Case reports

Congenital oesophagobronchial fistula in an adult involving left main bronchus

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Abstract

Congenital fistulae between the tracheobronchial tree and oesophagus usually originate from the lower end of the trachea or right main bronchus. The case history is presented of a man in whom a fistula between the oesophagus and left main bronchus was not diagnosed until the age of 48.

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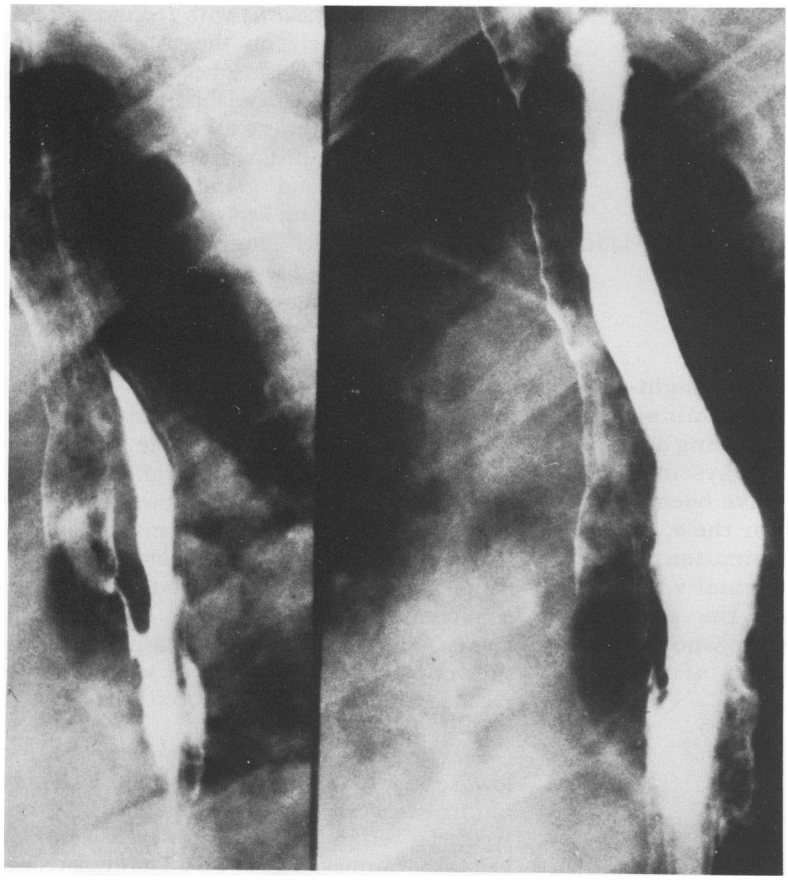

Barium swallow showing a fistulous tract in the middle third of the oesophagus with contrast passing into the left main bronchus.

Tracheo-oesophageal fistula is a congenital malformation which represents less than 1% of oesophageal anomalies[1] and was first described by Lamb in 1873. Congenital fistulae between the airway and the oesophagus are usually diagnosed in the neonatal period.[2] In a quarter of cases, however, they may only be diagnosed in adolescence[3] and occasionally in adult life. Anatomically they connect the oesophagus with the lower end of the trachea or the right main bronchus. Connection with the left main bronchus is very unusual. We present one case of a broncho-oesophageal fistula located between the anterior aspect of the middle oesophagus and the left main bronchus.

Case report

A 48 year old man with a past medical history of regurgitation and dysphagia for liquids since infancy was evaluated because of worsening symptoms over the previous two years. Physical examination was unremarkable. Full blood count, blood biochemistry, chest and abdominal radiograph were normal. A barium swallow showed a hiatus hernia and oesophageal reflux, and endoscopy demonstrated severe reflux oesophagitis. Oesophageal manometry revealed moderate pharyngo-oesophageal incoordination with tertiary waves in the oesophagus and a hypertensive cardia consistent with reflux, confirmed by 24 hour pH monitoring.

The patient underwent a Nissen funduplication and the postoperative period was unremarkable.

