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Case report

Congenital broncho-oesophageal fistula: An unusual cause of persistent pneumonia in a young adult

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ABSTRACT

A 19-year-old female with a 3-day history of high temperature, productive cough and dyspnoea was admitted due to diabetic ketoacidosis and pneumonia of the right lower lobe. Antibiotics (amoxicillin-clavulanic acid), insulin, fluids and electrolytes were administered, as appropriate. The patient was doing well (normal temperature, normal glucose levels, normal acid-base balance) until the sixth day of hospitalization, when she reported bouts of cough when swallowing liquids. Barium oesophagography revealed the presence of a broncho-oesophageal fistula (BOF). Congenital BOFs are rare developmental malformations (only just over 100 reported cases in the literature), which are attributable to persistent attachments between the tracheobronchial tree and the oesophagus. When not combined with oesophageal atresia, symptoms may not appear until adult life. History of recurrent respiratory infections, bronchiectasis, haemoptysis and chronic cough associated with eating, may indicate investigation with conventional or multi-positional oesophagography. At the time that surgery was decided, our patient developed persistent pneumonia of both the middle and the lower right lobes. A fistulous tract between the medial segmental bronchus and the oesophagus was removed, along with right middle lobectomy. Post-operative clinical course was excellent.

1. Introduction

The common embryonic origin of the respiratory tract and the oesophagus may rarely give rise to anomalous congenital communications such as tracheo-oesophageal (TOF) or broncho-oesophageal (BOF) fistulas.[1–3] Most cases present in infancy, and are accompanied by oesophageal atresia; however, in the so-called "H-type" fistula, which represents only 3–6% of all cases, the oesophagus is otherwise normal, and the communication exists either to the trachea or less commonly to a bronchus (25% of all cases).[1,4] In these cases, diagnosis may be delayed until childhood or adult life.

Typical symptoms, although non-specific, include recurrent cough, chest pain and dysphagia.[1,3] The so-called "Ohno's sign", defined as a choking sensation when ingesting liquids, has been estimated to be pathognomonic in 65% of patients.[5] Clinical suspicion has to be raised also in patients with recurrent pneumonia and bronchiectasis.[1,2]

This study reports of an interesting case of a congenital BOF in a 19-year-old female, who presented with diabetic ketoacidosis due to a community-acquired pneumonia. Systematic review concerning the epidemiology, aetiopathogenesis, clinical presentation, diagnosis and outcome of BOFs is further discussed.

2. Case report

A 19-year-old female presented in the emergency department with a 3-day history of productive cough, dyspnoea and high temperature of 39 degrees Celsius (°C). Clinical examination revealed a respiration rate of 36 breaths per minute, oxygen saturation of 96% while breathing ambient air, and late inspiratory rales on auscultation of the right pulmonary base. Chest X-ray confirmed pneumonia of the right lower lobe (Fig. 1a). Laboratory investigation, shown in Table 1, revealed, among others, a remarkable hyperglycemia of 392 mg/dl, while pH oxymetry was indicative of diabetic ketoacidosis.

The patient reported having no history of diabetes mellitus or other co- morbid conditions. She was Caucasian in origin, lived in Greece, and reported no recent travelling abroad. She had no pets, never smoked, and did not use systematically any medication. Her

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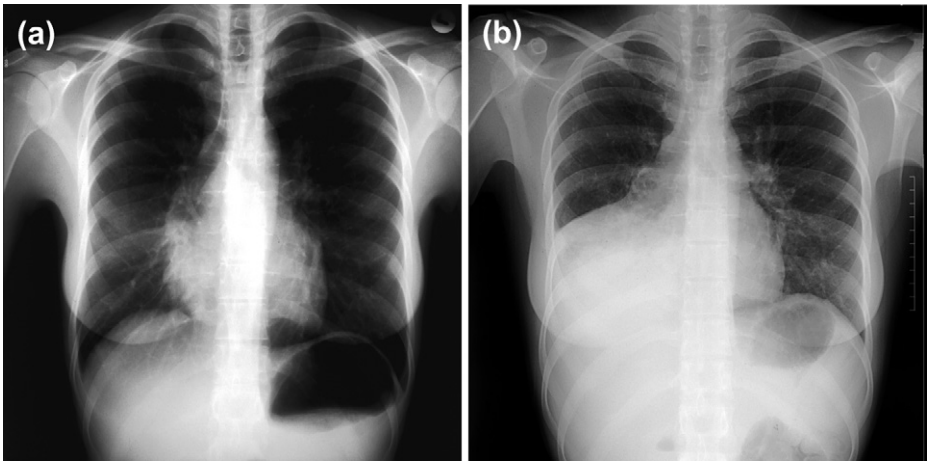


