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Congenital Bronchoesophageal Fistula Associated with Esophageal Diverticulum in the Adult

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Congenital bronchoesophageal fistula is a rare clinical entity in adults. This anomaly may cause various symptoms such as respiratory infections, coughing bouts when eating or drinking, and even hemoptysis. The fistula can cause symptoms in childhood but may not appear until adulthood. We recently experienced a case of congenital bronchoesophageal fistula associated with esophageal diverticulum in an adult. A 63-year-old woman was admitted to our hospital due to chest discomfort, sore throat and coughing bouts when eating. An empyema with lung abscess had occurred eight years previously. Results of the physical examination were unremarkable. A Barium swallowing revealed a medium-sized diverticulum at the right anterior aspect of the esophagus, which had developed a fistulous connection with the right lower lobe bronchus. The patient was treated by fistulectomy and lobectomy of the right lower lobe. The postoperative course was smooth and uneventful.

Key Words: Congenital bronchoesophageal fistula, esophageal diverticulum

Congenital bronchoesophageal or tracheoesophageal fistula is a rare clinical problem in adults. It is usually associated with esophageal atresia and is readily diagnosed in the neonatal period. But if it is not associated with esophageal atresia, it may persist until adulthood before the diagnosis is established (Azoulay *et al.* 1992; Juhani et al. 1995). The diagnosis of this condition may be difficult due to its insidious clinical course. If a respiratory-esophageal fistula is not suspected and is therefore left untreated, it may lead to fatal complications despite the benign nature of this anomaly (Risher *et al.* 1990; Juhani *et al.* 1995).

We present a case of congenital bronchoesophageal fistula in an adult which was associated with esophageal diverticulum.

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CASE REPORT

A 63-year-old woman was admitted to our hospital on September 21, 1996, with the chief complaints of chest discomfort, sore throat and coughing bouts when eating which had first started 20 years before and had recently been aggravated for one month. She had been sufferring from the symptoms but did not seek proper treatment. In 1988, she had been admitted to another hospital due to pyogenic empyema with a lung abscess in the right lower lobe and

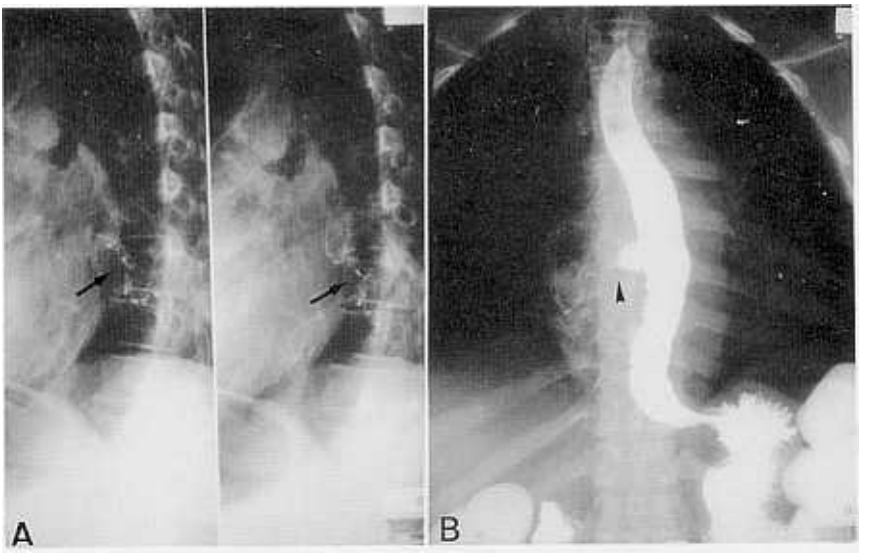

Fig. 1 A, B. Barium esophagogram demonstrates communication between a diverticulum arising from the right anterior wall of the mid-esophagus and the superior portion of the right lower lobe bronchus.

treated with antibiotics and closed thoracostomy. Eighteen months prior to admission to our hospital, during an upper gastrointestinal examination for a routine check-up, a mid-esophageal diverticulum was noted, but further evaluation and treatment were not performed and her symptoms were not aggravated.

From one month prior to admission, recurrent chest discomfort, sore throat, and coughing bouts when eating had developed.