Three months later the patient was re-evaluated because of worsening of his preoperative symptoms. He was unable to tolerate liquids due to cough and choking. A barium swallow revealed a fistula between the middle third of the oesophagus and an airway (figure). Bronchoscopy demonstrated a small orifice oozing blood stained material in the left main bronchus 1–2 cm beyond the carina. The patient underwent a right thoracotomy with closure of the fistula and interposition of a mediastinal pleural patch. The postoperative period was uneventful and the patient's symptoms disappeared.

Discussion

Congenital tracheo-oesophageal fistulae are usually diagnosed in the neonatal period because of their frequent association with oesophageal atresia. Only cases without atresia may remain silent until adulthood. Blackburn and Amoury[4] described 260 patients in whom the mean age of diagnosis was 33 years. Our adult case, with no significant symptoms until the age of 48, may be explained by three different

mechanisms: (a) presence of an occluding membrane that subsequently ruptured; (b) presence of an oesophageal fold working as a valve that became incompetent with ageing;[3] (c) progressive slope of the fistulous tract due to the different growth rates for the trachea and oesophagus.[6]

The location of the fistulous opening within the airway is usually the carina (91%).[7] Location in the right main bronchus is observed in 8% of patients. A congenital fistula between the oesophagus and left main bronchus has not been reported in the medical literature.

The diagnostic techniques are a barium swallow and bronchoscopy. In our patient bronchoscopy allowed identification of the fistulous opening. The sensitivity of both diagnostic methods is summarised in the table. Oesophagoscopy has a lower yield than the other two procedures as it can be very difficult to visualise the oesophageal opening. This should always be performed, however, to rule out other causes.[9,10] In some cases, including this one, the orifice of the fistula is hard to identify and hence a Nissen funduplication for gastro-oesophageal reflux is performed. The persistence of the symptoms after surgery led to a repeat barium swallow which demonstrated the lesion.

Percentage accuracy of bronchoscopy and barium swallow in the diagnosis of tracheo-oesophageal fistula as reported by different authors

	Bronchoscopy	*Barium swallow*
Bedard *et al*[1]	66	53
Kirk and Dicks-Mireaux[2]	100	47
Vasquez *et al*[3]	25	100
Johnston and Hastings[7]	50	66
Beaseley and Myers[8]	–	73
Mean	60	68

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Congenital cutis laxa with a dominant inheritance and early onset emphysema

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Abstract

Two cases (mother and daughter) are reported of autosomal dominant cutis laxa which are unusual in being associated with early onset emphysema. Both mother and daughter have been smokers and are heterozygotes for the α_1 antitrypsin genotype. The combination of cigarette smoking and subnormal α_1 antitrypsin levels may explain the pulmonary spread in these two women who have what is usually a benign form of cutis laxa limited to the skin.

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Case reports

CASE 1

A 51 year old woman had been noted at birth to have an abnormal "velvety" skin and had developed the typical features of cutis laxa in her early childhood. By the age of 51 she looked much older than her chronological age. Her skin was lax and inelastic with redundant folds present, particularly on the face, neck, and abdominal wall. Her ear lobes were pendulous and the eyelids and nasolabial folds drooping despite cosmetic surgery to the face in her late teens. Her family history revealed a similarly affected mother and grandmother, both of whom were non-smokers who survived into their eighth decade. There was no family history of respiratory disease or consanguinity.

The patient became progressively dyspnoeic on exercise in her late twenties and was diagnosed as having emphysema. She had smoked 10–15 cigarettes a day from her early teens until diagnosed. Her symptoms progressed until July 1991 when she received a double lung transplant. Pretransplantation spirometry results are given in the table, and the appearance of the chest radiograph before transplantation is shown in the figure. Histopathological examination of the explanted lungs showed extensive emphysema with a scarred and bronchiectatic lingula. She is currently well 12 months after surgery with a forced expiratory volume in one second (FEV_1) of 2·0 l and forced vital capacity (FVC) of 2·3 l.

Pretransplantation investigations revealed a low serum α_1 antitrypsin level of 1·65 mg/l (normal ranges 1·6–2·1 mg/l) and a PI^{mz} genotype. Her serum IgG and IgM levels were also below the normal range at 4·6 g/l (normal range: 6·4–16) and 0·3 g/l (normal range: 0·6–2·8), respectively.