

CASE REPORT

Congenital oesophageal respiratory tract fistula presenting in adult life

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ABSTRACT: Congenital oesophageal respiratory tract fistula presenting in adult life is rare. A tracheo-oesophageal and a broncho-oesophageal fistula presenting in a 22 year old female and 68 year male, respectively, are described. In both, symptoms of chronic recurrent pulmonary suppuration were initially attributed to alternative aetiologies, resulting in diagnostic delay and inappropriate management. Correct diagnosis was established by flexible bronchoscopy and oesophageal contrast studies. Surgical division of fistulae resulted in complete symptom resolution.

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The common origin of the respiratory tract and oesophagus from the same embryological structures sometimes gives rise to anomalous communications between them. The majority of congenital fistulae between the oesophagus and the respiratory tract are associated with oesophageal atresia, and present in infancy. Rarely the fistula may occur in the absence of oesophageal atresia. The condition is, therefore, compatible with life, and the patient can progress into adulthood without detection.

Two such cases of congenital oesophageal respiratory tract fistulae occurring in adults are presented. The paper highlights the need for thorough evaluation for occult oesophageal respiratory fistula in any patient with chronic pulmonary suppuration of unknown aetiology. Timely diagnosis and treatment will avoid serious complications and years of debilitating illness.

Case one

A 21 year old female waitress presented with a chronic productive cough. From childhood, she had suffered repeated chest infections and a persistent nocturnal cough, the frequency and severity of which had increased over the past 2 yrs. Because of the persistent respiratory symptoms, she had undergone investigations for cystic fibrosis, skin testing for allergies and had a tonsillectomy at 13 yrs of age. She had never smoked or had known exposure to tuberculosis.

On examination, she was of normal weight and height for her age and sex. Early finger clubbing was present. Auscultation of the chest revealed diminished breath sounds, with crepitations at the right lower zone. Chest X-ray revealed right middle and lower lobe collapse with consolidation.

In view of the lobar collapse, a flexible bronchoscopy was performed. This revealed a simple opening in the posterior wall of the trachea, 15 cm from the incisors. Contrast

injected into this opening demonstrated a communication with the oesophagus. At oesophagoscopy a normal, healthy looking oesophageal mucosa was visualized, together with a small opening in the area of the mid-oesophagus. Secretions and air bubbles were seen expressing from this opening into the oesophageal lumen, when the patient was asked to cough. A tube oesophagram was performed using water soluble contrast material. On swallowing, the contrast quickly escaped from the oesophageal lumen into the respiratory tract, but the definite fistulous tract could not be visualized (fig. 1).

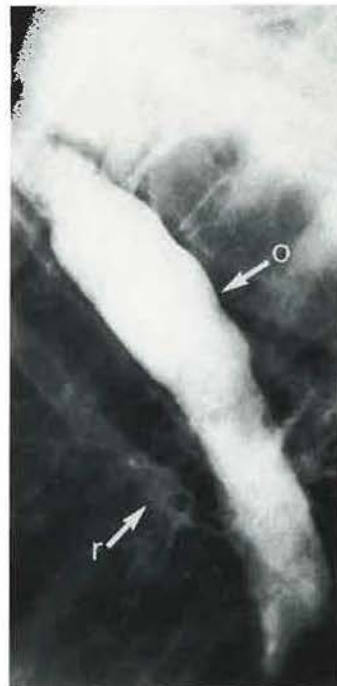


Fig. 1. - Tube oesophagram. Contrast within the oesophagus (O) spilling into the respiratory tract (r).

A right thoracotomy was performed. A fistulous tract, 2 cm long, was easily delineated running upwards from the mid-oesophagus to the lower trachea. There was no surrounding inflammation or adherent lymph nodes. The fistula was ligated and divided and vascularized intercostal muscle was interposed.

The patient's postoperative recovery was uneventful, and after one year her respiratory symptoms have fully resolved.

