Recurrent Pneumonia in a 51-Year-Old Woman Due to Congenital Bronchoesophageal Fistula

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Introduction

Recurrent pneumonia in adults can be due to various etiologies. Congenital bronchoesophageal fistula may go undiagnosed in adult patients for many years. In cases of delayed recognition there is a long history of intermittent bouts of cough and choking after swallowing liquid, which may eventually present as recurrent pneumonia, lung abscess, or bronchiectasis. We report a patient with congenital bronchoesophageal fistula who presented in adulthood and discuss the pathogenesis, diagnosis, and management of this condition.

Case Summary

A 51-year-old woman with a 20-pack-year history of smoking presented with chronic productive cough and regurgitation for the past 5 years. She had a severe pneumonia 10 years before, which required prolonged intubation and tracheostomy, followed by 3 episodes of right middle and lower lobe pneumonia. Her medications included omeprazole for gastroesophageal reflux disease and inhaled corticosteroids and β agonists for presumed cough-variant asthma. Her pulmonary function test results were normal. She had a history of substantial exposure to second-hand smoke. On physical examination she was afebrile, had normal oxygen saturation on room air, and there was no evidence of respiratory distress. She had a well healed tracheostomy scar, and no peripheral lymphadenopathy. Clubbing was present in all digits. Auscultation of the right lower lung field revealed bronchial breath sounds and rhonchi.

Computed tomogram showed scarring of the right middle and lower lobes, and traction bronchiectasis (Fig. 1). A barium swallow study revealed a bronchoesophageal fistula in the distal esophagus, with contrast entering the right lower lobe bronchus (Fig. 2). Subsequent esophagoscopy and bronchoscopy found no luminal lesions or fistulas. She underwent right posterolateral thoracotomy for fistula repair with surgical stapling and pedicled vascularized buttress, and wedge resection of the right lower lobe. The fistulous tract measured 3.5 cm in length and 5–8 mm in width and could be felt as a rubbery cord leading to the esophagus, splitting the esophageal muscle. There was no evidence of inflammation or adherent lymph nodes around the fistula.

Pathology of the resected specimen revealed that the fistula was lined by benign squamous epithelium with the muscularis mucosa, and there was no evidence of malignancy, infection, or chronic inflammation. Acquired causes for bronchoesophageal fistula were ruled out by negative cultures and biopsies obtained from bronchoscopy and absence of other pathology on imaging studies. Her postoperative course was uneventful, and there was no clinical or radiographic evidence of fistula recurrence at 12-month follow-up.

Discussion

Fistulous communication between the esophagus and bronchus can result from congenital, inflammatory, traumatic, or neoplastic etiologies.[1] Congenital bronchoesophageal fistula with associated esophageal atresia usually presents in infancy, but is not highly unusual in adult patients, especially if there is no associated esophageal atresia and the involved bronchus is lobar rather than main.[1] Congenital bronchoesophageal fistula in adults is most commonly reported in the 40–60 years age group.[2]

There 4 types of congenital bronchoesophageal fistula:

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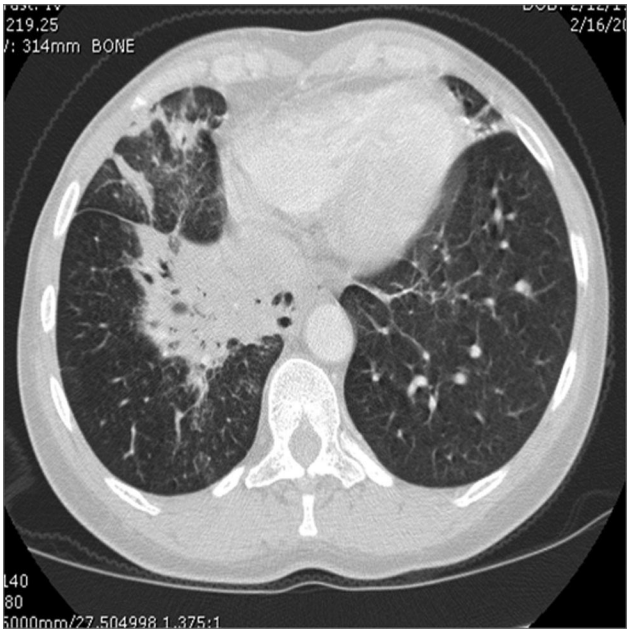

Fig. 1. Computed tomogram shows scarring of the right middle and lower lobes, and traction bronchiectasis.

