

Broncho-oesophageal fistula with vascular malformation

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ABSTRACT: We present a patient with a type III congenital broncho-oesophageal fistula and a connection between the systemic and the pulmonary circulation. The congenital fistula caused chronic bronchopulmonary suppuration with bronchiectasis which in turn was the cause of a left-to-right shunt, probably through multiple precapillary or capillary anastomoses.

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A connection between the oesophagus and the bronchial tree is an uncommon cause of chronic bronchopulmonary suppuration [1]. Sometimes symptoms are only present in adult life [1], though the origin of the connection may be congenital [2]. We present an elderly patient with a broncho-oesophageal fistula and also an anastomosis between the systemic and pulmonary circulation.

Case report

A 64 year old woman presented a thirty-year history of coughing, purulent expectoration, discomfort after swallowing, recurrent episodes of pneumonia of the right middle and lower lobes and of haemoptysis. She lost 10 kg in weight in the two months prior to admission caused by anorexia due to recurrent infectious episodes.

On examination she presented râles at the right base besides generalized minor expiratory wheezing. There was a slight finger-clubbing. Chest X-ray showed volume loss and ill-defined opacifications in the right lung base (fig. 1). A right bronchogram was performed which demonstrated an extra-bronchial collection of contrast medium (fig. 2). A barium swallow revealed a fistula from the oesophagus to a cystic structure in the right lower lobe (fig. 3). Endoscopy of the oesophagus confirmed a fistula with two orifices, macroscopically lined by a normal mucosa. Bronchoscopy demonstrated signs of chronic bronchitis, mainly with pus in the partially collapsed right lower lobe bronchi, but did not visualize the fistula. Aortography showed a prominent bronchial artery (fig. 4a) and an infradiaphragmatic artery branching into the consolidated right lower lobe and anastomosing to the right pulmonary artery (figs. 4b and c). Pulmonary angiography confirmed the absence of anterograde filling of the



Fig. 1. - Posterior-anterior chest radiograph showing, at the base of the right lung, a volume loss, ill-defined opacity and tubular condensations concomitant with bronchiectasis.

right lower pulmonary artery (fig. 4d). Right heart catheterization indicated that the pressure in the main pulmonary artery was within the normal range.

A right thoractomy was performed, showing a fistula 10 mm wide and 25 mm long between the oesophagus and the excavated consolidated area in the right lower lobe. A right middle and lower lobe resection was carried out, and the opening of the fistula to the oesophagus was closed. The postoperative period was



Fig. 2. - Bronchography shows bronchiectasis in the right middle and lower lobe together with a posteriorly located irregular cavity which is in connection with the dilated bronchi.



Fig. 3. - Barium study of the oesophagus reveals a broad fistula between the oesophagus and the cavity (arrowheads).

