Bronchoesophageal fistula due to broncholithiasis: a case series

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Received 28 October 2004

Summary

Objective: To describe the clinical presentation, diagnostic evaluation, and treatment of bronchoesophageal (BE) fistula due to broncholithiasis, a rare cause of chronic cough with specific therapeutic implications.

Methods: Retrospective chart review of 9 patients diagnosed with BE fistulae at our tertiary-care institution between 1964 and 2002.

Results: The median age of patients (3 men, 6 women) was 56 years (range, 34–72 years). Six patients had never smoked. Eight presented with intractable cough typically worse after drinking that was relieved by lying on either side; 1 patient presented with dysphagia. Six patients reported lithoptysis, and most had a history of recurrent pneumonia. Chest findings were nonspecific. Diagnosis was established by radiocontrast studies of the esophagus (5 patients), surgical procedures (3), or bronchoscopy (1). Fistulae were right-sided in 7 patients and left-sided in 2. All patients had surgical repair of the fistulae; 6 patients experienced symptom resolution.

Conclusion: BE fistula caused by broncholithiasis most commonly affects the right bronchial tree and should be considered in patients with chronic cough associated with drinking, lithoptysis, or recurrent pneumonia. The diagnosis is usually established by radiocontrast studies of the esophagus or incidentally during operations. Surgical repair is required.

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Introduction

Chronic cough is a common presenting complaint of patients in general medical practice. The most common causes of chronic cough include postnasal drip syndrome, asthma, gastroesophageal reflux disease (GERD), chronic bronchitis from cigarette smoking or exposure to chemical and environmental irritants, bronchiectasis, eosinophilic bronchitis, and use of angiotensin-converting enzyme inhibitors. These conditions account for about 95% of all cases of chronic cough.¹ The other 5% of
persons affected have less common conditions, such as bronchogenic carcinoma, tuberculosis, or pharyngeal dysfunction.2

Bronchoesophageal (BE) fistula caused by broncholithiasis is a rare condition that also may result in chronic cough. Broncholithiasis is defined as the presence of calcified material within a bronchus or within a cavity communicating with a bronchus.3 A number of case reports and case series describing patients with broncholithiasis have been published since the mid-1940s.3–5 However, broncholithiasis with secondary BE fistulization is extremely rare and has been described in only a few case reports.6,7 Thus, we sought to describe the clinical presentation, diagnostic evaluation, and treatment of patients with chronic cough caused by BE fistula secondary to broncholithiasis.

Methods
A computerized search of the medical records of all patients seen at Mayo Clinic in Rochester, Minnesota, between 1950 and 2002 was conducted using the terms “broncholithiasis,” “bronchoesophageal fistula,” and “tracheoesophageal fistula.” The search for broncholithiasis, which included codes for “calculus bronchus” and “calcification bronchus,” yielded 706 cases. Of these 706 patients with a diagnosis of broncholithiasis, 15 were identified who also had BE fistulae. Patients selected for this study were identified on the basis of (1) surgical, radiologic, or endoscopic evidence of broncholithiasis and a BE fistula and (2) a documented causal relationship between broncholithiasis and BE fistulization. Nine patients met these inclusion criteria.

Medical records were reviewed to identify the clinical characteristics of each case. In compliance with federal regulations, permission to perform this retrospective review was granted by the Mayo Foundation Institutional Review Board. Specific information that was extracted included presenting symptoms, examination findings, investigations that yielded the diagnosis, complications of therapy, and overall clinical outcome.

Results
The characteristics of the study population are outlined in Table 1. The median age of the 9 patients (3 men, 6 women) was 56 years (range, 34–72 years). Six patients (67%) had never been smokers, and 6 had other conditions known to
cause chronic cough, including GERD, asthma, hiatal hernia, and bronchiolitis obliterans with organizing pneumonia.

Eight patients (89%) presented with cough that was characteristically intractable, severe, worse after swallowing liquids, and relieved by lying on either side. One patient (patient 2) had dysphagia as the presenting symptom. Six patients (67%) reported expectoration of foreign bodies (lithoptysis). However, these patients usually did not volunteer a history of lithoptysis but rather affirmed it on specific questioning. Findings on physical examination were nonspecific and highly variable. Abnormal auscultatory findings did not correlate with the location of the pathologic condition.

Four patients (44%) were treated empirically for asthma before receiving a diagnosis of BE fistula caused by broncholithiasis. Six patients (67%) had a history of recurrent pneumonia. The time from the onset of symptoms to the diagnosis of a BE fistula was 1–72 months (mean, 21 months).

