

Congenital bronchobiliary fistula: a case report and literature review

Le Thuong Vu^{1†}, Nguyen Minh Duc^{2,3,4†}, Thieu-Thi Tra My⁴, Mai Tan Lien Bang², Le Tuan Linh⁴ & Tran Quynh Huong⁵

¹Department of Internal Medicine, University of Medicine and Pharmacy at Ho Chi Minh City, Ho Chi Minh City, Vietnam.

²Department of Radiology, Children's Hospital 2, Ho Chi Minh City, Vietnam.

³Department of Radiology, Pham Ngoc Thach University of Medicine, Ho Chi Minh City, Vietnam.

⁴Department of Radiology, Ha Noi Medical University, Ha Noi, Vietnam.

⁵Department of Respirology, Children's Hospital 2, Ho Chi Minh City, Vietnam.

Keywords

Bronchoscopy, computed tomography, congenital bronchobiliary fistula, fistulography.

Correspondence

Nguyen Minh Duc, Department of Radiology, Pham Ngoc Thach University of Medicine, Ho Chi Minh City 700000, Vietnam. E-mail: bsnguyenminhduc@pnt.edu.vn

Received: 9 December 2020; Revised: 4 February 2021; Accepted: 15 February 2021;

†These authors contributed equally as co-first authors. Associate Editor: Cameron Sullivan.

Respirology Case Reports, 9 (4), 2021, e00731

doi: 10.1002/rcr.2.731

Introduction

Congenital bronchobiliary fistula (CBBF) is a rare, anomalous fistula that occurs between the respiratory and biliary systems, with unknown aetiology [1]. The first CBBF case was reported in 1952 by Neuhauser [2]. The tract often connects trachea or bronchus with the intrahepatic bile duct or common bile duct [3]. CBBF can occur at any age, from newborns to adults, and the presence of respiratory symptoms is thought to be associated with the fistula diameter [4]. Patients with CBBF may present with symptoms including respiratory distress, recurrent pneumonia, cough with dark-yellow sputum, and choking [5]. Surgical resection is the most appropriate and effective treatment for most cases [4].

Case Report

A full-term, female, two-day-old baby was admitted to the hospital with symptoms of choking, coughing, vomiting, bilious sputum, and cyanosis. She had a heart rate of

Abstract

Congenital bronchobiliary fistula (CBBF) is a very rare malformation associated with clinical symptoms of dyspnoea, vomiting, cyanosis, persistent pneumonia, and poor response to antibiotics. Typical imaging techniques used to diagnose this malformation include bronchography, computed tomography (CT), cholangiography, hepatobiliary nuclear imaging, and magnetic resonance imaging (MRI). We diagnosed a case of CBBF that was initially diagnosed as non-resolving pneumonia. CT and fistulography were used to obtain the correct diagnosis. The fistula was confirmed by fistulography under the guidance of bronchoscopy. Surgical excision of the fistulous tract was performed, with complete recovery. The aim of this report was to emphasize the epidemiology and clinical features of CBBF patients.

100 beats per minute, a temperature of 37°C, a rapid respiratory rate of 60 breaths per min, and oxygen saturation of 96% when breathing room air. Physical examination showed subcostal retraction, with crackling sounds in the right lung. Laboratory studies revealed normal white blood cell count and neutrophils. Three sputum cultures failed to detect any bacteria. Sputum was obtained by suctioning through the oropharynx using a sterile, suction catheter. Sputum respiratory syncytial virus polymerase chain reaction (PCR) was negative. A chest X-ray showed opacity in both hilar regions and the right lower lobe area (Fig. 1A). The patient was diagnosed with pneumonia and was treated with ampicillin and cefotaxime. One month later, the symptoms improved but did not disappear. Therefore, the patient was treated with broader spectrum antibiotics, including ciprofloxacin and imipenem, but the respiratory symptoms continued after two months. Because the symptoms did not respond to antibiotic therapy, congenital pulmonary or upper gastrointestinal anomalies were

