Bronchobiliary fistula: a clinical and surgical challenge. Presentation of a pediatric case

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Abstract

Introduction. Congenital bronchobiliary fistula is an extremely rare malformation with high morbidity and mortality rates. Up to 2016, 36 cases had been reported worldwide.

Clinical case. 11-year-old male patient with history of chronic lung disease and respiratory insufficiency, bile ptyalism and 66-80% arterial saturation, jaundice, asymmetric thorax, finger clubbing, and disseminated crackling rales. He was diagnosed through fibrobronchoscopy and CT-scan. After fistula closure and right pneumonectomy, recurrence occurred due to bile duct hypoplasia as evidenced by endoscopic retrograde cholangiopancreatography. Left lateral hepatic segmentectomy and fistula closure from the abdomen were carried out. Bronchopleural fistula persisted following intensive nutritional and antibiotic treatment. It was surgically closed using a bovine pericardial patch. Six months later, the patient had no symptoms.

Discussion. Given how extremely rare this malformation is, cross-disciplinary treatment and a high grade of suspicion are needed. The presence of bile duct hypoplasia is to be considered, since it requires a thoracoabdominal approach.

Key Words: Fistula; Bronchus; Biliary tract disease.

Fístula biliobronquial, un desafio clínico-quirúrgico. Presentación de un caso pediátrico

Resumen

Introducción. La fístula biliobronquial congénita es una muy rara malformación con alta morbimortalidad. Hasta 2016 se habían reportado 36 casos en el mundo.

Caso clínico. Paciente masculino de 11 años de edad, antecedentes de neumopatía crónica más insuficiencia respiratoria, bilioptisis, saturación arterial entre 66-80%, ictericia, tórax asimétrico, dedos hipocráticos, estertores crepitantes diseminados. Diagnóstico positivo por fibrobroncoscopia y tomografia axial computarizada. Tras cierre de fístula y neumonectomía derecha, se produjo recidiva por hipoplasia de la vía biliar comprobada con colangiopancreatografía retrógrada endoscópica. Se realizó segmentectomía hepática lateral izquierda y cierre de fístula desde el abdomen. Mantuvo fístula broncopleural persistente, luego de tratamiento nutricional y antibiótico intensivo, se cerró quirúrgicamente con parche de pericardio bovino. Asintomático tras 6 meses.

Comentarios. La muy baja frecuencia de esta malformación obliga a un alto índice de sospecha y un tratamiento multidisciplinario. Es importante considerar la presencia de hipoplasia de la vía biliar pues obliga a un abordaje toracoabdominal.

Palabras Clave: Fístula; Bronquio; Enfermedad del tracto biliar.

INTRODUCTION

In 1850, Peacock's described the first case of bronchobiliary fistula (BBF) in a 20-year-old woman with hepatic echinococcosis[(1)]. BBF has also been called biliary bronchial fistula, hepatobronchial fistula, pleurobiliary fistula, and cholecystobronchial fistula. It can be congenital or acquired as a result of trauma, liver disease, or biliary obstruction, and it is characterized by an abnormal connection between the respiratory system (the trachea or the bronchi) and the biliary tract[(1-3)].

The presence of bile ptyalism, which can be mistaken for purulent expectoration, is pathognomonic, and it is typically associated with coughing and fever. In the chronic stage, bronchiectasis occurs in the pulmonary segments involved[(1-4)]. Episodes of recurrent pneumonia are frequent, associated with pleural effusion, especially on the right side. Owing to the chemical pneumonitis caused by the bile, bacterial infections and resulting sepsis occur[(1,2,5)].

Bronchoscopy is the most frequent diagnostic technique, followed by contrast CT-scan with 3D reconstruction (CT angiography)[(5)]. Surgery is the treatment of choice. Ligation is performed, with or without excision of the fistulous tract, and in cases of associated biliary hypoplasia, with resection of the liver tissue involved and biliodigestive diversion if required[(3,5-7)].

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Date of submission: June 2020 *Date of acceptance:* July 2020

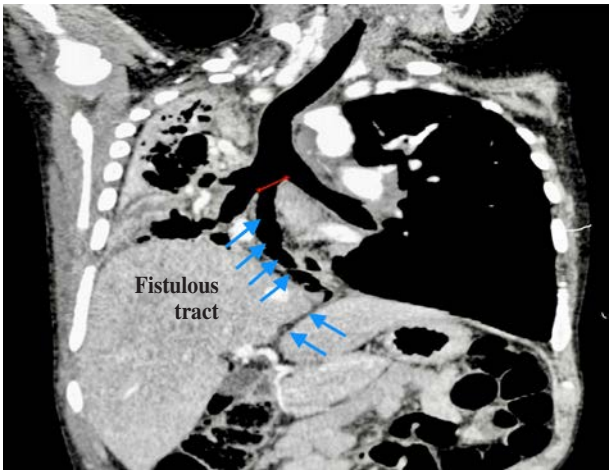

Figure 1. Thoracoabdominal CT-angiography. Bronchobiliary fistulous tract (blue) and occurrence (red) at the right bronchus close to the carina.