The diagnostic and therapeutic features and outcomes of the 9 patients are summarized in Table 2. A definitive diagnosis was established in most patients by multiple studies, including chest radiography (9 patients), radiocontrast studies of the esophagus (7 patients), computed tomography (CT) of the chest (5 patients), bronchoscopy (5 patients), esophagogastrodudenoscopy (EGD) (5 patients), and pulmonary function testing (3 patients). Of these tests, only the radiocontrast studies of the esophagus and bronchoscopy were diagnostic of BE fistula caused by broncholithiasis. Notably, 7 patients had abnormal chest radiographs, and the most common findings were calcified granulomas or calcified hilar or mediastinal lymph nodes. The most common abnormal chest CT findings were calcified granulomas in the lungs, liver, or spleen, calcified lymph nodes, and pulmonary infiltrates. Findings on pulmonary function testing varied widely from normal to evidence of reversible obstruction in patients diagnosed with asthma.

The diagnosis of BE fistula caused by broncholithiasis was established by radiocontrast studies of the esophagus in 5 patients (56%), during surgical procedures in 3 (33%), and by bronchoscopy in 1 (11%). Figure 1 shows the bronchoscopic view of a broncholith that had eroded into the right main stem bronchus of a patient with cough and lithoptysis.

Seven patients (all except patients 2 and 5) had radiocontrast studies of the esophagus. For 5 of the 7 patients, these studies established the diagnosis of BE fistula. For the other 2 patients, the diagnosis was established by bronchoscopy in 1 patient (patient 8) and during a surgical procedure in the other (patient 4).

Eight patients (all except patient 2) underwent bronchoscopy. Of these 8, only 4 patients had the presence of a BE fistula confirmed. All 5 patients in whom radiocontrast studies of the esophagus identified a BE fistula caused by broncholithiasis subsequently underwent bronchoscopy. In these patients, bronchoscopy confirmed a BE fistula in only 3 (patients 1, 6, and 9). Indeed, bronchoscopy was the diagnostic test in only 1 patient (patient 8).

The diagnosis of BE fistula caused by broncholithiasis was established during surgical procedures in 3 patients. Patient 2 was referred to our institution for evaluation of dysphagia after a mass, presumed to be an esophageal leiomyoma, was identified on a radiocontrast study of the esophagus. On EGD, the mass was found to be a submucosal lesion. During a surgical procedure, the mass was identified as a large group of lymph nodes with broncholithiasis and an associated BE fistula. Patient 4 was referred to our institution for an intractable cough made worse by drinking. CT of the chest revealed large calcified subcarinal and left hilar lymph nodes. No fistula was found on EGD, a radiocontrast study of the esophagus, or bronchoscopy. The patient was taken to the operating room for treatment of symptomatic broncholithiasis, and the BE fistula was found during that procedure. Patient 5 was referred to our institution for recurrent pneumonia and an intractable cough made worse by drinking. CT of the chest revealed calcified hilar and mediastinal lymph nodes. Bronchoscopy showed a large broncholith involving the right main stem bronchus. No fistula was found. The patient was taken to the operating room for treatment of symptomatic broncholithiasis, and the BE fistula was found during that procedure.

BE fistulae were located at the right main stem bronchus in 3 patients (33%), the bronchus intermedius in 4 patients (44%), and the left main stem bronchus in 2 patients (22%). Overall, 7 patients (77%) had right-sided fistulae and 2 patients (22%) had left-sided fistulae.

All 9 patients in this series ultimately had surgical interventions to repair the fistulae. After these interventions, however, some patients experienced complications (e.g., wound infection, respiratory failure, and atrial fibrillation). Of the 8 patients who presented with cough, 6 had complete resolution of the cough after surgery. Patient 2, who presented with dysphagia, continued to experience persistent mild dysphagia. Two patients experienced a late recurrence of fistulae after their initial
Table 2  Diagnosis, therapy, and outcome of 9 patients with bronchoesophageal fistula caused by broncholithiasis.