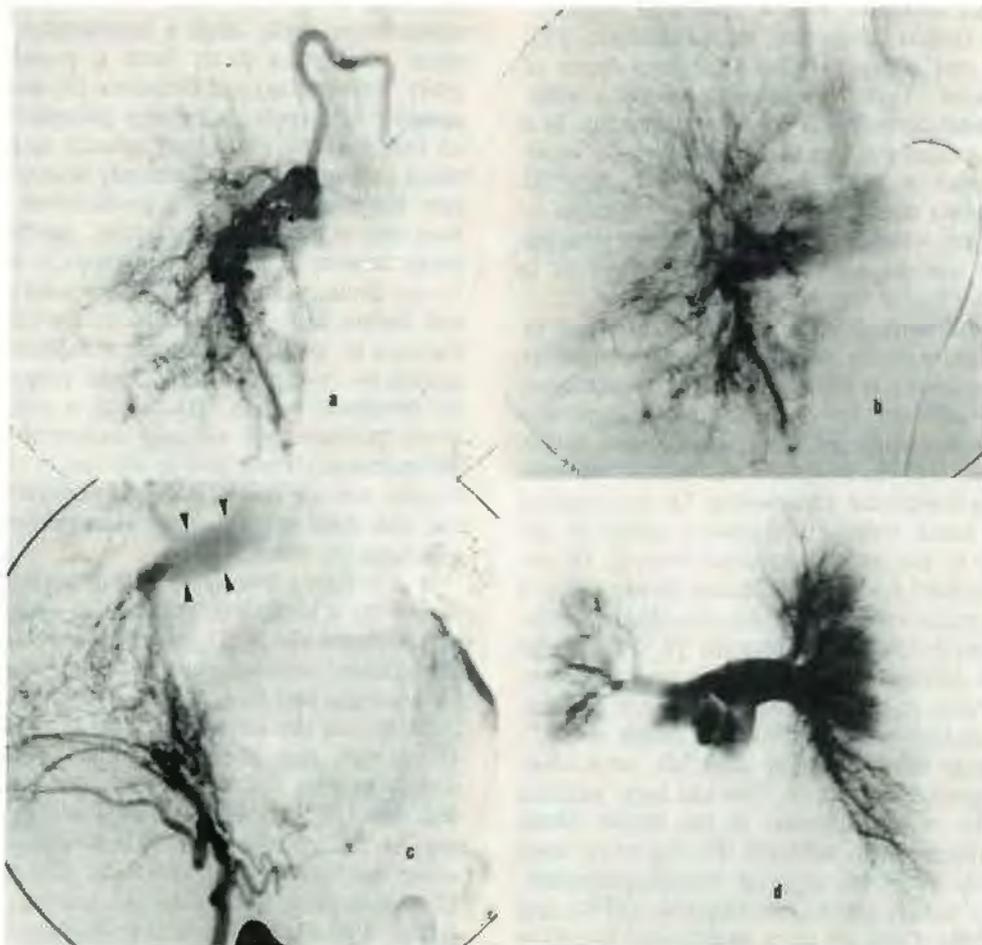


Fig. 4. - Contrast angiography shows: (a) selective opacity of a hypertrophic and tortuous right bronchial artery branching from the thoracic aorta. This bronchial artery branches into several smaller arteries with irregular walls in the region of the lung consolidation; (b) opacity of the right pulmonary artery during the same procedure, suggesting a shunt between this artery and the hypertrophic bronchial arteries; (c) injection of contrast in a hypertrophic subdiaphragmatic artery also shows many tortuous branches and also filling of the right pulmonary artery (arrowheads); (d) pulmonary angiogram, injection into the pulmonary trunk. There is predominant flow to the left lung and to the right upper lobe.

uneventful. The patient then mentioned that it was the first time as far as she remembered, that she did not have thoracic discomfort and cough when drinking with her meals, and she realized that these sensations had not been normal.

The pathological specimen showed that the fistula was lined with a squamous epithelium, and entered a cystic structure in the right lower lobe, which was lined partly with a squamous and partly with a columnar epithelium. The submucosal area was infiltrated by inflammatory cells, partially interrupting the muscularis mucosae.

Discussion

Since her youth this patient had complained of recurring pulmonary infections, cough and discomfort after swallowing, attributable to a broncho-oesophageal fistula with cavitating consolidation in the right lower lobe. In view of the complaints, the endoscopic finding of two oesophageal orifices of the fistula and the pathology of the specimen, we assumed it was a type III congenital fistula. There are only about 30 patients described in the literature with a congenital broncho-oesophageal fistula detected in adult life [3]. BRAIMBRIDGE and KEITH [1] described four types of congenital fistulae. Type I is associated with a wide-necked congenital diverticulum of the oesophagus. In a type II fistula a short track runs directly from the oesophagus to a lobar or segmental bronchus. In type III there is a fistulous track connecting the oesophagus to a cyst in the lobe, which in turn communicates with the bronchus. This was the case in our patient. Type IV is a fistula running into a sequestered segment.