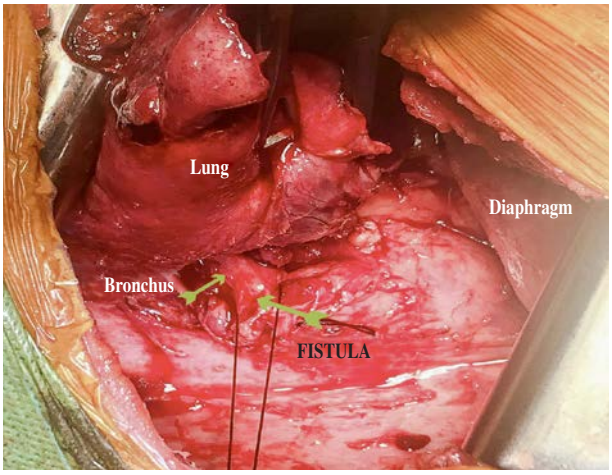

Figure 2. Intraoperative view of the thoracic cavity. Communication of the thick fistula ascending from the diaphragm to the right bronchus.

It is worth noting that BBF is a rare malformation with high morbidity and mortality rates if not timely diagnosed. 36 cases have been reported worldwide since it was first described in 1952 by Neuhauser et al.[4,5].

CLINICAL CASE

11-year-old male patient, with history of chronic lung disease and respiratory insufficiency requiring hospitalization several times at other Ecuadorian healthcare facilities. He presented night coughing, bile expectoration, and fever. Arterial oxygen saturation ranged from 66% to 80%. At physical exploration, he had overall paleness, slight jaundice staining, asymmetric thorax, decreased vesicular murmur, and disseminated crackling rales, primarily on the right side. The abdomen was distended and globular, and there was finger clubbing in the limbs. Laboratory tests showed anemia, leukocytosis, thrombocytosis, and mixed acidosis with high lactate levels.

Thoracoabdominal CT-angiography demonstrated right pulmonary fibrosis, bronchiectasis, and a fistulous tract from the left bile duct to the right main bronchus (Fig. 1). Flexible fibrobronchoscopy showed chronic tracheobronchitis, with an abundant greenish secretion suggestive of bile content from the fistula orifice, thus confirming BBF diagnosis.

As a result of severe and irreversible chronic pulmonary damage, right pneumonectomy was decided upon. An atrophic and fibrotic lung, a 10mm bronchobiliary fistula communicating with the right main bronchus, and multiple adhesions to the diaphragm and the parietal pleura were found (Fig. 2). Communication was divided, and they were both closed using non-absorbable running sutures. The pathological study revealed dilated bronchoalveolar structures, fistulous areas, and bile-looking yellow-pigmented material, with the lung parenchyma being replaced by fibrohyaline tissue.

On postoperative day 3, a yellowish liquid was observed. Total bilirubin levels at cytochemical study were 20.3 mg. A somatostatin analogue was applied for 15 days, without success. Given the suspicion of bile duct hypoplasia, and in order to facilitate drainage to the duodenum and probably reduce BBF output, combined endoscopic retrograde cholangiopancreatography (ERCP) and sphincterectomy were carried out (Fig. 3A), with no clinical improvement. The intra-abdominal fistulous tract involving liver segments II and III was resected and closed underneath the diaphragm (Fig. 3B).

The corrosive action of the bile on the bronchial suture caused dehiscence and thus bronchopleural fistula, which was treated with water seal chest drainage, aggressive nutritional support, long-term broad spectrum antibiotic therapy, and antifungals. As a result of this, clinical signs improved, and 21 days later, the short and friable bronchial stump was closed with a bovine pericardial patch and a synthetic polymer sealant (Fig. 4). Six months later, the patient had no symptoms or fistula recurrence.

DISCUSSION

Congenital BBF is a rare abnormality characterized by an abnormal connection between the respiratory tree and the left hepatic bile duct. In most cases, the fistula originates at the right main bronchus or the carina[3,5,7]. The fistulous tract descends through the posterior mediastinum and crosses the diaphragm through the esophageal hiatus. The decreased abdominal portion of the fistula leans towards the right, typically ending at the left bile duct. Exceptionally, it flows into the common hepatic duct[5].