On physical examination the patient was found to be a well-developed, thin woman, who did not appear acutely or chronically ill. Her temperature was 36.8°C, blood pressure was 140/90 mmHg, her heart rate was 70/min, and respiration was 20/min. Positive physical findings were limited to the chest, where a few coarse rales were noted in the right posterior lung base.

Laboratory tests and pulmonary function studies were all within normal limits. Esophageal manometry and 24-hour pH moniter study did not reveal any pathologic findings.

A barium swallow demonstrated a communication between a diverticulum arising from the right anterior wall of the mid-esophagus and the superior portion of the right lower lobe bronchus (Fig. 1). Bronchoscopic examination did not reveal endobronchial lesion or bronchoesophageal fistula outlet opening.

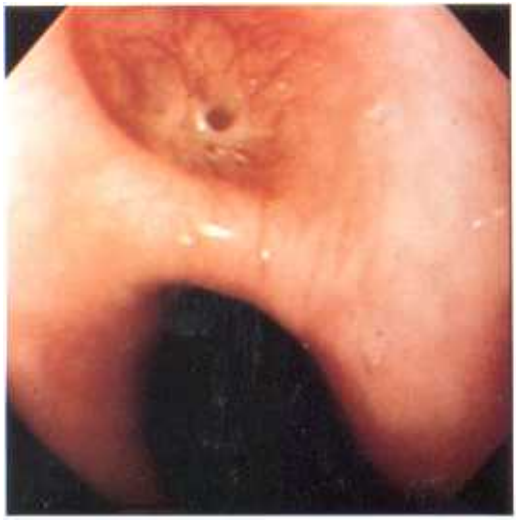

Fig. 2. Esophagoscopic examination shows fistulous opening orifice at 30cm from upper incisor teeth.

On esophagoscopy, a fistulous opening orifice was observed at 30 cm from the upper incisor teeth (Fig. 2). The plain film of the chest showed haziness in the right lower lobe and blunting of the right costophrenic angle (Fig. 3). The chest CT scan showed complete collapse of the right lower lobe basal segment with bronchiectatic change and calcifications,

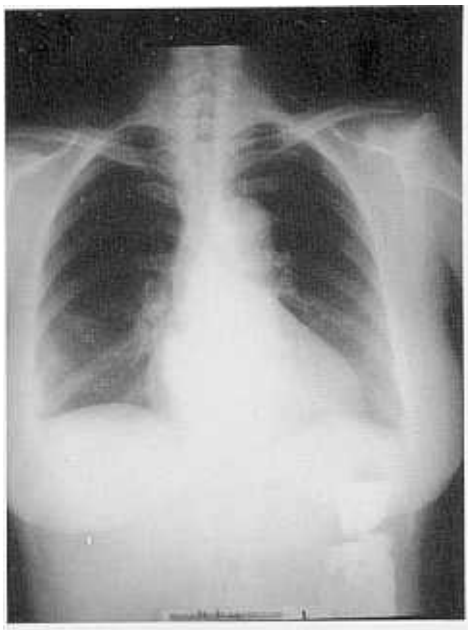

Fig. 3. Initial plain film of the chest reveals haziness in the right lower lobe and blunting of the right costophrenic angle.

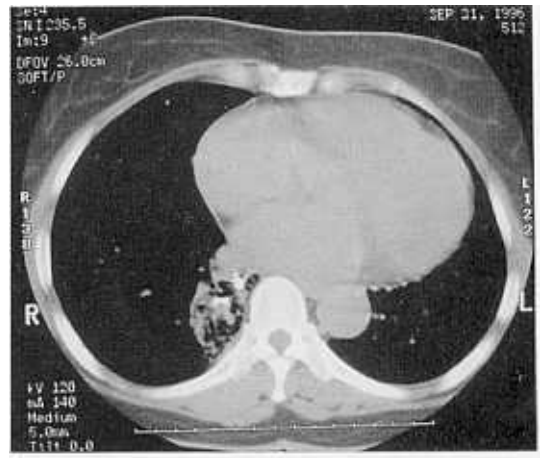

Fig. 4. Chest CT scan shows complete collapse of the right lower lobe basal segment with bronchiectatic change and calcifications, probably due to old inflammatory sequelae, but no definite fistulous tract is shown.

Fig. 5. Operative photograph of the fistula. The fistula was well separated and both openings were closed with sutures and the right lower lobectomy was performed. The tract arose from the base of the mid-esophageal diverticulum, which had developed a fistulous connection with the superior segment of the right lower lobe bronchus.

probably due to old inflammatory sequelae, but no definite fistulous tract was shown (Fig. 4).

