Two Unusual Cases of Adult Onset Congenital Bronchoesophageal Fistulas Treated With Fistula Division

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Adult onset congenital bronchoesophageal fistula is a very rare entity. We report 2 cases of adult onset type II congenital bronchoesophageal fistula between the distal thoracic esophagus and the lower lobe superior segmental bronchi surgically treated through a right and left thoracotomy, respectively. In both cases the fistula was transected and sutured with no parenchyma resection. Both patients had an uneventful recovery. Resection of the underlying parenchyma during surgery for bronchoesophageal fistula is not always necessary as the lung can heal in time after performing just fistulectomy.


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Bronchoesophageal fistula (BEF) is an abnormal communication between the esophagus and the bronchial tree. It can be acquired or congenital. Congenital BEF when associated with esophageal atresia presents early in life. In the absence of coexisting esophageal malformations symptoms may not present until adult life and is often intermittent. Congenital BEF in adult life is rare and most literature comprise of case reports [1, 2]. Most reported literature advocate fistulectomy and resection of the underlying parenchyma. We report the surgical treatment of 2 cases of adult onset BEF where we preserved the underlying parenchyma with both patients having a well expanded lung during follow-up.

The first patient was a 39-year-old male and the second, a 55-year-old female. Both patients had symptoms of bouts of cough when swallowing liquids since childhood. Barium swallow was diagnostic in both patients showing a fistula tract between the esophagus and the lower lobe bronchi (Fig 1). Chest computed tomography scan revealed consolidations and atelectasis in the right lower lobe superior segment, and infiltrations in the left lower lobe superior segment, respectively. Both patients were operated through a thoracotomy. In the first patient bronchoscopy and esophagoscopy confirmed the orifices of the fistula, whereas in the second patient only bronchoscopy revealed the fistula in the left lower lobe superior segmental bronchus. Both patients underwent thoracotomy and had significant adhesions in their lower lobes. Careful dissection of the posterior mediastinal pleura from the level of the carina to the inferior pulmonary vein was done. Large collateral vessels were noticed on the aorta in the female patient. The fistula tract was just above the inferior pulmonary vein between the esophagus and the lower lobe superior segmental bronchi (Fig 2).

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In both patients the fistula tract was divided with scissors and the bronchial defect was sutured with interrupted 4-0 polyglactin, whereas the esophageal defect was closed in 2 layers with interrupted sutures; 3-0 polyglactin for the mucosa and 3-0 silk for the esophageal muscle. Pericardial flap reinforcement on the bronchus was done in the female patient. There was minimal inflammation surrounding the fistula and a clearly visible esophageal muscular wall, having a horizontal course, with no sequester or anthracotic lymph nodes were present. Based on this operative findings diagnosis of type II congenital BEF according to Braimbridge and Keith [3] was confirmed (Table 1). The lower lobes expanded nicely after positive pressure ventilation in both patients, thus no lung resection of the underlying parenchyma was performed.

The postoperative period of both patients was uneventful. Oral feeding was started on postoperative day 5 when contrast swallow was normal. Both patients were discharged on postoperative day 6. The outpatient chest X-rays of both patients 2 months postoperatively revealed a fully expanded lung with complete resolution of the lower lobe atelectasis and infiltrations. There was no morbidity or mortality. The male patient is in the tenth, and the female patient in the fourth postoperative month, and are asymptomatic.

Comment

Congenital bronchoesophageal fistula is rare in adults. Symptoms are nonspecific, bouts of cough when swallowing liquids (Ohno’s sign) is considered pathognomonic and is present in 65% of all cases. The duration of symptoms before diagnosis varies from 6 months to 50 years [4]. Both patients had been tolerant to their symptoms for many years. Many mechanisms explaining the late presentation of congenital BEF have been reported, including the anti-gravitational effect of the fistula tract, imperforated web or fold of the mucous membrane, spasm of smooth muscles in the fistula wall, or patient tolerance to minimal symptoms [5]. Bronchoscopy, esophagoscopy, and barium swallow are essential in the diagnosis, with the latest considered the most sensitive [2]. Definitive diagnosis of congenital BEF include long standing respiratory signs and symptoms from infancy, presence of bronchial or esophageal epithelium lining the interior of the fistula tract, and the absence of relevant inflammation surrounding the fistula [1, 5]. The causes of acquired BEF must be excluded before diagnosis of congenital BEF is made. Both of our cases fulfilled congenital BEF type II criteria. Surgery is the treatment of choice and should not be delayed after diagnosis to avoid respiratory complications or bleeding [1, 2]. There have been reports of less invasive closure techniques with less success [6]. Interventions other than surgery should be limited to patients who cannot undergo surgery. The most frequent site of BEF in the lung has been reported to be the lower lobe, particularly the superior segment as was in our 2 cases [1]. The predilection of BEF in the superior segmental bronchus of the right lower lobe was postulated by Shimada and colleagues [7] to be due to a thinner esophageal musculature and the lack of a soft tissue envelope around the esophagus below the carina. Braimbridge and Keith classified congenital BEF into 4 types with type II been the most common [3]. Most of the reported cases of BEF surgical treatment had the underlying parenchyma resected together with fistulectomy. In our cases, both patients had a recoverable underlying parenchyma hence no parenchyma resection was necessary and the atelectasis and infiltrations in the lower lobes disappeared 2 month after surgery.