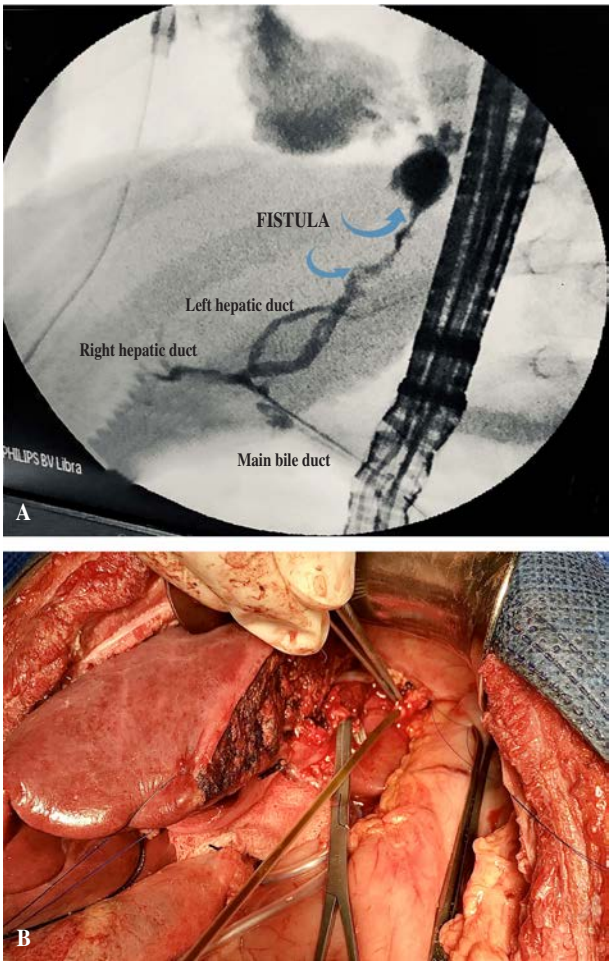

Figure 3. A) Endoscopic retrograde cholangiopancreatography. Fistula tract from the beginning of the left hepatic duct to the thorax. B) Surgical picture of the resection of liver segments II and III. Fistula cannulation, with the probe draining at the thorax through the diaphragm.

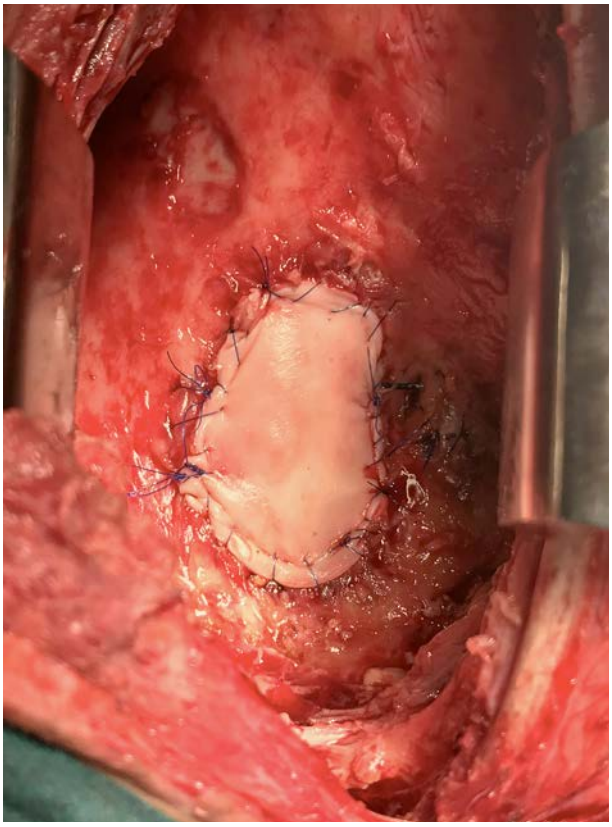

Figure 4. Short and friable bronchial stump closure with a bovine pericardial patch.

Pathogenesis is unclear. However, two potential embryological mechanisms have been proposed: (a) duplication of the upper gastrointestinal tract; and (b) merger of an abnormal bronchial bud with an abnormal bile duct. These hypotheses are based on histological evaluations of the fistulous tracts resected[3-5].

The most frequent symptoms are respiratory – coughing, dyspnea, cyanosis, bilious sputum, persistent rhinitis, pneumonia, atelectasis, and emphysema. However, these symptoms are unspecific, which means differential diagnosis with tracheoesophageal fistula, gastroesophageal reflux, gastrointestinal obstruction, gastrointestinal fistula, and aspirating pulmonology should be carried out[5,6].

Bronchoscopy stands as the most common and effective diagnostic technique. Other diagnostic techniques include bronchography, cholangiography, CT-scan 3D reconstruction, and magnetic resonance cholangiography, which provide functional information[3,5,8].

In acquired fistulas, especially those under 3 mm in size, conservative treatment is initially attempted, with endoscopic occlusion since the therapeutic bronchoscopy has been carried out, and biliary drainage measures such as ERCP, associated or non-associated with somatostatin analogues. However, in congenital fistulas, especially those with associated abnormalities of the main bile duct, surgery is the treatment of choice – either pulmonary resection, diaphragmatic fistula defect closure, liver resection, or bilioenteric anastomosis[1-3,5, 9-10]. Fistulous tract resection is preferred to ligation[3,5]. In half of the cases reported, concomitant malformations have been described, 66% of which are congenital abnormalities of the biliary system (hypoplasia or atresia), which makes surgical treatment more complex and risky[3,5,7].

This rare congenital disease is to be considered in newborns with respiratory distress and airway bile output without intestinal obstruction, since late diagnosis leads to irreversible lung parenchyma damage[6,8].

Cross-disciplinary management is key to effective diagnosis and treatment. When BBF is suspected, bronchoscopy has demonstrated to be the diagnostic technique of choice. The presence of bile duct hypoplasia should be ruled out, since it requires a thoracoabdominal approach in one or multiple surgical maneuvers.

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