Medical Journal of the
Islamic Republic of Iran

Volume 12
Number 3
Fall 1377
November 1998

Case Reports

CONGENITAL BRONCHO-ESOPHAGEAL FISTULA: A CASE REPORT

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ABSTRACT

Congenital broncho-esophageal fistula (BEF) is a rare anomaly usually detected in adulthood. In one of the latest reviews of this anomaly, no more than 150 cases were found in the world literature. We report our experience with a 49 year old male patient referring with a classic presentation of chronic cough and choking episodes upon liquid intake. Broncho-esophageal fistula was confirmed by an esophagogram and the congenital nature of the fistula was established by the pathologic report notifying the presence of squamous cell lining of the fistula tract.

We conclude that the esophagus should be investigated more frequently when patients have respiratory complaints at their initial presentation, since pulmonary symptoms may indirectly reflect esophageal disease.
MJIRI, Vol. 12, No. 3, 295-298, 1998

INTRODUCTION

Congenital fistulous communications between the esophagus and trachea or one of the main bronchi are usually associated with esophageal atresia and easily diagnosed in the neonatal period. A tracheobroncho-esophageal fistula without esophageal atresia (H-type) is compatible with life and may occasionally persist until adulthood before the definitive diagnosis.[1] Most probably there are more cases of congenital BEF encountered in adults than have been reported. To date, no more than 150 cases have been reported in the world literature,[2] including only those patients with fistulous communications between the esophagus and bronchial tree and excluding those cases with tracheo-esophageal fistula.

Our own experience with a 49 year old male patient with a classic presentation forms the basis of this article. Special emphasis is placed on the more extensive esophageal studies in patients presenting with chronic respiratory symptoms, particularly in adult life.

Case report

A 49 year old man was admitted to the hospital with cough and fever due to right lower lobe pneumonitis. He was doing well up to 6 months prior to admission. In the past 6 months he was hospitalized 3 times with recurrent non-migratory right lower lobe pneumonia. During this period of time, repeated episodes of coughing and choking, especially after liquid intake, would occur. Physical examination revealed diffuse rales and decreased breathing sounds at the base of the right lung. Chest roentgenogram showed right lower lobe consolidation and air in the esophagus (Fig. 1).

Barium esophagogram revealed an oblique fistulous tract between the lower esophagus and right lower lobe bronchus (RLLB) (Figs. 2,3). Bronchoscopic study was

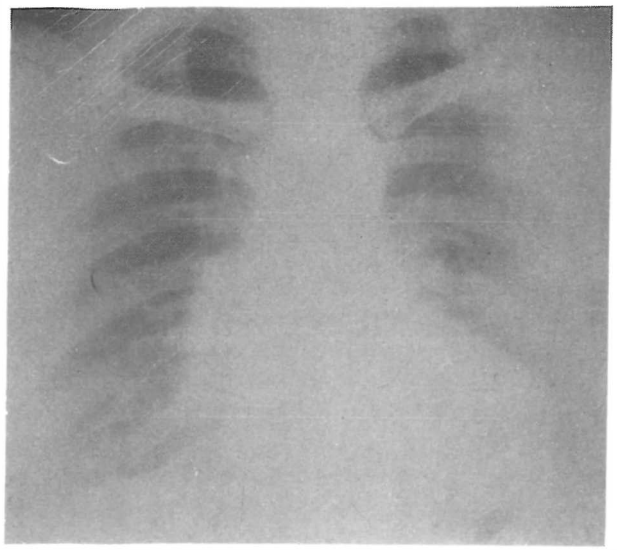


Fig. 1. Chest roentgenogram of the patient showing right lower lobe consolidation.

