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Congenital bronchoesophageal fistula in an adult: a case report and radiologic review

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Congenital bronchoesophageal fistula (BEF) is rarely reported in adults in the English literature. Herein, we present a rare case of congenital BEF in a 43-year-old man that was incidentally found on esophagogastroduodenoscopy. Chest CT and barium esophagography revealed a fistula between his lower esophagus and the right lower lobe segmental bronchus. After the fistula was surgically treated, the fistula was no longer noted on follow-up barium esophagography.

Key Words: Bronchoesophageal fistula, Congenital, CT Esophagography,

Bronchoesophageal fistula (BEF) in adults is commonly caused by malignancy (1). Benign BEF is rare in adults and can be congenital or acquired. Acquired BEF is more common than congenital BEF and can be of traumatic, inflammatory, infectious, or iatrogenic origin. BEF may appear at any time during life.[1-3] Congenital BEF is usually associated with esophageal atresia and is readily diagnosed during the neonatal period. However, in cases where congenital BEF is not associated with esophageal atresia, it may persist into adulthood before becoming clinically apparent.[4,5] Congenital BEF is rarely reported in adults, and CT findings of adult BEF are rarely reported in the English literature. Therefore, we present a case of congenital BEF occurring in an adult with a radiologic review.

CASE

A 43-year-old man was referred to our clinic after an incidental finding of a fistulous opening at the lower third of the esophagus on esophagogastroduodenoscopy that was conducted during routine health screening (Fig. 1A). He had no significant medical or surgical history, but had

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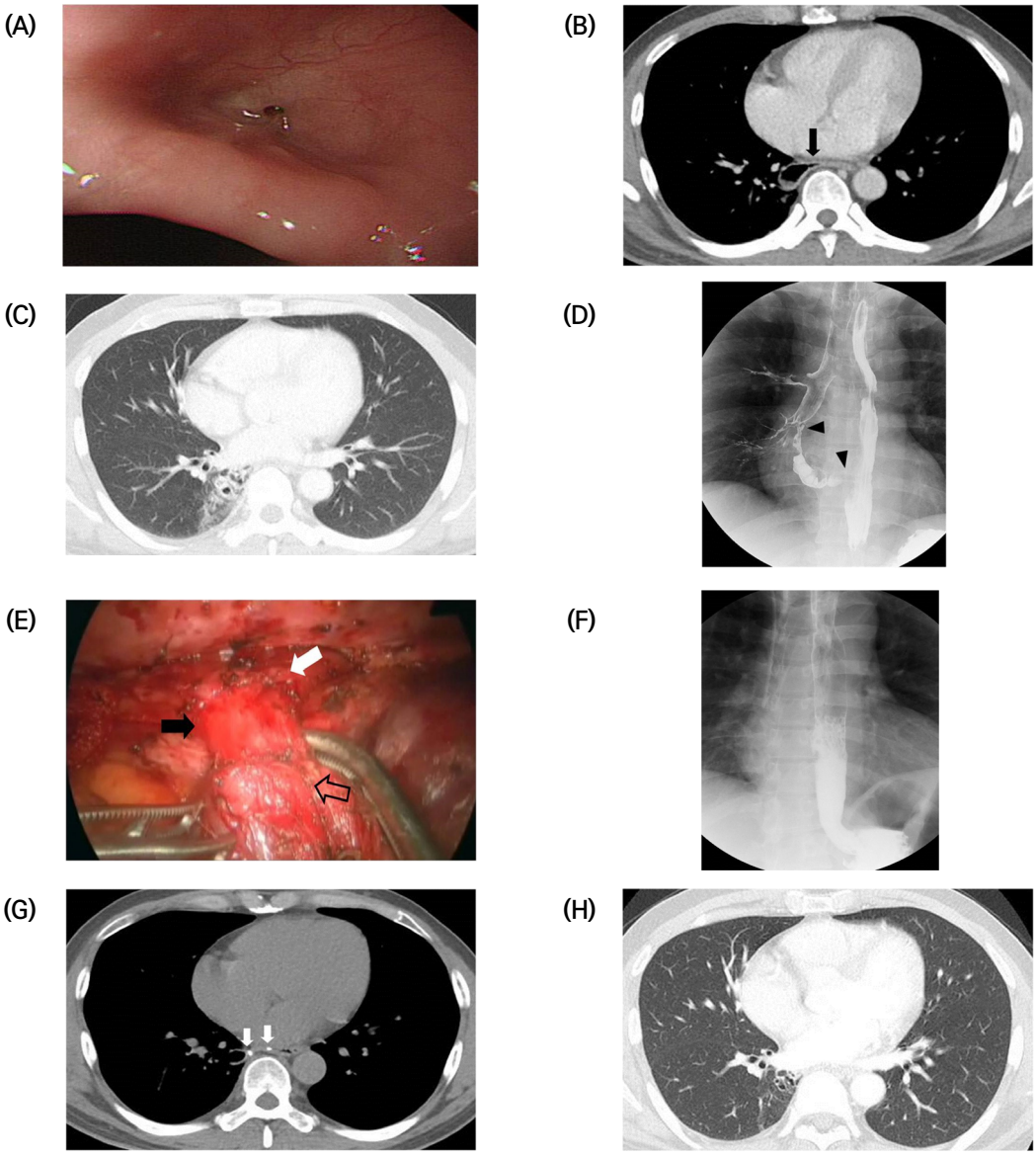

