Congenital Bronchoesophageal Fistula in the Adult

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A case of congenital bronchoesophageal fistula was reported with an updated review of the literatures. Congenital bronchoesophageal fistula may persist into adult life causing chronic cough or choking on swallowing or can be an uncommon source of chronic bronchopulmonary sepsis. Diagnosis is relatively easy and operative treatment is very successful. This anomaly should be considered when there are typical symptoms or chronic pulmonary sepsis.

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Most bronchoesophageal fistulas are acquired in nature and the congenital one is rare.[1,2] The congenital fistula associated with esophageal atresia usually requires immediate diagnostic and therapeutic action. The fistula without atresia often has insidious clinical course and may persist into adult life. Chronic aspiration through the fistulous tract may lead to repeated pulmonary infection or sepsis. If the diagnosis is made and handled properly, permanent damage of the lung can be prevented. A recent case prompted this report and an updated review of the literatures.

CASE REPORT

A twenty-two years old woman came to the medical attention because of chronic periodic and intermittent epigastric discomfort unrelated to meals and no nocturnal attack for a few months. She had had frequent regurgitation and excessive cough during rapid swallowing, particularly water or liquid diet but no dysphagia, since early childhood. During this period, her general health remained stable and as a matter of fact she had the tendency to gain weight. Her past history, family history and review of other systems were non-contributory.

Physical examination revealed a healthy young woman without any acute distress. Her vital signs were normal. Both lungs were clear on auscultation. No difinite tenderness at epigastrium. The essential laboratory tests included hemogram, urinalysis, blood sugar, blood urea nitrogen, creatinine and the screening liver biochemical tests were all within normal limits. The chest roentgenogram was normal. The barium upper gastrointestinal series clearly demonstrated a bronchoesophageal fistula between midesophagus and the lower part of left main bronchus (Figure 1). Bronchoscopy and esophagoscopy were not performed on this patient.

On January 16, 1980, a left thoracotomy was performed. Minimal adhesion was noted in the pleural cavity. A fistulous tract 0.5 x 1.7 cm connected the middle third of the esophagus to inferior surface of the distal left main bronchus was found. (Figure 2) There was no lymphadenopathy or evidence of inflammation around the fistulous tract or the esophagus. There was mild degree of bronchiectasis of

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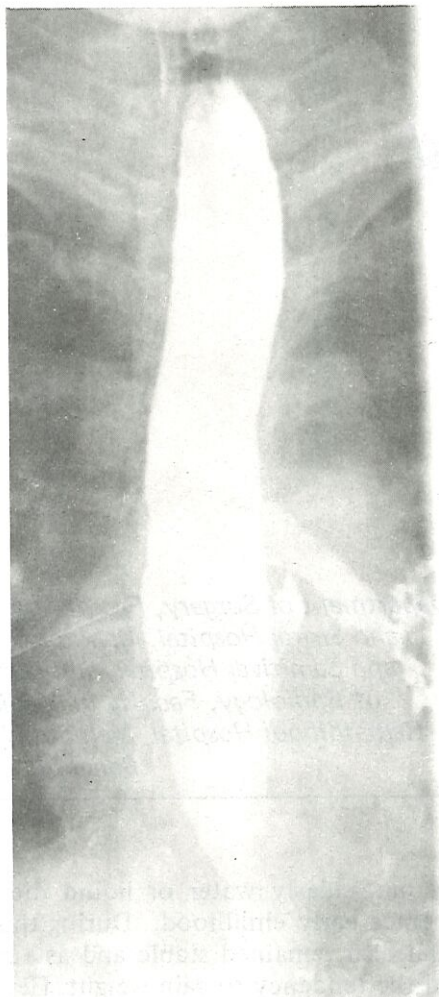

Fig. 1 The barium esophagogram demonstrated a fistulous tract between the mid-esophagus and the lower part of left main bronchus.

