tracheoesophageal fistula. The study revealed severe gastroesophageal reflux and nasopharyngeal incoor-dination, but no evidence of tracheoesophageal fistula was noted. Her secretion in the endotracheal tube did not decrease despite continuous treatment. The secretion was mucoid and yellow-green in color. Analysis of specimen of the secretion showed 25.0

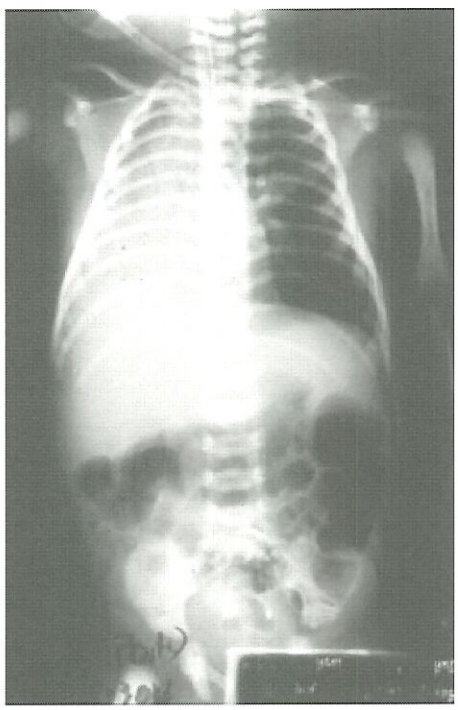

Fig. 1 Chest film after intubation showed right lung atelectasis.

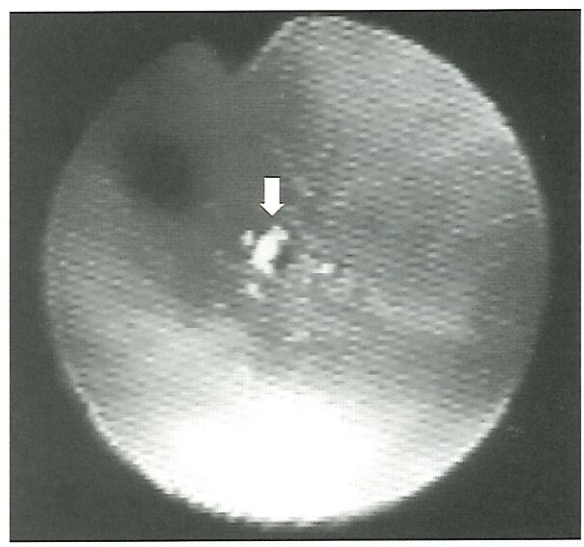

Fig. 2 View of tracheal bifurcation during flexible broncoscopy. The anomalous opening of the broncobiliary fistula was present at the carina (denoted by the arrow).

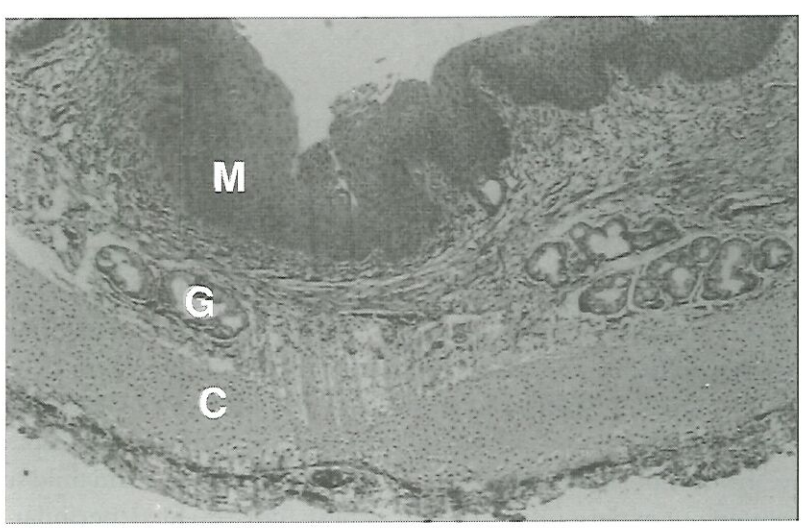

Fig. 3 Histologic finding of the excised fistulous tract
M = mucosa
G = grandular structure
C = cartilage