Fig. 1. A 43-year-old man with an incidentally detected fistulous opening at the lower third of esophagus on esophagogastroduodenoscopy.
(A) Esophagogastroduodenoscopy shows a fistulous opening on the lower third esophagus.
(B) Chest CT shows an outpouching cavitary lesion, which had thin enhancing wall contiguous with mucosal layer of the esophagus (arrow) at the right side of the lower esophagus.
(C) Chest CT with lung window settings shows patchy areas of ground glass opacities and bronchial dilataion with wall thickening, which represents inflammatory change due to chronic and recurrent infection in the superior segment of the right lower lobe.
(D) Barium esophagography confirms the fistulous tract (arrowheads) between the lower third esophagus and the right lower lobar superior segmental bronchus.
(E) Video-assisted thoracoscopic surgery (VATS) demonstrates the fistulous tract about 1cm in diameter (black arrow) between the esophagus (open arrow) and the lung (white arrow).
(F) Follow-up barium esophagography after the fistula close operation. There is no evidence of contrast leakage or remnant fistula tract.
(G, H) Follow-up CT checked at 3 months after the patient discharged from the hospital. Metallic staples (arrows in G) at the surgically closed site of lower esophagus are noted. In the lung window CT, previous noted inflammatory changes (C) in the right lower lobe of the lung is markedly improved.

reported a chronic cough over a period of 20 years. His physical exam and results of laboratory tests were unremarkable.

Our clinical impression was of an abnormal communication between the airway and esophagus. We first performed chest radiograph and CT. The chest radiograph was unremarkable. On CT we observed an outpouching sac, which had thin enhancing walls contiguous with the mucosal layer of the esophagus at the right side of the lower esophagus (Fig. 1B). This finding suggested a mucosal layering fistulous tract arising from the esophagus. Bronchial dilation with wall thickening and peripheral patchy ground glass opacities were noted in the superior segment of the right lower lobe on CT with lung window settings (Fig. 1C), and such findings can represent chronic and recurrent infection. There was no evidence of malignancy or lymphadenopathy around the fistulous tract on chest CT. We performed barium esophagography for further evaluation of communication between the esophagus and airway, which confirmed BEF between the lower esophagus and the right lower lobe superior bronchus (Fig. 1D). We decided to perform surgical treatment to prevent secondary complications such as recurrent pneumonia or abscess. The fistula identified was identified using video-assisted thoracoscopic surgery (VATS) and was 1 cm in diameter (Fig. 1E). The fistulous tract was divided, and each end was closed using surgical staples. No remnant fistula was noted on follow-up barium esophagography after surgery (Fig. 1F). The patient was discharged from the hospital without any complications. After 3 months, follow up CT showed improvement of the inflammatory changes in the superior segment of the right lower lobe and no evidence of any complications (Figs. 1G and 1H).

DISCUSSION

Congenital BEF, originally described by Braimbridge and Keith,[1] results from failed tracheoesophageal separation in the early stages of embryonic development. The congenital nature of such fistulae is characterized by the absence of adherent lymph nodes and surrounding inflammation,[5] by the presence of mucosa and definitive muscularis mucosa within the fistulous tract,[4] and by the onset of symptoms in childhood.[1] In our case, we could not obtain pathologic results of the fistula tract through surgical treatment. However, our patient reported experiencing chronic coughing since childhood and we observed suspicious findings of the mucosal layer within the fistulous tract on CT. Therefore, we diagnosed our patient with congenital BEF. Although there were inflammatory changes around the fistulous tract on CT, we regarded these findings to represent secondary infection due to congenital esophagobronchial fistula. Moreover, some reports of congenital BEF also revealed chronic inflammatory changes around fistula tracts on CT.[6,7]

Patients with BEF may have nonspecific signs over long periods of time, such as vague thoracic or epigastric pain, heart burn, mild dysphagia, weight loss, and recurrent pulmonary infections.[5] Coughing after liquid ingestion (Ohno's sign) is a more specific sign, frequently associated with productive sputum, fever, and hemoptysis.[5] The duration of symptoms varies from 6 months to 50 years before diagnosis.[3-5,8] Usually symptoms depend on fistula size, being more evident in short, wide communications and more discreet in narrow fistulae. Such fistulae may become life threatening, with repeated infections leading to pneumonia, bronchiectasis, and abscess formation.[4,9]

Theories to explain the delay in onset of symptoms include the presence of a membrane, which subsequently ruptures, an occluding proximal fold of esophageal mucosa (flap valve) that becomes less mobile and occlusive with age, and a fistulous tract running upward from the esophagus to the bronchus that may close during swallowing.[1]

Barium esophagography usually aids diagnosis and is the initial study of choice. Esophagoscopy and bronchoscopy may not always demonstrate the fistulous orifice.[5] CT may demonstrate fistulous tracts between the esophagus and airway, but is usually not confirmative.[3] CT can show consolidation, ground glass opacity, masses, or bronchial dilation; these findings represent chronic and recurrent infection, secondary to esophagobronchial fistula. CT is utilized to rule out neoplasm or lymphadenopathy and to define the extent of coexisting pulmonary disease, which may require resection.[3,4]

Once BEF is confirmed, early surgical intervention is considered definitive and results in successful outcomes. Surgical approaches can be divided into two main approaches: division of the fistulous tract, and excision of the entire communication.[3-5] In our case, the fistulous tract was divided and each opening was closed. Additional pulmonary resection is often needed in patients with coexistent pulmonary disease. Some authors have suggested endoscopic and even medical treatment methods to avoid invasive surgical treatment.[10]

Congenital BEF is a rare anomaly in adults with insidious and inconspicuous symptoms, which require a high degree of suspicion for successful diagnosis. Congenital BEF should be considered in the diagnosis of adult patients with suspicious abnormal communications between the esophagus and bronchus without evidence of malignancy or lymphadenopathy around the fistulous tract on chest CT. In these patients, barium esophagography is useful for confirming the presence of the fistula tract and consequently this rare diagnosis.

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