Case two

A 68 year old male retired factory manager presented with chronic productive cough and haemoptysis. Respiratory symptoms first began at the age of 40 yrs, when he complained of a productive cough and right-sided pleuritic pain. Chest X-ray revealed a shadow involving the right lower lobe, suggestive of a neoplasm. A rigid bronchoscopy was performed, but did not identify an endobronchial lesion. Subsequently, his symptoms and X-ray appearance resolved on antibiotic treatment. Respiratory symptoms had been quiescent until 2 yrs ago, when he again developed repeated right-sided chest infections. Furthermore, over the past 12 months, he had noticed a tendency to cough when eating and drinking. No food particles were identified in the sputum. Because of increasing exertional dyspnoea he had been commenced on anti-anginal medication, with little benefit. He was a nonsmoker, with no known tuberculosis exposure.

On physical examination, the patient appeared well. No lymphadenopathy was found. Diminished breath sounds and coarse crepitations were heard at the right lower lung zone. There was no digital clubbing or cyanosis.

A chest X-ray revealed consolidation, with cavitation of the right lower lobe. A barium swallow examination



Fig. 2. - Flexible bronchoscopy. Contrast injected into the right main bronchus entering the oesophagus via the broncho-oesophageal fistula.

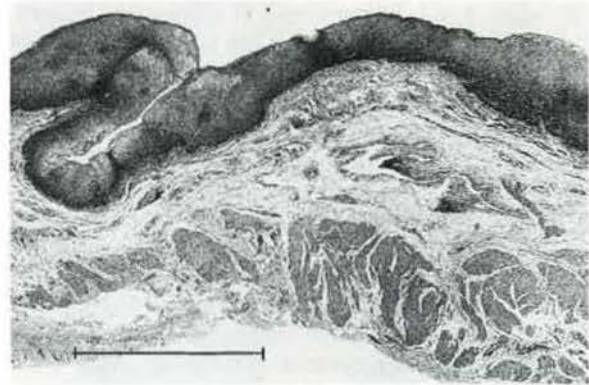


Fig. 3. - Histology of the excised fistula, demonstrating squamous epithelium and muscularis mucosa, with no evidence of inflammation or malignancy. (Scale bar = 0.33 mm.).

showed an oesophageal diverticulum in the middle third of the oesophagus. Contrast medium spilled into the right lower lobe, through a fistulous communication between the oesophageal diverticulum and the right lower lobe bronchus.

Flexible bronchoscopy revealed an essentially normal respiratory tract, with healthy looking respiratory epithelium. No opening in the bronchial wall could be identified. Upon injection of contrast into the right main bronchus the fistulous communication was confirmed (fig. 2).

A right thoracotomy was performed. The pleural cavity contained numerous adhesions from previous pleurisy. A fistulous tract, about 3 cm in length, ran between the mid-oesophagus and the right lower lobe bronchus. There was no surrounding inflammatory reaction, tumour or enlarged lymph nodes. The fistula was excised and the oesophageal and bronchial defects closed in layers.

Histological examination demonstrated stratified squamous epithelium, with surrounding muscularis mucosa, and with no evidence of neoplastic or inflammation processes (fig. 3).

The postoperative course was uneventful and symptoms fully resolved.

Discussion

In adults, fistulae between the oesophagus and the respiratory tract are commonly the result of malignant disease. Non-malignant oesophageal respiratory fistulae are relatively rare, and may be congenital or acquired from trauma [1], or inflammation, such as tuberculosis, syphilis or pulmonary abscess [2]. Congenital fistulae have been described throughout the entire respiratory tree. Congenital tracheo-oesophageal fistulae, without oesophageal atresia, was first anatomically recognized by LAMB [3] in 1873, and described in the adult by NEGUS [4] in 1927. To date, 20 such cases of congenital tracheo-oesophageal fistulae presenting in adults have been described. More distally sited congenital fistulae, between bronchus and oesophagus are more common, and 70 such adult cases have been described previously [5]. As these fistulae are being reported with increasing frequency in recent years, it is likely that cases with this abnormality were not recognized in past years.