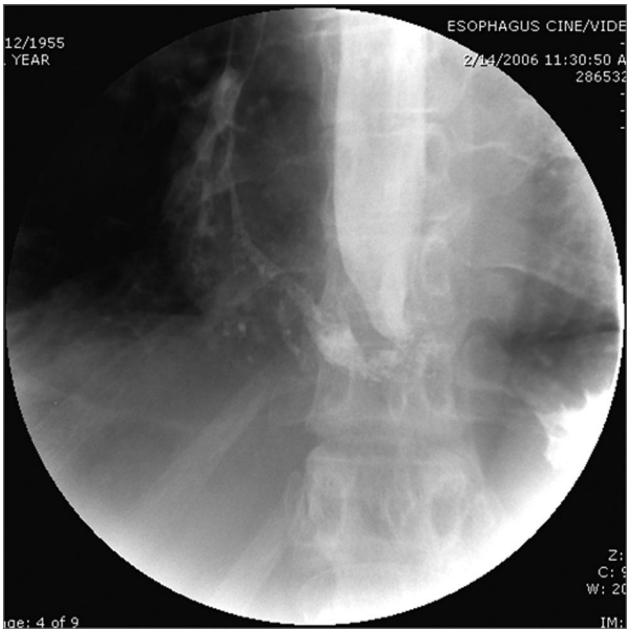

Fig. 2. Barium swallow shows bronchoesophageal fistula in the distal esophagus, with contrast entering the right lower lobe bronchus.

Type 1 is associated with a diverticulum of the esophagus, which may get inflamed due to stasis within the dependent tip of the diverticulum. The diverticular inflammation leads to its erosion and perforation into the bronchus. Although this type of fistula is congenital, its clinical presentation and manifestation are secondary to the underlying inflammatory changes.

Type 2 is the commonest and is due to a direct fistulous communication between the esophagus and the lobar/segmental bronchus.

Type 3 is due to a fistulous track between a cyst within the lung and the esophagus.

Type 4 is due to communication between a sequestrated lung segment (which has its own systemic arterial supply from the aorta) and the esophagus.

The existence of adult congenital bronchoesophageal fistula is controversial[3] and thought to be due to a persistent attachment between the tracheobronchial tree and the esophagus, and abnormal differential growth of the trachea and esophagus in the embryologic period. The eventual location of the fistula with respect to the bronchus depends on the degree of tracheoesophageal separation that occurs before the caudal migration of the trachea.[2] Fistulas are more common on the right side, mainly to the right lower lobe bronchus.[1] The absence of inflammation and adherent lymph nodes around the fistulous track and the histologic confirmation of mucosa and muscularis mucosa suggest a congenital etiology for the fistula.[1]

These patients are rarely symptomatic until late childhood or adulthood. Various explanations[1,2,4] for this lengthy asymptomatic interval have been proposed:

- Presence of a membrane overlying the fistula, which subsequently ruptures
- Presence of a proximal fold of esophageal mucosa overlapping the orifice, which subsequently becomes less mobile
- Anti-gravitational or upwards extension of the fistulous tract from the esophagus, which may close during swallowing
- Spasm of the smooth muscle of the fistula

Cough is the commonest symptom, followed by recurrent respiratory infections and pneumonia. A bout of coughing/choking on swallowing liquids (Ohno's sign) is pathognomonic,[2] but these symptoms are often mild and elicited in retrospect, after the diagnosis has been made.[1] Barium swallow is the diagnostic test of choice, although computed tomogram is often performed to look for complications such as bronchiectasis or associated pathology. Bronchoscopy and esophagoscopy sometimes find the fistula orifice, which is often small and recognizable only when the exact site is known.[1]

Early and definitive surgery, with meticulous exposure and division or resection of the fistula and primary repair of the bronchus and esophagus, is the treatment of choice. Resection of the affected segment or lobe should be performed if the fistula communicates with a lung cyst or is associated with bronchiectasis, to prevent pulmonary complications.[3,5,6]

Esophagoscopic and bronchoscopic occlusion of the fistula with a tissue sclerosant, such as silver nitrate or sodium hydroxide, can be attempted in patients who are poor surgical candidates.[7]

Teaching Points

Bronchoesophageal fistula in adults is usually associated with malignancy, infection, inflammation, or trauma. Bronchoesophageal fistula of benign or congenital etiology can present in adulthood. Clues to the diagnosis of congenital bronchoesophageal fistula include symptoms of aspiration, chronic wheeze, recurrent bronchopulmonary infections, hemoptysis, and hematemesis. The diagnosis is confirmed with barium esophagography and absence of other causes on clinical workup and imaging. Division of the fistula and reinforcement of the esophageal staple with a vascular pedicle graft, via thoracotomy or video-assisted thoracoscopic surgery, is curative. Lung resection is required if the fistula communicates with a lung cyst or is associated with bronchiectasis.

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