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Time to diagnosis (month)</th>
<th>Diagnostic test</th>
<th>Fistula location</th>
<th>Treatment</th>
<th>Outcome or complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12</td>
<td>Radiocontrast study of the esophagus</td>
<td>Bronchus intermedius</td>
<td>Repair of the fistula, broncholithectomy</td>
<td>Unresolved symptoms or no complications</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td>Operation</td>
<td>Left main stem</td>
<td>Repair of the fistula, broncholithectomy</td>
<td>Persistent mild dysphagia or no complications</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>Radiocontrast study of the esophagus</td>
<td>Right main stem</td>
<td>Repair of the fistula, broncholithectomy, repeat repair of fistula</td>
<td>Resolved or no complications</td>
</tr>
<tr>
<td>4</td>
<td>6</td>
<td>Operation</td>
<td>Bronchus intermedius</td>
<td>Repair of the fistula, broncholithectomy, bronchoplasty</td>
<td>Resolved or no complications</td>
</tr>
<tr>
<td>5</td>
<td>20</td>
<td>Operation</td>
<td>Right main stem</td>
<td>Repair of the fistula, broncholithectomy, sleeve resection</td>
<td>Resolved or no complications</td>
</tr>
<tr>
<td>6</td>
<td>60</td>
<td>Radiocontrast study of the esophagus</td>
<td>Bronchus intermedius</td>
<td>Repair of the fistula, broncholithectomy</td>
<td>Resolved or wound infection</td>
</tr>
<tr>
<td>7</td>
<td>11</td>
<td>Radiocontrast study of the esophagus</td>
<td>Left main stem</td>
<td>Mediastinal lymphadenectomy, repair of the fistula, broncholithectomy</td>
<td>Resolved or no complications</td>
</tr>
<tr>
<td>8</td>
<td>6</td>
<td>Bronchoscopy</td>
<td>Bronchus intermedius</td>
<td>Surgical closure, serratus anterior flap, and thoracic lymphadenectomy</td>
<td>Persistent cough or respiratory failure, postoperative anemia and fever, repeat bronchoscopy for retained secretions, jejunostomy, FT, tracheostomy, wound infection, AF with RVR</td>
</tr>
<tr>
<td>9</td>
<td>72</td>
<td>Radiocontrast study of the esophagus</td>
<td>Right main stem</td>
<td>Endoscopic coagulation with suturing, fistula cautery with fibrin glue, surgical closure with muscle flap to esophagus</td>
<td>Resolved or no complications</td>
</tr>
</tbody>
</table>

AF, atrial fibrillation; FT, feeding tube; and RVR, rapid ventricular response.

operations (patient 1 after 2 years and patient 3 after 5 years). These recurrent fistulae were repaired successfully and symptoms resolved. There were no operative or perioperative deaths.

Discussion

Broncholithiasis is a condition caused by the healing of granulomatous pulmonary infections that spread from the lung parenchyma to peribronchial and hilar lymph nodes and then calcify. Constant respiratory motion causes simultaneous pressure atrophy of the associated bronchial wall. This process may result in erosion and migration of the lymph nodes into the bronchus. The “spitting of stones” associated with this condition has been described for centuries. Some of the earliest descriptions of lithotomy were provided by Aretaeus, Galen, and Aristotle. The most common symptoms of broncholithiasis are cough,
lithoptysis, hemoptysis, and pneumonia. Erosion of broncholiths into the esophagus results in BE fistula.

BE fistulae are uncommon; those caused by broncholithiasis are extremely rare. Indeed, our search yielded only 9 cases from more than 700 patients with broncholithiasis treated in a period of more than 50 years at our institution. Cough associated with drinking liquids or with migratory pneumonia is particularly suggestive of BE fistula, and broncholithiasis should be suspected in patients with aerodigestive symptoms and calcifications in the hilum or mediastinum on chest radiographs. Eight patients had a characteristically intractable, severe cough that was worse after swallowing liquids and that was relieved by lying on either side. Furthermore, 6 patients reported lithoptysis. However, patients usually did not volunteer a history of lithoptysis. Hence, patients with chronic intractable cough should be asked about possible lithoptysis.

The median age of our patients was 56 years. The occurrence of broncholithiasis in the sixth decade is consistent with data from other series. Patients were predominantly from the midwestern United States, but the population is perhaps skewed because our facility is located in a region of the country known as the “histoplasmosis belt” with easy access by regional patients.

Infectious agents most commonly associated with broncholithiasis are *Mycobacterium tuberculosis* and *Histoplasma capsulatum*. Other causes include aspiration of bone tissue or in situ calcification of aspirated foreign material and erosion by and extrusion of calcified or ossified bronchial cartilage plates, silicosis, and rarely actinomycosis. In these 9 patients, we were unable to ascertain clear pathogenic agents of broncholithiasis. The most common implicated agent was histoplasmosis. One patient had a remote history of ocular histoplasmosis and 3 others had a history of histoplasmosis. Such inability to identify the causative organism in broncholithiasis has been reported by other authors. In a study by Vollmer and Flye, the causative organism was infrequently identified in surgically resected specimens. However, Potaris and colleagues obtained positive cultures of *H. capsulatum* in 12 of 47 patients who had broncholithiasis, and 1 patient had both *H. capsulatum* and *Mycobacterium avium-intracellulare*.