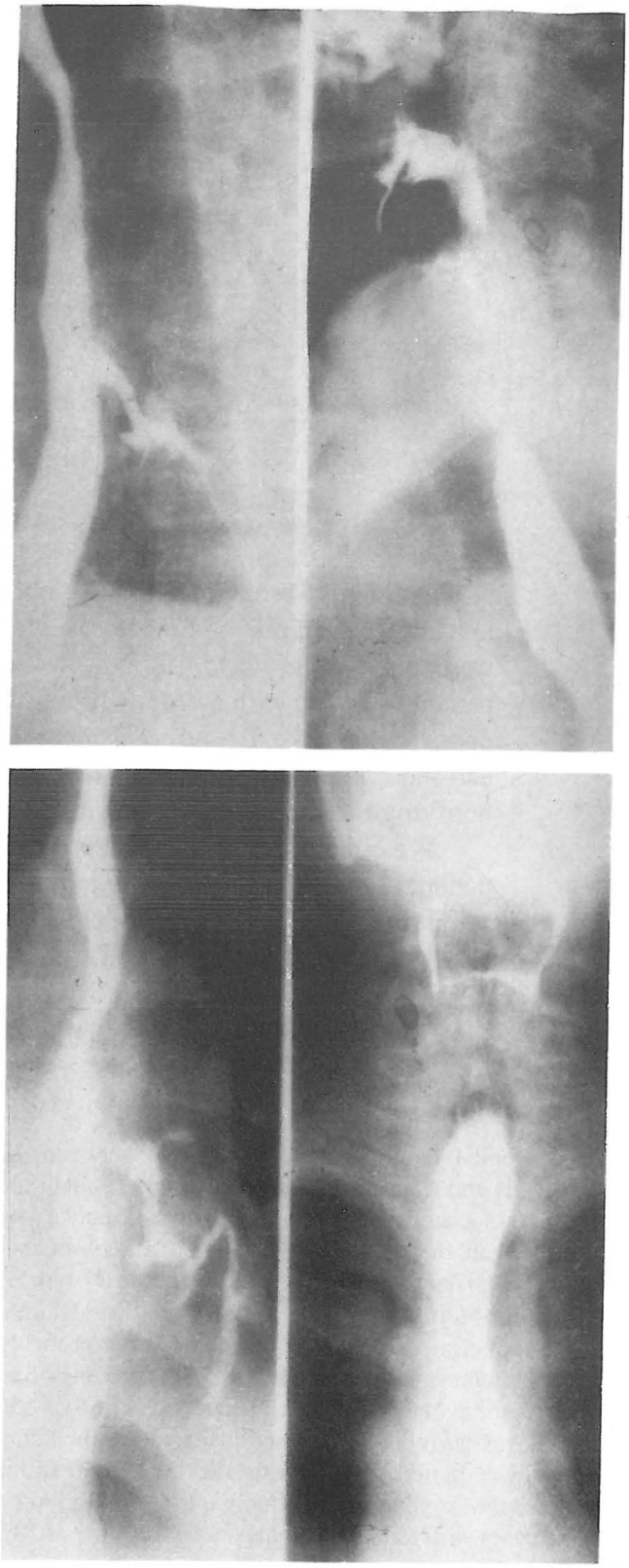
done and an area of inflammation seen in the RLLB. Biopsy was taken and secretions sent for acid fast staining. Esophagoscopy was done and a small area of inflammation was found in the lower esophagus, from which a biopsy was taken. Both esophageal and bronchial biopsies were negative for malignancy. Smear and culture was negative for tuberculosis. With an impression of broncho-esophageal fistula, he underwent a right thoracotomy with division and partial excision of the fistulous tract and interposition of a pleural flap. The dissection was simple and performed meticulously with no evidence of past or present inflammation around the fistulous tract, bronchus, or esophagus. The fistula was 2.5 cm in length and 1 cm in its outer diameter. Repair of bronchial and esophageal sides was accomplished with interrupted 4-0 vicryl.

He had an uneventful and smooth postoperative course and was discharged one week after surgery. Pathologic report of the fistulous tract was squamous epithelium and muscularis mucosa with mild non-specific inflammation.