mg/dl of total bilirubin and 21.7 mg/dl of direct bilirubin. Then, bronchoscopy was performed with a suspected diagnosis of a bronchobiliary fistula. At bronchoscopy, green-yellow mucoid material was seen on the tracheal wall. An anomalous orifice was identified at the carina, mid way between the right and the left main bronchi (Figure 2), hence the diagnosis of bronchobiliary fistula was confirmed. An operation was undertaken on the following day. A right transpleural thoracotomy was performed and a fistulous tract connecting between the two main bronchi was identified and transected. The proximal orifice at the carina was repaired with 5-0 Vicryl interruptedly. The distal orifice of the tract was cannulated and fistulography was done under fluoroscopy. The study revealed diffuse filling of the contrast medium over both lobes of the liver which fainted over delayed time. Then the distal tract was dissected further down and excised 2.5 cm below the carina. The distal stump was suture-ligated with 5-0 Vicryl. Histological finding of the tract showed the respiratory tract lining epithelium (Figure 3). The patient was extubated on the seventh postoperative day. Unfortunately she had postoperative surgical wound infection. The causative organism was methicillin-resistant Staphylococcus aureus, sensitive only to vancomycin. Later, she developed osteomyelitis of the adjacent ribs requiring a long period of antibiotic administration. Finally, she was discharged in good condition on the 58th postoperative day when she was 79 days old. At the last follow-up when she was vaccinated at the age of 6 months, her clinical condition was good and healthy. Her body weight was 6 kilograms.

Table 1 Summary of the previously reported cases.

Case No	Year	Reporter	Sex	Age of onset	Age at diagnosis	Presenting symptoms	Diagnostic method	Result
1	1952	Neuhauser[2]	F	Since birth	5 mo	Recurrent pneumonia	Bronchoscopy	Dead
2	1958	Haight[6]	F	Since birth	3 mo	NA	Bronchoscopy	Survived
3	1963	Enjoji[3]	M	Since birth	7 mo	Recurrent pneumonia	Bronchogram autopsy	Dead
4	1966	Stigol[4]	F	Since birth	14 mo	Recurrent pneumonia	Bronchogram	Survived
5	1968	Weitzman[5]	M	Early infancy	33 mo	Recurrent pneumonia vomiting	Bronchoscopy	Survived
6	1970	Sane[6]	F	Since birth	4 mo	Bilious vomiting	Bronchoscopy	Survived
7	1970	Wagget[7]	F	Since birth	3 mo	Cough, vomiting	Bronchoscopy	Survived
8	1974	Caudros[8]	NA	NA	6 yr	NA	NA	Survived
9	1976	Kalayoglu[9]	F	NA	4 days	Drooling	Autopsy	Dead
10	1984	Chan[10]	F	NA	4 days	Dyspnea and drooling	Bronchogram	Dead
11	1984	Dyon[11]	NA	NA	12 hr	NA	NA	Dead
12	1985	Chang[12]	M	12 hr	1 mo	Bilious vomiting	Bronchoscopy	Survived
13	1986	Lindahl[13]	F	1.5 days	3 days	Respiratory distress	Bronchoscopy	Survived
14	1988	Carvalho[14]	F	2 mo	32 yr	Fever, productive cough	Bronchoscopy, HIDA scan	Survived
15	1989	Mavunda[15]	F	3 days	1yr	Bilious regurgitation	Bronchoscopy	Survived
16	1992	Barlocca[16]	F	Since birth	21 mo	Respiratory distress	HIDA scan Bronchoscopy	Survived
17	1992	Barlocca[16]	M	First month	30 mo	Chronic cough	Bronchoscopy	Survived
18	1993	Gauderrer[17]	M	1 day	23 mo	Recurrent pneumonia	Bronchoscopy	Survived
19	1994	Tekant[18]	F	1 day	> 2 mo	Respiratory distress	Bronchoscopy	Survived
20	1996	Egrari[19]	F	3 days	Soon	Respiratory distress	HIDA scan	Survived
21	1998	Fisher[20]	F	NA	New born	GER	Bronchoscopy	Survived
22	2000	Duong[21]	F	NA	3 yr	Respiratory problem	Bronchoscopy	Survived

NA = no available data, F = Female, M = Male, GER = Gastroesophageal reflux, hr = hour (s), wk = week (s), mo = month (s), yr = year (s)

Table 2 Summary of the previously reported cases.