Our impression was bronchoesophageal fistula with destructive lung change and the patient was transferred to a chest surgeon.

Through right thoracostomy, the fistular tract was easily separated and excised. Right lower lobe lobectomy was also performed due to a destructive lung change. The fistulous tract arose from the base of the mid-esophageal diverticulum, which had developed a fistulous connection with the superior segment of the right lower lobe bronchus (Fig. 5). Pathologic examination of the resected specimen revealed a lining of the proximal portion of the fistulous opening with intact esophageal mucosa (squamous epithelium) and a lining of the distal portion with intact respiratory epithelium. An area of direct contact between the squamous epithelium of the divertiulum and the pseudostratified columnar bronchial epithelium was also demonstrated (Fig. 6). There was no evidence of inflammatory cell infiltration. We confirmed it as a congenital bronchoesophageal fistula (Braimbridge type I), even though it was found in a 63-year-old woman. The patient's postoperative course was uneventful and she was discharged, asymptomatic, on her 15th postoperative day.

DISCUSSION

Bronchoesophageal fistulae may be either congenital

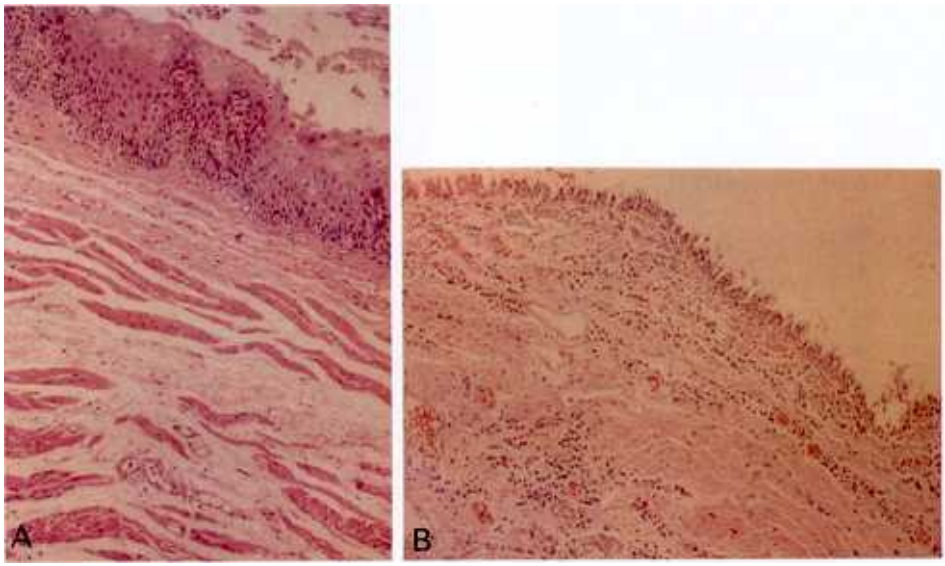

***Fig. 6** A, B. Photomicrograph of the specimen resected shows lining of the proximal portion of the fistula opening with intact esophageal mucosa (squamous epithelium) and lining of the distal portion with intact respiratory epithelium (pseudostratified columnar epithelium). Organizing and adhesive inflammation around the fistulous tract and adjacent lung parenchyma with bronchiectatic change is shown.*

or acquired, with the latter being divided into benign and malignant types. Congenital bronchoesophageal fistulae are 25% to 50% less common than tracheoesophageal fistulae (Smidth, 1970; Scheiner and Lachance, 1980; Hendry *et al.* 1985). Tracheoesophageal fistulae are usually present in early infancy, whereas bronchoesophageal fistulae are diagnosed after age 15 in 75% of cases. The reason is that tracheoesophageal fistulae are often associated with esophageal atresia and connected with a large airway (Chu and Mullen, 1978; Scheiner and Lachance, 1980; Hendry *et al.* 1985).

The congenital nature of these fistulae is still controversial, but it may be due to an abnormal growth of the trachea during its separation from the esophagus; this abnormal growth produces a persistent communication between the tracheoesophageal tree and the esophagus (Gerle *et al.* 1968; Azoulay *et al.* 1992).