Fig. 1. Chest X-ray (a) on admission and (b) sixth day of hospitalization.

family history was only of a grandfather with a history of type 2 diabetes mellitus.

The patient was admitted to the department of internal medicine for the management of diabetic ketoacidosis triggered by a community-acquired pneumonia, where she initially received intravenously antibiotics (amoxicillin-clavulanic acid), as well as insulin, fluids and electrolytes, as appropriate. Two days later, temperature dropped to 37.7 °C, serum glucose and blood gasses normalized, and the patient began eating on subcutaneously administered insulin.

On the sixth day, recurrence of high temperature with persistence of productive cough prompted new radiographic investigation with chest X-ray (Fig. 1b) and computed tomography (Fig. 2), which both revealed significant increase of the inflammatory infiltrates, with expansion of the bronchopneumonia in both the middle and lower right lobes, as well as accumulation of small amount of pleuritic fluid. Antibiotic treatment changed from amoxicillin-clavulanic acid to piperacillin-tazobactam, in order to cover possible hospital-acquired pathogens, but the patient remained feblile. Simultaneously, the patient reported new onset, worsening cough, and, particularly, bouts of cough when swallowing liquids. This new symptom raised clinical suspicion of a communication between the respiratory tree and the upper gastrointestinal tract. The patient was advised to stop eating, and was subjected to a barium-oesophagography, which confirmed the presence of a BOF (Fig. 3).

The patient was referred to a specialized department of Cardio-Thoracic Surgery, where open right thoracotomy and resection of the fistulous tract, along with right middle lobectomy, were performed. The fistula was about 3 cm long, connecting the right middle segmental bronchus and the lower third of the oesophagus, while the right middle lobe macroscopically resembled liver parenchyma. The histopathological examination reported that the whole lobe removed was atelectatic with evidence of chronic pneumonitis, while medium-sized bronchi were moderately dilated and full of cellular debris. The fistula was covered with squamous epithelial tissue. No evidence of malignancy or granuloma formation was found in the samples taken from the reactively hyperplastic hilar lymph nodes.

Post-operative course of the patient was excellent and the patient was discharged 8 days later.

Table 1
Laboratory data.

Variables*	Ref. range	On admission	2nd day	6th day	Before surgery
White-cell count (/mm^3)	4000–10,500	19,800	10,600	5440	3660
Neutrophils (%)	40–70	83.9	77.3	58.3	63.5
Haematocrit (%)	36–43	42.9	32.5	33.5	29.3
Haemoglobin (g/dl)	12–16	14.4	11.2	11.0	9.7
Glucose (mg/dl)	75–115	392	167	143	119
Na (mmol/l)	135–145	132	136	136	140
C-reactive protein (mg/dl)	<0.8	16.4	6.3	1.9	5.4
pH oxymetry					
pH	7.35–7.45	6.943	7.396		7.404
pO_2 (mmHg)	60–100	106	191		233
pCO_2 (mmHg)	35–45	9	25		28
HCO_3 (mmol/l)	22–28	1.9	17.9		19.8
BE (mmol/l)		−28.3	−9.2		−6.5
Anion gap (mmol/l)	8–12	22.5	6.5		5.5
Lac (mmol/l)	<1	1.1	0.6		0.6

*Measured variables of platelet count, prothrombin time, urea, creatinine, aminotransferases, alkaline phosphatase, total protein, albumin, potassium, calcium, were within normal values, at all times.

