

Short reports

Congenital bronchobiliary fistula: first case in an adult

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ABSTRACT The first adult case of a congenital communication between the biliary tract and the right main bronchus is reported. Treatment by surgical excision and pneumonectomy was successful.

Since 1952, when the first case of a congenital fistula between the respiratory and biliary tract was reported by Neuhauser *et al*,¹ only 14 cases of this malformation have been described.²⁻⁷ All have been in infants or children and the abnormality has led to respiratory distress, with a fatal outcome in some children. Three patients had other biliary malformations.^{2,4,5} The following case is the first to be described in an adult.

Case report

A 32 year old white woman was admitted to hospital in January 1985 because of fever and severe productive cough with greenish yellow sputum. She gave a history of continuous expectoration of golden yellow sputum, which increased greatly after meals. She had had frequent episodes of bronchopulmonary infections since she was two months old and had required two or more courses of antibiotics a year for acute chest infections. On examination she was thin and had the appearance of being chronically unwell. Her temperature was 38°C and she was expectorating large quantities of yellow mucus.

On examination there was chest wall retraction, rhonchi, and crepitations in the right hemithorax. The chest radiograph showed opacification of the right lung and a shift of the mediastinum to this side. The sputum pH was 8.0 and the bilirubin concentration was 2.2 mg/100 ml (37.6 µmol/l). Antibiotics were administered with little improvement. Bronchoscopic examination showed considerable distortion

of the bronchial tree and carinal trifurcation. On chole-scintigraphic examination, technetium-99m labelled diethyl IDA (HIDA) appeared in the right lung projection. Cholangiography via the papilla of Vater showed an anomalous communication between the left biliary duct and right lung.

An elective thoracotomy was performed to excise the fistula and the right lung. The chest was entered through a right posterolateral thoracotomy. There were a few loose

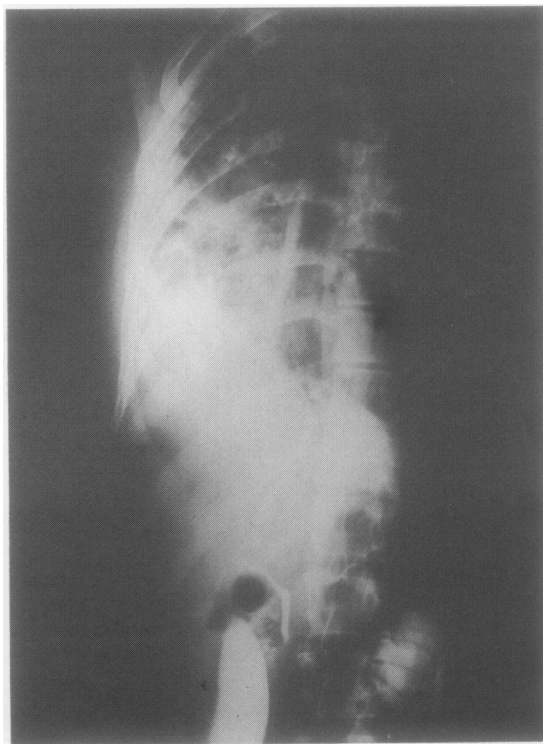


Fig 1 Fistulocholangiogram showing the communication between the left biliary duct and the right anomalous bronchus.

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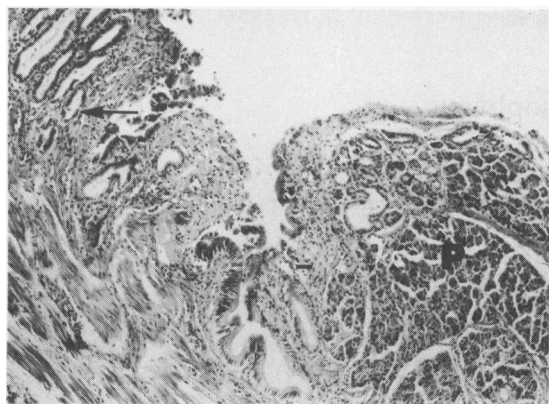