An bronchial-oesophago fistula may be acquired or congenital. If there is no evidence of past or present inflammation around the fistulous track or oesophagus, if there are no adherent lymph nodes and if there is a mucosa and a muscularis mucosae, a congenital nature may be assumed [1]. The lesions in our patient are most suggestive of a congenital abnormality. Of the acquired fistulae, 60% result from a malignancy either of the oesophagus or of the tracheobronchial tree [1]. Of the remainder, one-third result from trauma (oropharyngeal intubation, chemical corrosion of the oesophagus, endoscopy) and two-thirds from infections [4, 5] such as histoplasmosis, aspergillosis, candidiasis, actinomycosis, tuberculosis and syphilis. Other causes include perforations from broncholithiasis and silicotic lymph nodes.

Symptoms may not begin until adult life, even when there is a congenital fistula [4]. This has been ascribed to the presence of a membrane in the fistula which subsequently ruptures [6], although this has never been proven. Usually there are atypical bronchopulmonary symptoms [1]: cough (36%), haemoptysis (17%) and pneumonia (56%). Choking upon swallowing liquids is present in about two-thirds of the patients [1], but it is often so mild that it is only elicited after the diagnosis has already been made, as was the case in our patient. Gastro-intestinal symptoms may also be present: reflux (13%), dysphagia (4%) and epigastric discomfort (13%).

In 65% the diagnosis is made by a barium swallow [1]; yet it is important that during the examination the patient should be placed in the position which causes most of the symptoms. Bronchoscopy and oesophagoscopy sometimes demonstrate the orifices of the fistula. In 35% the diagnosis may only be made at operation for recurrent pulmonary pneumonias and sepsis [1].

In addition, our patient presented an anastomosis between the systemic and pulmonary circulation. This was found on routine aortography, which was performed to demonstrate a possible sequestration. A left-to-right shunt is a common finding in chronic bronchopulmonary suppuration, especially bronchiectasis [7]. Commonly these systemic-pulmonary artery anastomoses occur between bronchial arteries and small pulmonary artery branches. Several hypotheses have been postulated to explain this phenomenon [8]: dilatation of normal precapillary or capillary anastomoses, anastomosis of vessels in granulation tissue arising from both arterial systems, and recanalization of the thrombosed pulmonary artery branches by enlarged vasa vasorum. WEBB and JACOBS [8] stated that "rarely, anastomotic systemic branches arise within the abdomen" and they described three such cases. In our patient the anastomoses arose partly from a hypertrophic infradiaphragmatic vessel and partly from a prominent bronchial artery. ROOSENBURG and DEENSTRA [9] mention that angiography in chronic pulmonary affections often reveals no filling of the pulmonary arterial system of the diseased part of the lung. Similarly in our patient pulmonary angiography showed a predominant flow to the left lung and to the right upper lobe. Another of the diagnostic criteria of left-to-right shunts is a raised arterial wedge pressure in the pulmonary artery to the abnormal region and often a rise in the pulmonary artery pressure as well; furthermore, a definite raised oxygen saturation is often found in the pulmonary artery to the abnormal region. We found a normal pulmonary artery pressure, but we only measured the pressure in the pulmonary trunk and at the origin of the main pulmonary arteries due to technical reasons imposed by the fact that these measurements were performed during the angiographic procedure.

It is believed that systemic-pulmonary artery anastomoses are a protective mechanism against desaturation of peripheral blood [7]. Since an inflamed part of the lung cannot ventilate satisfactorily, bronchopulmonary shunts open and thus permit saturated bronchial blood to flow into the inflamed part of the lung. BOTENGA [7] proved that bronchopulmonary anastomoses can disappear after treatment of the inflammatory process, and therefore can serve as an indicator for the intensity of the inflammation. These anastomoses do not cause any symptoms, and usually they are a coincidental radiological finding in the routine check-up of a patient with chronic bronchopulmonary suppuration.

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RÉSUMÉ: Les auteurs présentent un cas de fistule broncho-oesophagienne d'origine congénitale, type III. Il existe aussi une communication entre la circulation systémique et la circulation pulmonaire. La fistule congénitale a causé une supuration bronchopulmonaire chronique avec bronchiectasies, qui elles mêmes son à l'origine d'un shunt gauche-droit, probablement du à des anastomoses précapillaires et capillaires.

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