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CASE REPORT | ESOPHAGUS

A Rare Cause of Dysphagia and Cough: Bronchoesophageal Fistula from Silicosis

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ABSTRACT

Dysphagia and cough in an older male smoker raise concern for malignancy. However, a history of environmental exposures led to a much more interesting diagnosis in this case of pneumoconiosis due to silicosis. Silicosis is an uncommon pulmonary disease with rare associated gastrointestinal symptoms. We report a bronchoesophageal fistula resulting from silicosis causing dysphagia and cough. This is the first report of using endoscopic stenting to manage an esophageal fistula from silicosis. This case highlights how common symptoms of cough and dysphagia can masquerade as a pulmonary or oropharyngeal problem, when they are actually gastrointestinal manifestations of a rare disease.

INTRODUCTION

Inhalation and deposition of silica particles into the terminal bronchioles and alveoli can ignite an inflammatory cascade that causes local injury, ultimately leading to fibrosis and calcification. In silicosis, severe pulmonary fibrosis can lead to massive calcification of peribronchial lymph nodes and structures adjacent to the esophagus. Over time, repetitive motions from respiration, swallowing, and normal circulation lead to broncholithiasis eroding into nearby structures, giving rise to a bronchoesophageal fistula. While infectious causes of broncholithiasis are well described, including fungal, nocardial, and mycobacterial lymphadenitis, silicosis is an extremely rare cause of bronchoesophageal fistula. Gastrointestinal (GI) manifestations of silicosis are extremely unusual and poorly described in the literature.[1-3]

CASE REPORT

A 73-year-old man with several weeks of progressive dysphagia, cough, and hoarseness presented with difficulty tolerating oral intake, with coughing immediately after ingestion of solids and liquids. He denied hematemesis, hemoptysis, or weight loss. He had several hospitalizations for recurrent pneumonia, requiring intermittent antibiotics, inhalers, and steroids. He also had a large empyema requiring video-assisted thoracoscopic surgery several months prior to presentation. He was a former 10 pack-per-year smoker who quit 20 years prior; he had worked as a dental technician with significant exposure to silica. He served in the Navy and was stationed in Cuba for several years, where he worked in the shipyards. His medical history included obstructive lung disease requiring home oxygen, bronchiectasis, gastroesophageal reflux, and silicosis.

On physical exam, the patient was afebrile and in no acute respiratory distress. His vital signs were blood pressure 108/66 mm Hg, heart rate 107 beats per minute, respiratory rate 20 breaths per minute, with oxygen saturation 95% on 3 L nasal cannula. He was a slender, well-appearing male, with no oropharyngeal findings, halitosis, tracheal deviation, or elevated jugular venous pressure. He had a notably hoarse, soft-spoken voice, and rales were

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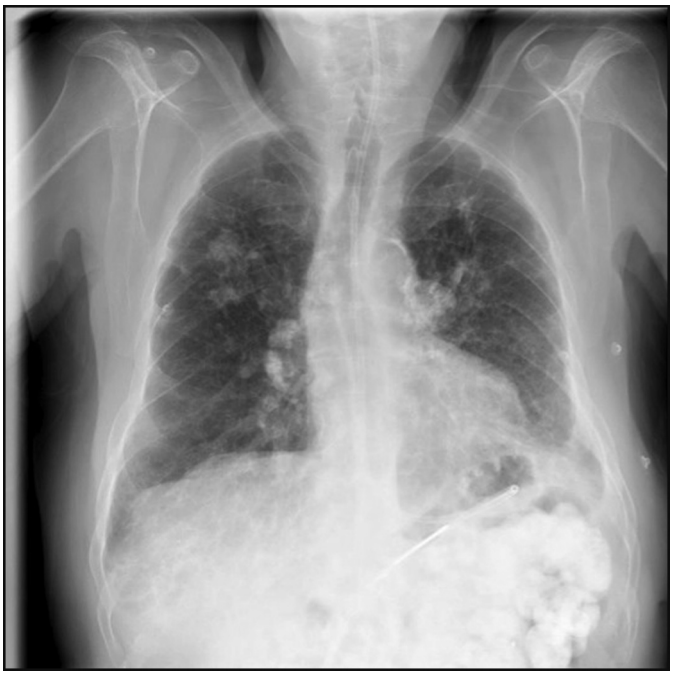

Figure 1. Chest radiograph demonstrating bilateral opacities in both mid to upper lung fields consistent with changes from silicosis. Calcified hilar lymph nodes, consistent with granulomatous disease from silicosis, are also seen bilaterally. Note a nasogastric tube is also in place.