DISCUSSION

Congenital broncho-esophageal fistula is very rarely seen in adults and its true incidence is basically unknown. However, broncho-esophageal fistula may be neoplastic, congenital, traumatic, or inflammatory.[3] The most common cause is malignant disease.[4] The rarity of the congenital type of the disease in adults has been documented by various authors.[5]

A review of the literature shows that BEFs occur in a roughly equal frequency in male (53%) and female patients (47%).[3] In a review of 100 cases of BEFs by



Figs. 2,3. Barium esophagogram clearly shows an oblique fistulous tract between the lower esophagus and the right lower lobe bronchus.

Rishor et al.,[3] the locational distribution of fistula tracts with the bronchial tree were as follows: right lower lobe bronchus (41 patients); left lower lobe bronchus (21), right main bronchus (18), bronchus intermedius (10), left main bronchus (16), right middle lobe bronchus (2), and right upper lobe bronchus (2), with no fistulas to the left upper lobe.

The embryological explanation of the origin of BEF has been addressed by Smith.[6] They may be due to abnormal growth of the trachea during its severance from the esophagus; this abnormal growth produces persistent communication between the tracheo-bronchial tree and the esophagus.[1] The congenital nature of the fistula in an adult is also suggested by the following features;

a) Duration of symptoms, usually since childhood;

b) Absence of a past history of infection or malignant diseases;

c) Tracheal or proximal bronchial level of fistula;

d) Complete recovery after resection of the fistula;

e) Association with other congenital malformations;

f) Presence of mucosa (usually of the squamous type) and muscularis mucosa, together with absence of adherent lymph nodes.[8-10]

In 1966, Braimbridge and Keith[9] proposed the following classification for congenital BEFs, dividing them into four distinct types;

Type I: Esophageal diverticulum with a large ostium and a fistula at its tip;

Type II: A short tract running directly from the esophagus to the lobar or segmental bronchus. This is the simplest and most common type.

Type III: A fistulous tract connecting the esophagus to a cyst in the lobe, which in turn communicates with the bronchus.

Type IV: A fistula that runs into a sequestrated segment or lobe which is recognized by the presence of a systemic arterial supply from the aorta.

The single patient in our experience had a type II anomaly. Among 150 cases of congenital respiratory fistula reported by Azoulay et al,[2] only 10 cases of associated lung sequestration were found.[11]

Symptoms may occasionally begin in childhood but rarely at birth. The cause of the delay has been attributed to: (I) the presence of a membrane that subsequently ruptures,[12] (II) the proximal fold of esophageal mucosa initially overlapping the orifice but subsequently becoming less mobile,[13] and (III) to the fact that the fistulous tract runs upward and may close during swallowing.[14]

The duration of symptoms may vary from 6 months to 50 years before detection and treatment. They usually consist of chronic broncho-pulmonary suppuration, cough (almost universal), pneumonia, and hemoptysis. The presence of food in the sputum or choking on swallowing liquids may lead to a diagnosis of BEF.[3]

The most useful diagnostic investigations are the esophagogram with water soluble contrast medium or dilute barium, and cine-esophagography with the patient in the prone or anterior oblique position.[15] These investigations usually provide a diagnosis in more than 65% of cases[16] and one of the series confirmed the diagnosis in 78% of cases.[2]

Other diagnostic techniques include esophagoscopy, bronchoscopy and occasionally bronchography supplemented with a CT-scan for assessment of the extent of coexistent pulmonary lesions.[17] Associated pulmonary sequestration may be documented by aortography.[11]

The treatment of choice is surgical intervention, mainly by either division and suturing of the fistula or by complete resection of the fistulous tract.[6] A pleural flap or muscular flap is inserted between the esophagus and the respiratory tract to prevent any refistulization, as has been reported in about 10% of cases.[18] Obviously, any diseased lung tissue and any sequestrated lung lobes should be resected at the same time.[17]

Cure has been reported after obliteration of the esophageal end of the fistula with silver nitrate, but this method seems to be a less efficient and unreliable technique which is reserved for those patients whose condition precludes a thoracotomy.[19] There has been no operative mortality in the review of cases of BEF.[3]

In conclusion, we believe that the esophagus should be investigated more extensively and with a high index of suspicion when patients initially present with respiratory complaints, since pulmonary symptoms may reflect esophageal disease.

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