Case No.	Operation	Other anomalies	Location of the fistula	
			Proximal	Distal
1	Dead before operation		Rt main bronchus	Lt hepatic duct
2	Excision of fistula		NA	NA
3	Roux-en-y	Stenosis of CBD	Tracheal bifurcation	Hepatic duct
4	T, DOF		Rt main bronchus	NA
5	T, DOF		Rt main bronchus	Lt hepatic duct
6	T, DOF		Tracheal bifurcation	Intrahepatic biliary tree
7	T, DOF, Lt lobectomy		Tracheal bifurcation	Lt hepatic duct
8	T, DOF		NA	NA
9	T, DOF, CE	EA with distal TEF	Rt main bronchus	NA
10	T, DOF	Absence of CBD	Rt main bronchus	Hepatic duct
11	NA	Choledochal hypoplasia	NA	NA
12	DOF		Rt main bronchus	NA
13	T, DOF		Rt main bronchus	Lt lobe of liver
14	T, DOF, pneumonectomy		Rt main bronchus	Lt biliary duct
15	T, DOF, Roux-en-Y Fistulojejunostomy		Lt main bronchus	Lt hepatic duct
16	T, DOF	Tracheal bifurcation	Intrahepatic duct	
17	T, DOF	Rt main bronchus	NA	
18	T, DOF, Lt hepatectomy	Tracheal bifurcation	Lt hepatic duct	
19	T, DOF Cholecystoduodenostomy	Absence of CBD	Tracheal bifurcation	Intrahepatic duct
20	T, DOF, IOC		Tracheal bifurcation	NA
21	T, DOF		Tracheal bifurcation	NA
22	T, DOF		Tracheal bifurcation	Lt hepatic duct

NA = no available data, T = Thoracotomy, DOF = Division of Fistula, IOC = Intraoperative cholangiography, CBD = Common bile duct, Lt = Left, Rt = Right, EA = Esophageal atresia, TEF = Tracheoesophageal fistula, CE = Cervical esophagostomy

Discussion

Congenital bronchobiliary fistula is a rare developmental abnormality. The pathogenesis of this anomaly is not clear. Two possible embryological mechanisms had been postulated. One was the union of an anomalous bronchial bud with an anomalous bile duct. The other was duplication of the upper gastrointestinal tract.[6] Summaries of the 22 cases previously reported in the literature[2-21] are shown in Tables 1-3. The clinical presentation of most cases was respiratory symptom. Female was more commonly affected than male (15:5). Bronchoscopy with contrast studies had been the mainstay of diagnosis in most cases. HIDA scan had been employed in 3 patients. In all but one case the diagnosis had been made at an early age while all of them developed the symptoms during neonatal period. Thoracotomy and division of the fistula was the definitive treatment. The most common location of the proximal fistula was at the right main bronchus, while the distal fistula communicated with the left hepatic duct. Associated anomalies were noted in five patients, four of whom involving the biliary tract. On microscopic examination of the fistula, the proximal portion resembled the bronchus in 12 cases, resembled the esophagus in one case and resembled the biliary system in one case. Five patients died during the first year of life, four from pulmonary complication as a consequence of the malformation, the other from multiple anomalies and septicemia.

In conclusion, the early onset of respiratory distress together with copious bile-appearance secretion from

Table 3 Microscopic finding of the fistula.

Case No.	Proximal portion	Distal portion
1	Stratified squamous epithelium	Stratified columnar epithelium
2	Stratified squamous epithelium	Stratified squamous epithelium
3	Pseudostatified columnar epithelium	Columnar epithelium
4	Resembling a bronchus	Resembling an esophagus
5	Squamous columnar epithelium	Modified columnar
6	Resembling a bronchus	Resembling an esophagus or bile duct
7	NA	NA
8	NA	NA
9	Resembling a biliary tract duct	NA
10	Resembling a bronchus	NA
11	NA	NA
12	Resembling a bronchus	Stratified squamous epithelium
13	Stenosed dysplastic bronchus	NA
14	Resembling a bronchus	Resembling a biliary tract duct
15	NA	NA
16	NA	NA
17	Resembling a bronchus	NA
18	NA	NA
19	Respiratory epithelium	Respiratory epithelium
20	NA	NA
21	Esophageal origin	NA
22	NA	NA

NA = no available data

the trachea directed us to the clinical diagnosis of congenital bronchobiliary fistula in our patient. Bronchoscopy was mandatory in establishing the location of the proximal fistulous opening. Excision of the fistula definitely relieved the patient's deadly condition.

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