Bronchoesophageal fistula should be remembered in the differential diagnosis of persistent chronic cough especially after liquid ingestion, and barium swallow should be included in the diagnostic investigation. Surgical management with transection and suturing of the fistula is the treatment of choice. Resection of the underlying parenchyma is not always necessary as the lung can heal in time after performing just fistulectomy.
Cardiac Cavernous Hemangioma and Multiple Pulmonary Cavernous Hemangiomas

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We describe for the first time a rare coexistence of a cardiac cavernous hemangioma with multiple pulmonary cavernous hemangiomas. Computed tomography revealed bilateral pulmonary nodules, left pleural effusion, and pericardial effusion. Positron emission tomography showed a pericardial neoplasm. Pathologically, multiple large dilated vascular spaces, lined by a single layer of endothelial cells and filled with blood, were revealed in both the cardiac tumor and the pulmonary nodules. Immunohistochemical examination of the lining cells showed positivity for CD31, FLI1, FVIII, and CD34. Taken together, these findings led to the diagnosis of cardiac cavernous hemangioma and multiple pulmonary cavernous hemangiomas.

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Cavernous hemangiomas are characterized by sharply defined yet unencapsulated masses, which are composed of large dilated vascular spaces lined by a single layer of endothelial cells and filled with blood. They are benign vascular tumors that are frequently located in the skin, subcutaneous tissues, and liver. Cavernous hemangiomas can occur in various internal organs, but they rarely occur in the heart and lungs. We report a rare case of a co-occurring cardiac cavernous hemangioma and multiple pulmonary cavernous hemangiomas (PCHs) and review the literature.

A 36-year-old man with a 1-month history of pericardial effusion was admitted to our hospital. Imaging with ultrasonography and chest roentgenography (Fig 1A) showed bilateral pleural effusion and pericardial effusion. Computed tomography (CT) of the chest and heart (Figs 1B, 1C) showed bilateral pulmonary nodules, left pleural effusion, and pericardial effusion. Positron emission tomography showed a pericardial neoplasm. After extensive workup, a thoracotomy, pericardial tumor biopsy, and right lung biopsy were performed. A dark red, easily bleeding cystic tumor, adherent to the right atrium and measuring 8 × 6 cm, was identified in the pericardial cavity. It could not be separated as a result of myocardial invasion. A small edge was removed for pathologic examination. The right mediastinal pleura was opened, and several nodules were found in the subpleural lung tissue. Some of this tissue was also removed for pathologic examination.

The samples were fixed in 10% buffered formalin, embedded in paraffin, sectioned conventionally, and then stained with hematoxylin and eosin. CD31, FLI1, FVIII, CD34, calretinin, WT-1, D2-40, smooth muscle actin, desmin, and caldesmon were detected using immunohistochemical staining methods.

Gross examination revealed the cardiac neoplasm to be a dark red lamellar mass measuring 2 × 2 × 1 cm; it was soft and bled easily. The pulmonary tissue was a dark brown mass measuring 2.5 × 2 × 1 cm and contained multiple sharply defined but not encapsulated nodules. The cut surface showed focal hemorrhagic and cystic areas. Microscopically, multiple irregular dilated vascular spaces lined by a single layer of endothelial cells were observed in the cardiac tumor (Fig 2A). Similarly, several nodules composed of large dilated vascular spaces were scattered in the lung tissue; these were variably filled with blood. The spaces were lined by a single layer of thin endothelial cells (Figs 2B, 2C). Immunohistochemical examination of the lining cells revealed positivity for CD31 (Fig 2D), FLI1, FVIII, and CD34, consistent with a lesion of endothelial origin. In addition, some dilated thick vascular walls in the pulmonary nodules were positive for smooth muscle actin. The results of staining for calretinin, WT-1, D2-40, desmin, and caldesmon were negative.

Comment

Hemangiomas are benign tumors that are mainly composed of blood vessels. The histologic classification...