In our series, findings of clinical examinations were highly varied and nonspecific. To establish a definitive diagnosis, multiple investigations were conducted in most patients. Such investigations included chest radiography, radiocontrast studies of the esophagus, chest CT, pulmonary function testing, bronchoscopy, and EGD. Calcified granulomas or lymph nodes were commonly observed on radiographs or CTs of the chest, but BE fistulae are unlikely to be demonstrated with such techniques. Although these tests may provide useful data, priority should be given to radiocontrast studies of the esophagus and perhaps to bronchoscopy because they provide the highest diagnostic yield.

In our series, BE fistulae were located on the right side in 7 of 9 patients (77%). This right-sided preponderance of broncholiths is consistent with data reported by Head and Moen in 1949, Arrigoni et al. in 1971, Faber et al. in 1975, and Potaris et al. in 2000. In the review by Potaris et al. of 47 patients with broncholithiasis who had surgical treatment, 64% of patients had abnormal bronchoscopic findings on the right side, 19% had left-sided findings, and 17% had abnormalities on the carina. We postulate that the high prevalence of fistulae in the right main stem bronchus occurs because the lymph node, bronchus, and esophagus commonly converge at this site. Although the esophagus is also associated with the left main stem bronchus, this area rarely contains as many lymph nodes.

Ultimately, all 9 of our patients had surgical repair of their fistulae. BE fistula caused by broncholithiasis is a well-established indication for surgery. Other general indications for operative management are hemoptysis, suppurative lung disease, bronchiectasias or bronchial stenosis, aortotracheal fistula, possible malignancy, uncertain diagnosis, and failure of bronchoscopy.

Surgical treatment of a BE fistula includes repair of the fistula, broncholithectomy with or without bronchoplasty, and pulmonary resection.
47 patients, Potaris and colleagues\(^3\) reported intraoperative complications in 12.8% of patients. These complications included lacerations of the pulmonary artery, esophagus, or bronchus and pulmonary infarction caused by pulmonary artery thrombosis. Potaris et al.\(^3\) reported postoperative complications in 34% of patients. These complications (7 major, 9 minor) included hemothorax, empyema, wound dehiscence, pulmonary artery thrombosis, respiratory insufficiency, pneumothorax, and atrial fibrillation. The reported rate of complications is similar to that observed in our small series.

Although surgery is considered the standard therapy for BE fistulae, endoscopic techniques were attempted twice in one of our patients (patient 9) before surgical repair provided symptomatic relief. The published data on endotherapy of BE fistulae are limited. Case reports show varying results. One case report described endoscopic suturing and coagulation of an esophageal fistula.\(^1\) Stenting of malignant esophageal fistulae has also been accomplished.\(^1\)\(^2\)

Limitations of our case series include its small size, which makes it difficult to extrapolate these findings to the general population. However, BE fistula caused by broncholithiasis is such a rare condition that large sample sizes would be difficult to obtain. Indeed, we were able to identify only 9 cases in the period between 1950 and 2002. Another possible limitation is that patients presented to our tertiary-care facility after complications developed. Hence, these patients may have been more ill than those in the general population with BE fistulae who could be asymptomatic or experiencing relatively mild symptoms.

BE fistula caused by broncholithiasis is a rare but clinically significant cause of chronic cough. A history of lithotripsy is an important clue to diagnosis. Although examination findings are nonspecific and vary highly, mediastinal or hilar calcifications are often present. In our series, the investigations that had the best diagnostic yield were radiocontrast studies of the esophagus that revealed fistulae, often on the right side. Surgical intervention is required for this condition, and endoscopic techniques have been used with limited success. Even after an operation, symptoms may persist and postoperative complications may occur.

### Acknowledgments

We wish to thank Dr. Paul R. Daniels and Dr. Mark Liebow from the Division of General Internal Medicine at Mayo Clinic for their guidance in the preparation of this case series. We also thank Dr. James P. Utz from the Division of Pulmonary and Critical Care Medicine at Mayo Clinic for providing us with bronchoscopy photographs. We are grateful as well to Dr. Christopher J. Gostout of the Division of Gastroenterology and Hepatology at Mayo Clinic for providing information on endotherapy of esophageal fistulae.

### References