3. Discussion

Congenital communications between the respiratory tree and the oesophagus are rare developmental anomalies (1:3000 to 1:4500 live births) resulting from failure of the lung bud to separate

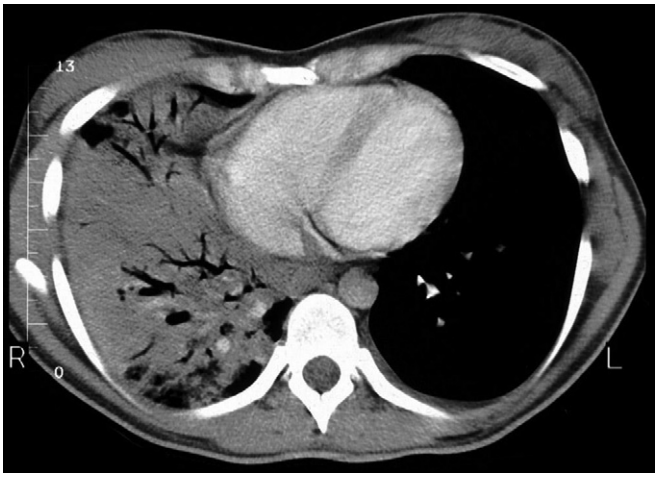


Fig. 2. Computed tomography scan.

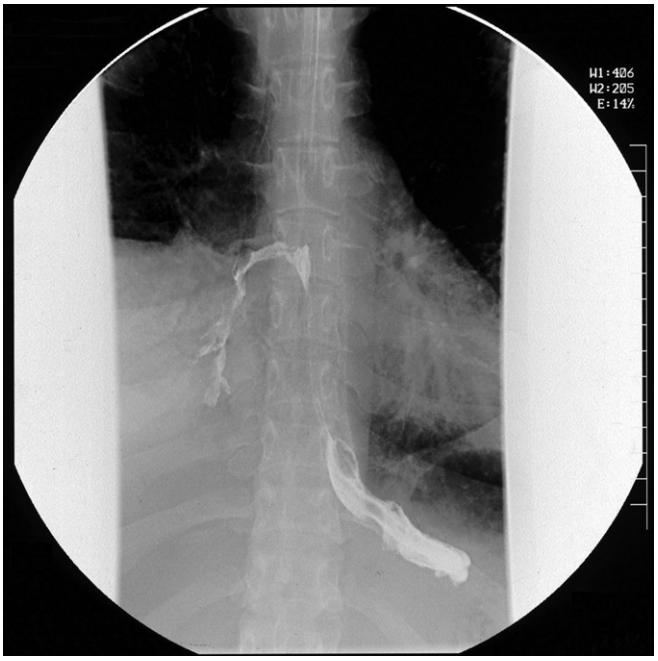


Fig. 3. Barium oesophagography.

completely from the foregut, a procedure that takes place between the 4th and the 6th weeks of gestation.[4] In the majority of cases, this malformation is accompanied by oesophageal atresia, and is typically presented in infancy.[1–4] However, in the so-called "H-type" fistula, which comprises only 3–6% of all cases, the oesophagus is otherwise normal, communicating with either the trachea or a bronchus, formulating either a TOF or a BOF, respectively.[1,2]

3.1. Epidemiology

Congenital BOFs were first described by Negus in 1929[2,5] and were classified into four categories by Braimbridge and Keith in 1965.[3] So far, only about 100 cases have been reported in the literature.[1] Their presentation may be delayed until childhood or adult life, with a median age of 33 years old, while the duration of symptoms can vary from 6 months to 50 years, with a mean of 17 years.[1,2] No sex predominance has been described.[1,2]

The majority (90%) of fistulas are type II according to Braimbridge classification.[5] The communication is usually short and is running directly from the oesophagus to a main or segmental bronchus. In other types, the fistula is communicating with a congenital oesophageal diverticulum (type I), an intralobar cyst (type III) or a pulmonary sequestration (type IV).[5]

3.2. Aetiopathogenesis

Congenital BOFs comprise rare developmental anomalies with an unknown aetiopathogenesis. Presence of a membrane or a tissue fold of the oesophagus which can work as a "flap valve" on the fistulous opening, and spasm of the smooth muscle in the fistula wall are the two most favourable theories that have been developed in order to explain why congenital BOFs can remain silent and asymptomatic for several decades.[1]