Fig 2 Photomicrograph of the fistular wall showing gastric epithelium (arrowed) and pancreatic tissue (P).

pleuropulmonary adhesions. The lung was totally collapsed and had a fibrotic appearance. The fistulous tract emerged as an anomalous branch from the right main stem bronchus. It was 15 mm wide and had cartilaginous rings throughout its course except for the last 2 cm. The duct entered the abdomen through the oesophageal hiatus, and the intraoperative fistulogram showed free communication with the left hepatic duct (fig 1). There was no other congenital anomaly of the bronchial or vascular supply to the right lung.

Pathological examination showed an end stage fibrotic right lung, with chronic inflammation of the bronchial tree, an increased number of mucus secreting cells, and hyperplasia of bronchi associated lymphoid tissue. Fusiform and saccular bronchiectasis was observed, mainly in the lower part of the right lung. Histopathological examination of the fistula showed normal cartilaginous rings and respiratory epithelium in the first part with areas of squamous metaplasia 4 cm from the right main bronchus, gastric mucosa and pancreatic tissue in the middle part, and biliary epithelium in the last part (fig 2).

The patient's postoperative course was uneventful. She was discharged on the 10th postoperative day and after one year was well, with none of her original respiratory symptoms.

Discussion

Congenital bronchobiliary fistula has been reported exclusively in children, 10 of the 14 published cases having

been in girls. Because of the associated respiratory distress and pulmonary infections the diagnosis previously has always been made at an early age, ranging from 12 hours after birth⁷ to six years.⁸ The malformation causes progressive respiratory dysfunction with a high mortality rate. Five of the children died as a result of the condition, three after surgical treatment. In all but one case the bronchial opening was on the right side of the carina or from the right main bronchus. We are not aware of any cases of this malformation diagnosed in adult life.

The pathogenesis of this anomaly is not clear, and two possible embryological mechanisms have been postulated: (a) fusion of an anomalous bronchial bud with an anomalous bile duct¹; (b) duplication of the upper gastrointestinal tract.¹ The histological finding of gastric and pancreatic tissue in our patient favours the latter hypothesis.

In previous studies the diagnosis has been made by bronchoscopy and bronchography. Our case indicates that non-invasive procedures may also confirm the diagnosis, with determination of bilirubin concentration in the sputum or tracheal aspirate combined with pH measurement and cholescintigraphy with 99mTc diethyl-IDA. The endoscopic fistulogram was also useful, providing an outline of the intrathoracic portion of the fistula to help surgical resection, the major objective in the management of this anomaly.

The pulmonary destruction in this adult case demonstrates the severity of the lesions that bile causes to lung tissue.

References

- 1 Neuhauser EBD, Elkin M, Landing B. Congenital direct communication between biliary system and respiratory tract. *Am J Dis Child* 1952;**83**:654-9.
- 2 Sane SM, Sieber WK, Girdany BR. Congenital bronchobiliary fistula. *Surgery* 1971;**69**:599-608.
- 3 Kalayoglu M, Olcay I. Congenital bronchobiliary fistula associated with esophageal atresia and tracheo-esophageal fistula. *J Pediatr Surg* 1976;**11**:463-4.
- 4 Chan YT, Ng WD, Mak WP, Kwong ML, Chow CB. Congenital bronchobiliary fistula associated with biliary atresia. *Br J Surg* 1984;**71**:240-1.
- 5 Dyon JF, De Marliave H, Bacle B. Les fistules aerodigestives congenitales rares. *Chir Pediatr* 1984;**25**:234-44.
- 6 Chang CCN, Giulian BB. Congenital bronchobiliary fistula. *Radiology* 1985;**156**:82.
- 7 Lindahl H, Nymon R. Congenital bronchobiliary fistula successfully treated at the age of three days. *J Pediatr Surg* 1986;**21**:734-5.
- 8 Cuadros J, Aransay A, Nistral M, Monereo J. Fistula traqueo-biliar congenita. *An Esp Pediatr* 1974;**7**:256-62.