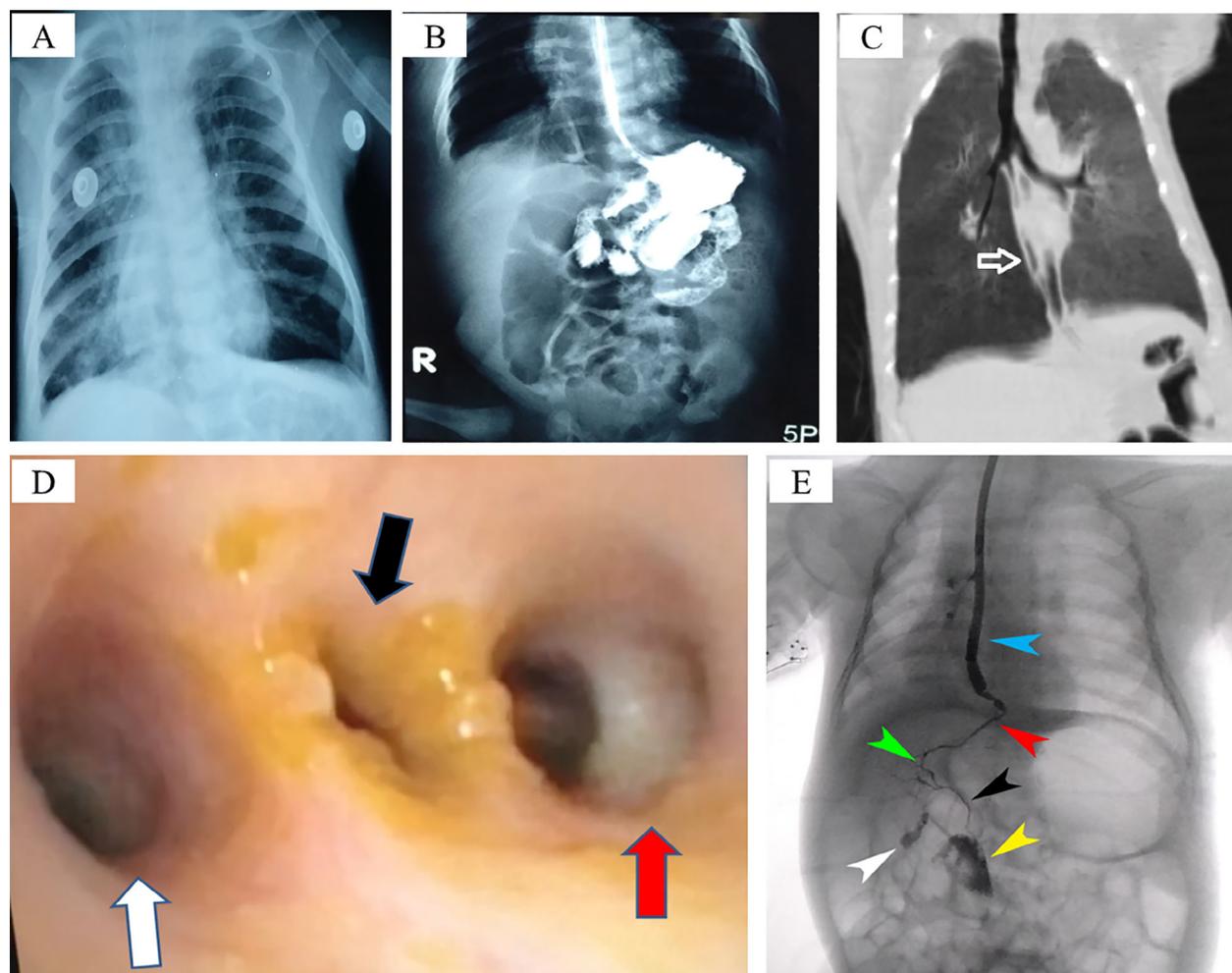


Figure 1. Chest X-ray showed opacification in both the hilar region and the right lower lobe area (A). No tracheoesophageal fistula, gastroesophageal reflux, intestinal malrotation, or bowel obstruction was observed on upper gastrointestinal tract radiography (B). Chest computed tomography revealed an abnormal, air-filled tract that opened to the carina and passed through to the diaphragm (arrow). The maximum and minimum diameters of the tract were 3 and 1 mm, respectively (C). Bronchoscopy showed a bile-like mucus being discharged from an anomalous orifice (black arrow) located between the right bronchi (white arrow) and left main bronchi (red arrow) (D). Fistulography image demonstrated that the contrast material flowed from the endoscopy tube (blue arrowhead) through anomalous tract (red arrowhead) to the left hepatic biliary duct (green arrowhead), gallbladder (white arrowhead), common bile duct (black arrowhead), and duodenum (yellow arrowhead) (E). The contrast in both main bronchi was also detected (white and red arrowheads).