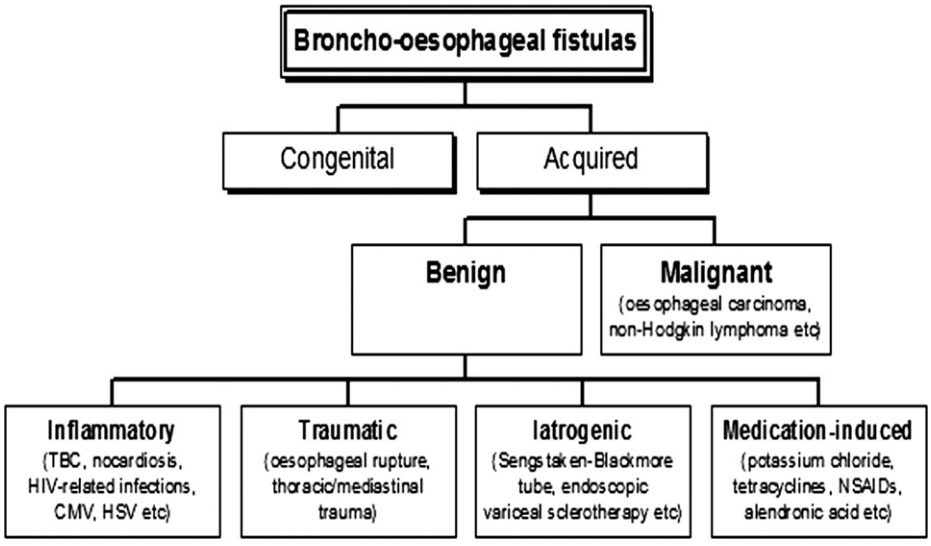
Unlike congenital, acquired forms of BOFs are increasingly encountered when reviewing the literature. Several reports, either as sole cases or as small cohorts of patients, describe broncho-oesophageal communications as the result of malignancy,[6] inflammation,[7–11] trauma,[12,13] iatrogenic procedures[14–16] or drug-induced oesophageal injury.[17] Graph 1 demonstrates a schematic classification of BOFs according to their aetiology.

3.3. Clinical presentation

When asymptomatic, BOFs are accidentically discovered in surgery affecting lobectomy for various reasons.[1] Non-specific symptoms include cough, dysphagia, chest pain and haemoptysis. Paroxysmal and recurrent cough when ingesting liquids ("Ohno's sign") can be pathognomonic in 65% of cases.[3,5] Clinical suspicion should further be raised in patients with recurrent respiratory infections and with non-tuberculous bronchiectasis.[1,2]

3.4. Diagnosis

Conventional barium oesophagography is the most sensitive and most "rewarding" tool in the diagnosis of BOFs.[1,3] In several cases, repetitive multi-positional scans may be necessary for definite diagnosis.[3] Instillation of methylene blue into the oesophagus



Graph 1. Classification of broncho-oesophageal fistulas.

during bronchoscopy, or finding the fistulous orifice during oesophagoscopy can be helpful in establishing diagnosis.[3,5] Nevertheless, as already mentioned, in several cases diagnosis was either made or confirmed intra-operatively.[1]

In all cases, pre-operatively scanning with computed tomography is essential in defining the extent of coexisting pulmonary infection, which may need resection.[1]

3.5. Treatment and prognosis

Thoracotomy with complete resection of the fistula and removal of any permanent co-existing pulmonary lesions is the "gold standard" in the treatment of BOFs.[1,3] Other techniques, mostly indicated in malignant or non-operable cases, are stenting or occlusion of the fistulous tract using specific glues.[18,19]

Despite the benign nature of congenital BOFs, if left untreated, fatal complications are to be expected.[1–3] On the other hand, when successfully and permanently resected, prognosis is excellent.

4. Conclusion

In conclusion, a high index of clinical suspicion for the presence of a possible BOF has to be raised by physicians in cases of unexplained recurrent respiratory infections, non-tuberculous bronchiectasis or chronic cough whenever swallowing liquids. Barium oesophagography is the gold standard in the diagnosis of BOFs, and surgical resection is the preferable choice for a definite treatment.

Conflict of interest

None.

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