There is no sex prevelance in its incidence and a relatively even distribution of cases in all age groups, with the highest incidence occurring in the third decade of life (Risher *et al.* 1990; Kim *at al.* 1995). The fistulae are almost three times more common on the right side than on the left, and the most frequent site of communication is the right lower lobe, particularly the superior segment (Risher *et al.* 1990; Juhani *et al.* 1995). In our patient, the fistula was on the right side and the communication was between the mid-esophagus and superior segment of the right lower lobe.

Braimbridge and Keith described four types of congenital fistulae (Braimbridge and Keith, 1965). Type 1 is associated with a wide-necked congenital diverticulum of the esophagus with an inflamed tip that perforates into the lung. Type 2 is a short tract running directly from the esophagus to the bronchus or the trachea. Type 3 is a fistulous tract connecting the esophagus to a cyst in the lobe, which in turn communicates with the bronchus. Type 4 fistulae run into a sequestrated segment or lobe. Type 2 is the most common. (Braimbridge and Keith, 1965). Our case, a bronchoesophageal fistula associated with esophageal diverticulum, was a type 1 fistula.

It may be difficult to differentiate a congenital form from an acquired form, especially if an advanced pulmonary disease exists. The type 1 fistula may represent an acquired fistula secondary to infection and a perforation of a congenital diverticulum of the esophagus. Generally, the criteria for the diagnosis

of a congenital fistula are as follows: (1) the absence of past or present surrounding inflammation; (2) the absence of adherent lymph nodes; and (3) the presence of mucosa and definite muscularis mucosa. The mucosa may be lined by squamous or columnar epithelium. Transition of the epithelial lining of the fistula from squamous (esophageal) to columnar (respiratory) is another criterion according to other reports (Smidth, 1970; Acosta and Battesbay, 1973; Chu and Mullen, 1978; Risher *et al.* 1990; Kim *et al.* 1995). But these criteria are not correct in all cases, especially in type 1 fistulae. Our patient had mucosa which consisted of squamous epithelium with a transition to columnar epithelium, and muscularis mucosa on microscopic examination. Our case was compatible with the above and we diagnosed it as a congenital type of bronchoesophageal fistula in an adult.

Presenting symptoms usually include coughing (96%), recurrent lung infections (56%), and bouts of coughing when swallowing liquids (Ohno's sign: 65%) (Braimbridge and Keith, 1965; Azoulay *et al.* 1992). Hemoptysis and dysphagia are less common. Other symptoms may include shortness of breath or wheezing and may be present for days-to-years before diagnosis.

The long, silent interval until adulthood and the irregular character of the signs have drawn various explanations. These include: (1) an occlusion of the opening by an esophageal tissue fold or a "flap valve"; (2) the presence of a thin membrane that subsequently ruptures; (3) the action of gravity (upward direction of the fistula from the esophagus to the bronchi) preventing spillage of the esophageal contents into the respiratory tree; (4) adaptation of patients to the minimal symptoms; and (5) spasm of the smooth muscle in the fistula wall. However, none of these is well supported by pathologic or radiologic findings (Smidth, 1970; Kim *et al.* 1995).

The most useful investigation is barium or cine-esophagography, which is diagnostic in over 65% of cases (Bekoe *et al.* 1974; Hendry *et al.* 1985). Other tests that have been used but are less successful include bronchography, esophagoscopy, and bronchoscopy (Yacoub *et al.* 1973; Scheiner and Lachance, 1980). Oral methylene blue is the least useful of the diagnostic tests (Hendry *et al.* 1985).

The usual treatment is excision of the fistula with closure of the esophageal bronchial defects and interposition of pleural or muscle flaps. Resection for a bronchiectatic or damaged lung is limited to the area of infection (Hendry *et al.* 1985). More hazardous and less efficient forms of suggested treatment have included occlusion of the esophageal opening with use of a silver nitrate, biologic glue, or Celestin tube (Laforet, 1978; Azoulay *et al.* 1992).

The most common complication is pulmonary suppuration, such as pneumonia, abscess, and bronchiectasis (Hendry *et al.* 1985).

In conclusion, bronchoesophageal fistula, either congenital or acquired, is benign but may be life threatening if left untreated, and the possibility of a bronchoesophageal fistula should be suspected when recurrent pulmonary suppuration occurs. Esophagography may confirm the diagnosis and surgical treatment should be taken as soon as possible after the diagnosis is established.

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