When an oesophageal respiratory tract fistula is diagnosed

in the adult, it is not always easy to differentiate congenital origin from acquired origin. The congenital nature of such fistulae is suggested by the clinical, operative and histopathological findings. In the two cases presented, there was no clinical evidence of trauma or inflammation before the onset of symptoms. The endoscopic and contrast studies, together with the operative findings, identified simple fistulae between the oesophagus and the respiratory tract, in the absence of any inflammatory or neoplastic processes. Histological assessment of the excised fistula from Case 2 identified a stratified squamous mucosa, with a surrounding muscular layer; findings proposed by BLACK [6] to be strongly suggestive of a congenital origin.

The two cases differ in the age of first clinical presentation. In Case 1, the symptoms began in infancy and progressively increased with age, whereas in Case 2, symptoms did not begin until the age of 40 yrs, making the congenital nature of this fistula more suspect. However, such delayed presentation of congenital fistulae has been previously well-recognized, with symptoms often first becoming apparent in adulthood. OSINOWO *et al.* [7] observed that congenital fistulae sited in the distal respiratory tract tended to present at a later age compared with fistulae in a more proximal position.

BRAIMBRIDGE and KEITH [8] originally classified congenital broncho-oesophageal fistulae into four types. Type 1 is associated with a wide neck congenital diverticulum of the oesophagus, that perforates into an airway following an inflammatory process. It is difficult to differentiate this diverticulum from the traction type, more commonly seen. Type 2 is a simple tract joining the oesophagus to a lobar or segmental bronchus. Type 3 is a fistulous tract with a cyst interposed between the two hollow viscera. Type 4 is a fistula entering the bronchus of a sequestered pulmonary segment. Case 2 represents an example of a Type 1 fistula.

In the clinical presentation, respiratory symptoms predominate, chronic cough being the commonest manifestation [9]. Other common symptoms include haemoptysis and recurrent pneumonia, which can lead to the development of bronchiectasis [7]. Both Ono's sign, which describes paroxysms of cough following ingestion of liquids, and the presence of food within the sputum are highly suggestive of a fistula [8]. Upper gastrointestinal symptoms of dysphagia and gastro-oesophageal reflux result from disorders of oesophageal motility. These oesophageal motor abnormalities, characterized by unco-ordinated, low amplitude peristalsis, occur in association with congenital oesophageal fistula, and persist despite fistula repair [10]. Epigastric pain, abdominal discomfort and vomiting can result from involuntary aerophagia, as lung air is forced across the fistula into the oesophagus during periods of exertion or coughing [11].

The symptoms may date from childhood, as in case one or may first become manifest in adult life, as in Case 2. To explain this delay in presentation, several mechanisms have been proposed: 1) the upwardly oblique course of the fistulous tract tending to close on swallowing and prevent reflux [12]; 2) the presence of membrane that subsequently ruptures [13]; and 3) a flap valve of oesophageal mucosa that subsequently becomes incompetent. Furthermore, the widespread use of antibiotics may limit the

degree of pulmonary suppuration, allowing the symptoms to present at a later age. Because of the variation in age at presentation and the nonspecific nature of symptoms, other disease processes, such as cystic fibrosis and ischaemic heart disease, are often considered, as in the cases presented. This can result in further diagnostic delays and inappropriate management.

Once suspected, diagnostic confirmation can prove difficult. The plain chest X-ray will typically demonstrate only nonspecific infective changes. However, the presence of air within the oesophagus is highly suggestive of an oesophageal respiratory tract fistula. Oesophageal contrast study remains the most sensitive investigation, particularly when using a tube oesophogram technique with water soluble contrast and the patient in the prone or anterior oblique position. Complete evaluation requires oesophageal and bronchial endoscopy to directly view and biopsy the fistulous opening, particularly if alternative pathology is being considered.

The condition is surgically correctable, and involves interruption of the fistula with resection of any bronchiectatic lung segments. The fistula may be excised or simply stapled, thus avoiding potential mediastinal contamination. In patients unfit for thoracotomy, obliteration of the fistula with topical applications of sodium hydroxide and acetic acid *via* the endoscope has been described [14].

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