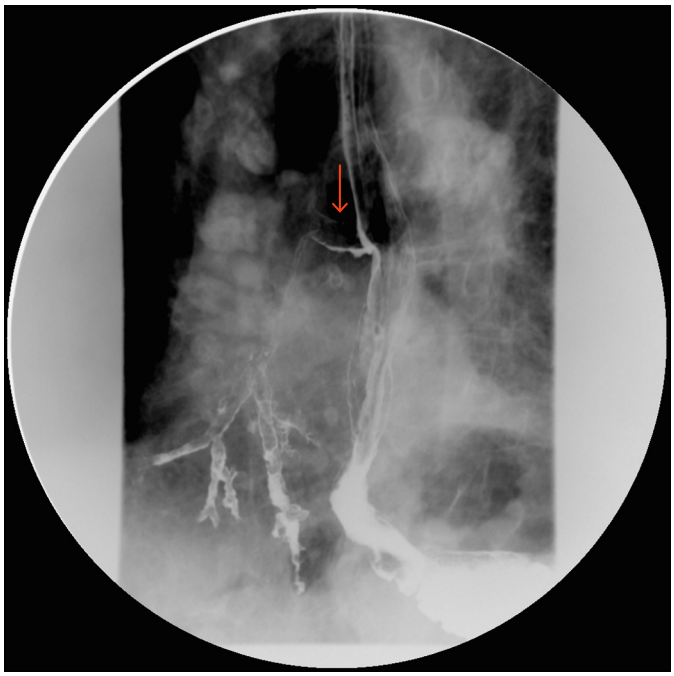

Figure 2. Upper gastrointestinal series demonstrating a widely patent fistula between the mid-esophagus and bronchus intermedius (arrow), with contrast extending into the right lower-lobe bronchi (left bottom) as well as the esophagus and stomach (right bottom). Mediastinal calcifications consistent with silicosis are also seen.

auscultated throughout both lung fields. Abdominal exam was unremarkable with no abdominal distension, tenderness on palpation, masses, or hepatosplenomegaly. The rest of the physical exam and labs were unremarkable. Tuberculosis and human immunodeficiency virus tests were also negative.

A swallowing evaluation was performed initially with a modified barium-swallow test, which showed moderate oropharyngeal dysphagia to solids. Aspiration occurred after the completed swallow, triggering cough and reflux of materials into the oral cavity. On chest radiograph, there were bilateral calcified hilar lymph nodes and opacities in the mid to upper lung fields consistent with his diagnosis of silicosis (Figure 1). Esophagogastroduodenoscopy revealed no esophageal abnormalities or a fistulous tract. A formal barium upper GI swallow study demonstrated a widely patent fistula between the mid-esophagus and bronchus intermedius, with contrast extending into the right lower-lobe bronchi (Figure 2). Mediastinal calcifications were also seen, consistent with silicosis. Chest computed tomography confirmed an 8-mm fistula extending from the subcarinal region into the esophagus, with progressive massive fibrosis and traction bronchiectasis in the upper lobes (Figure 3).

Esophageal stenting of the fistula was performed via endoscopy but was complicated by stent migration and eventual removal. An open partial esophagectomy was performed as definitive management. A month after surgery, the patient was tolerating small amounts of food by mouth without dysphagia or cough.

DISCUSSION

GI complications of silicosis are extremely uncommon and are rarely described in the literature. Despite recurrent episodes of aspiration pneumonia and prior negative computed tomography scans and endoscopy, a bronchoesophageal fistula went undetected in this patient until his symptoms progressed. This rare complication from silicosis only became obvious on imaging once his disease progressed. Vague symptoms of cough and dysphagia in a former smoker with a history of pneumoconiosis could easily be mistaken for a primary pulmonary or oropharyngeal problem. However, the key to this clinical diagnosis was the realization that a progressive and erosive granulomatous disease could lead to unexpected GI complications despite a negative initial workup.