suspected. Upper gastrointestinal tract radiography revealed the absence of tracheoesophageal fistula, gastroesophageal reflux, intestinal malrotation, and bowel obstruction (Fig. 1B). Chest computed tomography (CT) revealed an abnormal tubule extending from the carina that passed through the diaphragm into the left hepatic biliary tract (Fig. 1C). The tube contained gas and fluid; thus, bronchobiliary fistula (BBF) was suspected. A bronchoscopy with fistulography was performed on this patient. The bronchoscopy demonstrated yellow fluid secretion originating from an anomalous orifice between the right and left main bronchi (Fig. 1D). After moving

the endotracheal tube to the lumen of the lesion, the contrast material (iopromide) was instilled into the endotracheal tube. Radiological images revealed the contrast material passing through the tube to the left hepatic biliary duct, gallbladder, bile duct, and duodenum (Fig. 1E). A fistula that bridged the left hepatic bile duct with the trachea was confirmed. The preoperative diagnosis was CBBF. One day later, the patient underwent surgery, in which we performed a posterolateral right thoracotomy. The fistula formed from the carina to the diaphragm and was approximately 45 mm in diameter length. We removed the fistula tract, and both ends were securely ligated and sutured. A

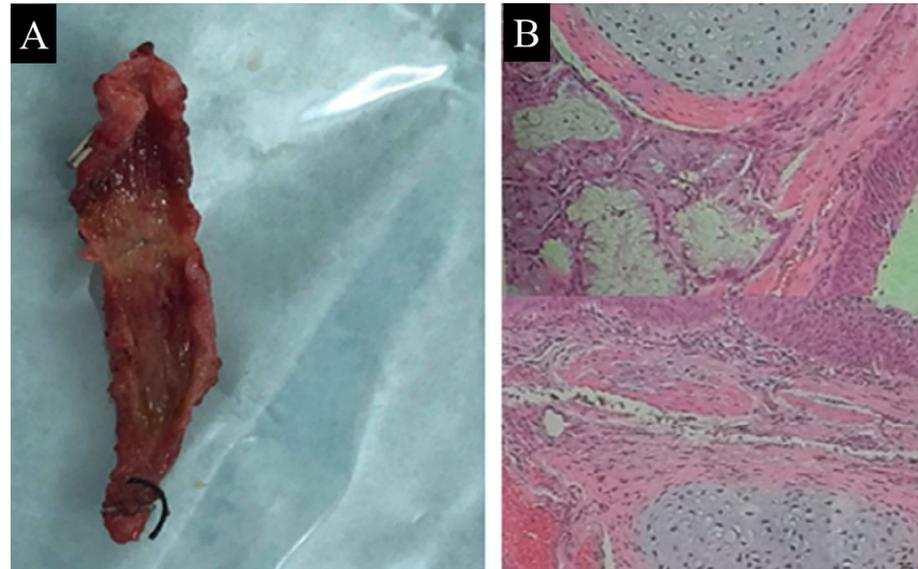


Figure 2. (A) The fistulous tract was resected. (B) Microscopic examination showed that the tract was lined with respiratory epithelium. In the submucosa, glands and muscle tissue from the external region were observed.

right thoracic drainage tube was placed. The histopathological findings demonstrated a tube of tissue that was lined with respiratory epithelium, without biliary epithelium. In the submucosa, the glands and muscle tissue from the external portion could be observed (Fig. 2). At one-week post-surgery, the child was successfully weaned from the mechanical ventilator. She recovered well and was discharged without oxygen.

Discussion

A BBF is a rare condition defined by the passage of bile into the bronchus or trachea [1]. BBF aetiology can be classified into two groups: CBBF and acquired BBF [6]. Acquired BBF is typically the consequence of local infection or trauma, such as hepatic abscess, trauma, biliary tract obstruction, hepatectomy, or radiofrequency thermal ablation of hepatic tumours [7]. CBBF occurs less frequently than acquired BBF, accounting for fewer than 6% of all BBF cases [7]. The development of BBF may be explained by: (1) union of an anomalous bronchial bud with an anomalous bile duct and (2) duplication of the upper gastrointestinal tract [4]. To the best of our knowledge, there have been no reports of a genetic background for this disease. Patients with BBF may have other congenital lesions such as diaphragmatic hernia or biliary atresia [5]. Recently, Li and Zhang identified only 44 reported CBBF cases [3]. The age of onset can range from 12 h to 65 years, and in most cases, the expression of symptoms occurs for less than one year [8]. CBBF occurs at a higher incidence in women than in men [8]. An abnormal fistula most commonly originates near the carina (43%) or the right main bronchus (46%), with only 11% of fistulas originating at the left main