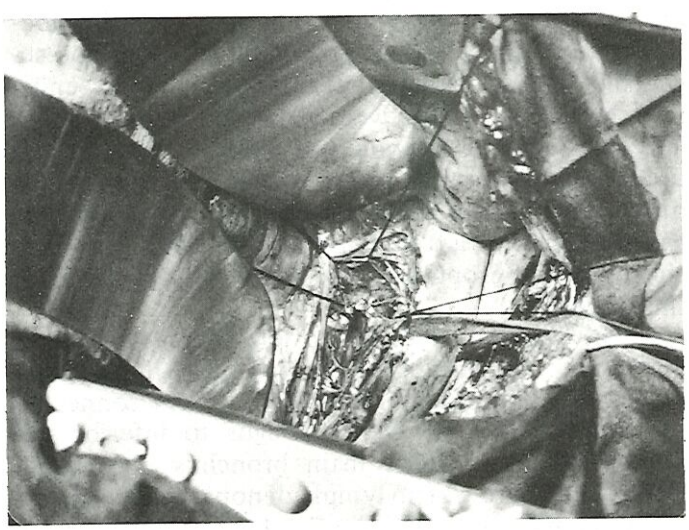

Fig. 2 The left lung was retracted medially and the descending aorta was encircled with an unbilical tape and pulled laterally to show the fistulous tract

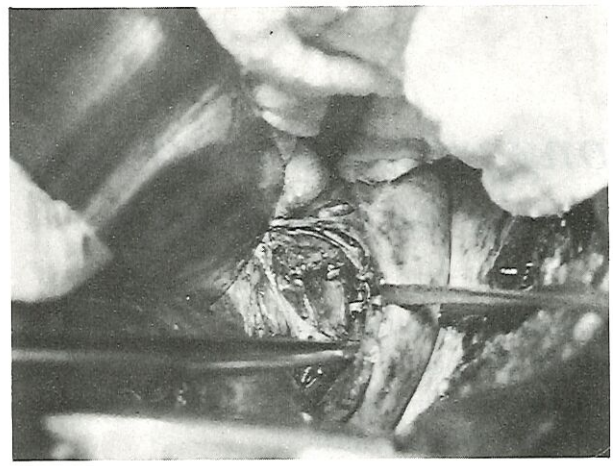

Fig. 3 The fistulous tract stumps after the division and suture. Notice an absence of inflammatory process or lymphadenopathy around the tract.

the left lower lobe bronchus as it was somewhat hard and irregular, but no pneumonitis was encountered. The fistulous tract was excised and the bronchial and esophageal defects were closed (Figure 3). Histologic examination showed a squamous mucosal lining with muscularis mucosa and no evidence of neoplastic or inflammatory processes in the fistulous tract (Figure 4, 5). Her postoperative course was uneventful and she remains asymptomatic during the follow-up.

DISCUSSION

Although the first congenital bronchoesophageal fistula was described in the autopsy report by Heiderich[3] in 1916, it was not until 1936 when Camplani[4] reported the first clinical diagnosis of such anomaly in an adult. Braimbridge and Keith[5] described four types of congenital bronchoesophageal fistula. Type 1 is associated with a wide-necked congenital deverticulum of the esophagus that perforates into the bronchus following an fnflammatory process. Although there is a congenital background, the fistula is inflammatory in origin and it may be difficult to distinguish this diverticulum from the traction variety. Type 2 is a simple tract joining the esophagus to a lobar or segmental bronchus. Type 3 is a fistulous tract with a cyst in the lung interposed between the esophagus and the bronchus. Type 4 is a fistula entering the bronchus of a sequestrated pulmonary segment. In thier review of 23 cases of congenital bronchoesophageal fistula, type 2 was the most common (57%) followed by Type 3 (26%) and Type 4 (17%). Blackburn and Amoury[6] reviewed the literatures in 1966 and reported on 51 cases of congenital bronchoesophageal fistula in adults. Since that review, 18 additional cases have