As demonstrated in this case, broncholithiasis is complicated by calcification of surrounding structures, including lymph nodes. Over time, these calcified peribronchial structures erode into nearby structures, including the esophagus, through the repetitive motions of swallowing, respiration, and normal circulation.[1] The most common causes of calcification include infections (fungus, nocardia, mycobacterium) and granulomatous diseases.[1,4,5] Bronchoesophageal fistulas are

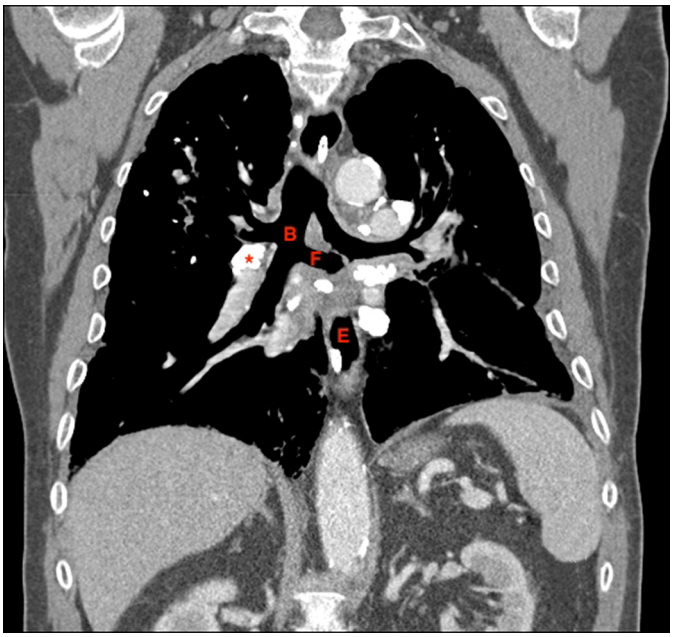

Figure 3. Coronal segment of a contrast-enhanced chest CT scan revealing an open bronchoesophageal fistula (8-mm wide) (F) at the subcarinal level involving the right bronchus intermedius (B) and extending into the esophagus (E) with retained contrast visualized. Extensive enhancing calcified lymph nodes (asterisk) are seen bilaterally throughout the anterior and middle mediastinum and hilar regions.

complicated by aspiration pneumonitis, pneumonia, lung abscess, and empyema, which complicated this patient's clinical presentation. Given the irreversible process of calcification and fibrosis in silicosis, broncholithiasis and associated fistulas are definitively managed with thoracic surgery, which can be associated with significant morbidity and mortality. However, esophageal fistulas themselves can be managed with less invasive strategies including endoscopic suturing and stenting.

Silicosis is a rare disorder with 3,000–7,000 cases reported annually between 1987 and 1996 in the United States.[6] The Department of Labor's Occupational Safety and Health Administration estimates that 2.2 million U.S. workers are exposed to silica.[7] Silica not only causes structural damage via an inflammatory process, but also is thought to be a carcinogen and a risk factor for lung cancer.[8] Roughly 1,437 individuals died from complications related to silicosis from 2001 to 2010.[9] Traditional occupational exposures to silica dust include mining, sandblasting, road construction, stone masonry, and pottery making. More recent hazardous exposures include dentistry, hydraulic fracturing of gas and oil wells, as well as fabrication and installation of engineered stone countertops. This patient's occupation as a dental technician involved exposure to respirable crystalline silica while mixing powders, removing castings from molds, and performing grinding and polishing maneuvers. The Centers for Disease Control and Prevention reported only 9 cases of silicosis in dental laboratory technicians from 1994 to 2000.[10]

Cough and dysphagia are common symptoms in older patients with a history of smoking. This patient, who had significant exposure to silica products as a dental technician and who also worked in a Navy shipyard, was at high risk for complications of pneumoconiosis, warranting further investigation of his dysphagia and chronic cough. Patients with significant environmental exposures and progressive symptoms require extensive evaluation and consideration of unique complications related to pneumoconiosis, including GI manifestations of rare pulmonary diseases.

DISCLOSURES

Author contributions: SR Lieber wrote the manuscript and is the article guarantor. S. Atlas and N. Ettenger edited the manuscript. N. Ettenger provided the images.

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