bronchus [3]. The fistula tract descends along the oesophagus and crosses the diaphragm through the oesophageal hiatus. The abdominal portion of the fistula typically ends in the left bile duct [9], although one case has been reported in which the fistula communicated with the common bile duct [9]. Patients with CBBF often present with respiratory symptoms, including cough, dyspnoea, cyanosis, bilious sputum, pulmonary infection, and poor response to antibiotic therapy [8]. However, these symptoms are non-specific. These anomalies have been diagnosed by bronchoscopy, fistulography, cholangiography, CT, magnetic resonance imaging (MRI), and hepatobiliary nuclear imaging [3,5,10,11]. Cholangiography has been used to perform both pre- and intra-operative diagnoses [8,12]. Bronchoscopy and CT are the most common methods used for diagnosis [3]. Bronchoscopy has been used to identify an abnormal opening to the carina of the bronchus from which bile was being secreted [4]. CT can also be employed to indicate the presence of an abnormal tubular structure that connects the respiratory system with the biliary tract [13]. CT can also provide information necessary for the establishment of a surgical plan for CBBF treatment [5]. The histopathological results have revealed that the upper part of the fistula resembles the respiratory tract; however, the lower part of the fistula was lined with epithelial cells from the bile duct [3]. The surgical excision of the tract is the first treatment option, and most cases recover fully [3]. Li and Zhang [3] described three patients who did not undergo surgical management; however, all of them died. It is noted that most of the patients treated with surgery had better outcome and prognosis.

In this case report, the patient presented with persistent pneumonia, vomiting, and bilious sputum. However, in

newborns, bile-stained sputum may pass through when vomiting. Therefore, an important symptom was likely dismissed. We recommend that congenital respiratory malformations should be considered by clinicians as a potential differential diagnosis in newborns with respiratory distress or persistent pneumonia.

In summary, CBBF should be considered as a possibility in neonates with pulmonary infection, respiratory distress, and bilious discharge through the airway. CT may suggest the presence of a BBF, which appears as an air-filled tube that originates from the bronchus or trachea and passes through the diaphragm into the liver. Bronchoscopy associated with fistulography has been demonstrated to be the preferred method of diagnosing BBFs.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Author Contribution Statement

Conceptualization: Le Thuong Vu and Nguyen Minh Duc; data curation: Le Thuong Vu and Nguyen Minh Duc; formal analysis: Le Thuong Vu and Nguyen Minh Duc; writing—original draft: Nguyen Minh Duc and Thieu-Thi Tra My; writing—review and editing: Nguyen Minh Duc and Thieu-Thi Tra My. All authors have read and agreed to the published version of the manuscript.

References

- Gauderer MWL, Oiticica C, and Bishop HC. 1993. Congenital bronchobiliary fistula: management of the involved hepatic segment. *J. Pediatr. Surg.* 28:452–455.
- Chan YT, Ng WD, Mak WP, et al. 1984. Congenital bronchobiliary fistula associated with biliary atresia. *Br. J. Surg.* 71:240–241.
- Li T-Y, and Zhang Z-B. 2019. Congenital bronchobiliary fistula: a case report and review of the literature. *World J. Clin. Cases* 7:881–890.
- Kim JS, Suh JH, Park CB, et al. 2015. Congenital tracheobiliary fistula in an adolescent patient. *Ann. Thorac. Surg.* 99:328–331.
- Na KJ, Jung JC, Hwang Y, et al. 2016. Minimally invasive surgical repair for congenital bronchobiliary fistula in an adult. *Ann. Thorac. Surg.* 101:1584–1587.
- Cao L, Song Z, Liu Q, et al. 2012. A case report of bronchobiliary fistula and literature review. *Infect. Int.* 1:149–150.
- Liao G-Q. 2011. Management of acquired bronchobiliary fistula: a systematic literature review of 68 cases published in 30 years. *World J. Gastroenterol.* 17:3842–3849.
- Yu H, Stavas JM, and Joseph M. 2011. Congenital tracheobiliary fistula combined with hypoplastic common hepatic duct: management by percutaneous transhepatic drainage. *Radiol. Case Rep.* 6:564.
- Pérez CG, Reusmann A, and Collaborators. 2016. Congenital broncho-biliary fistula: a case report. *Arch. Argent. Pediatr.* 114:e350–e353.
- Hourigan JS, Carr MG, Burton EM, et al. 2004. Congenital bronchobiliary fistula: MRI appearance. *Pediatr. Radiol.* 34:348–350.
- Egrari S, Krishnamoorthy M, Yee CA, et al. 1996. Congenital bronchobiliary fistula: diagnosis and postoperative surveillance with HIDA scan. *J. Pediatr. Surg.* 31:785–786.
- Velchik MG, Roth GM, Wegener W, et al. 1991. Bronchobiliary fistula detected by cholescintigraphy. *J. Nucl. Med.* 32:136–138.
- Salzedas Netto AA, Silva PCM, Vicentine FPP, et al. 2018. Congenital tracheo-biliary fistula: staged surgical treatment. *J. Pediatr. Surg. Case Rep.* 32:11–13.