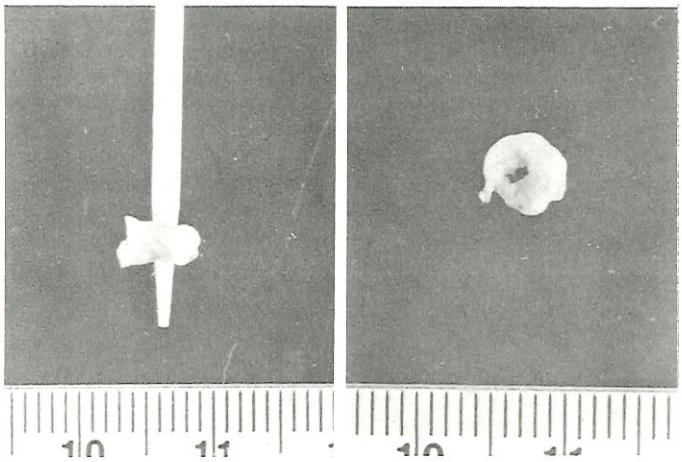

Fig. 4 Gross appearance of the segment of fistulous tract

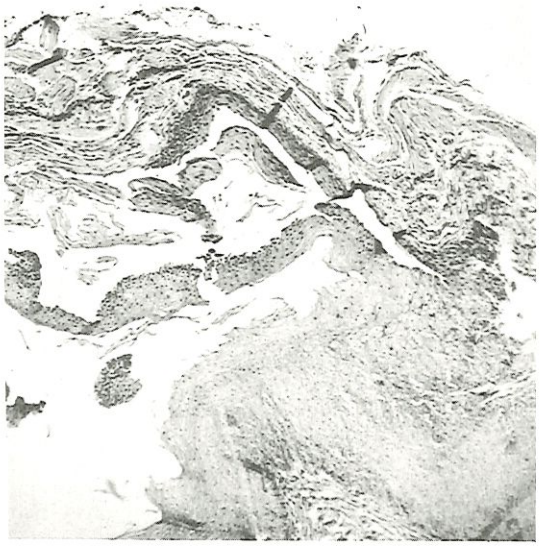

Fig. 5 Section revealed a tube lined by stratified squamous epithelium surrounded by fibrous connective tissue and bundles of smooth muscle. No evidence of neoplastic or inflammatory process was seen.

been described.[7] In this series of 70 cases (included ours), there was no overwhelming sex preponderance. The ages at the time of diagnosis varied from the late teens to the seventh decade without a predilection. Symptoms may not begin until adult life and are often intermittent, usually due to chronic bronchopulmonary suppuration. The most common symptom that prompted the patient to seek medical attention was regurgitation and chronic cough as seen in almost all cases reviewed by Braimbridge and Keith[5] whereas pneumonitis was seen in more than half of the cases and hemoptysis was seen in only 4 out of 23 cases. Other esophageal symptoms were uncommon and among those epigastric discomfort which occured in our patient was mentioned. The late appearance of symptoms and diagnosis has been explained in various ways : (1) The presence of membrane that subsequently ruptures[8] (2) a flap valve of esophageal mucosa that subsequently becomes incompetent[9] and (3) the frequent upwardly obligue nature of the tract prevents reflux.[10] In our patient the fistulous tract had an upward direction from midesophagus to the left main bronchus thus supporting the third explanation. The fistula itself gives no definite physical signs but chronic bronchial sepsis and pneumonitis can cause clubbing of the fingers, basal rhonchi, rales and pleural effusion. The diagnosis is usually made by barium swallow where the opague medium is seen in the fistulous tract or sometime in the bronchial tree. In Braimbridge and Keith's review,[5] they reported preoperative diagnosis of bronchoesophageal fistula in 65% of cases whereas the diagnosis was made at an operation for pulmonary sepsis in the remainning 35%. A cine esophagography was also used to confirm the diagnosis in one case.[7] Esophagoscopy, bronchoscopy and bronchography rarely demonstrate the fistula. Fistulas involving the right bronchial tree were over twice as frequent as those involving the left[6].

The most effective form of treatment is operative resection of the fistula and repair of esophageal and bronchial defects. In some cases, resection of permanently damaged pulmonary lobe or segments are necessary. There were 26% of patients in Braimbridge and Keith's review in whom the fistula only was closed but 74% of patients had to have lung resection in addition. This indicated that majority of patients with congenital fistula were diagnosed late when they already had pulmonary complication as result of repeated aspiration.

The congenital nature of a fistula may be assumed when there are no evidence of past or present inflammation around the fistulous tract or esophagus, no adherent lymph nodes and there must be a mucosa and muscularis mucosae histologically. The absence of inflammation makes operative dissection of the fistulous tract easy and simply excludes an acquired type with inflammatory nature.

The operative results have been excellent, with no operative death observed in the reviews. Most patients experienced complete symptomatic relief. Our patient has been followed up for 6 months with no evidence of recurrent symptoms.

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