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Surgical treatment of bronchiectasis in Kartagener's syndrome

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SUMMARY

A case of Kartagener's syndrome in which bilateral partial pulmonary resection was performed is reported. The literature concerning the surgical treatment of severe bronchiectasis is reviewed.

THE association of situs inversus and bronchiectasis was first described by Siewert in 1904, but it was Kartagener who, in 1933, published a study of 11 cases which presented the triad with which his name was to be associated: situs inversus, bronchiectasis and sinusitis.

Bronchiectasis and sinusitis are present in 25 per cent of patients who show situs inversus, and in less than 0·5 per cent of the remaining population according to Logan et al. (1965). In the series reported by Adams and Churchill (1937) these figures were 16 and 0·2 per cent respectively.

'Sinusitis' comprises the least distinctive element of the triad. The term 'sinusitis' includes the absence or hyperplasia of one or several sinuses, nasal polyposis as well as simple sinusal infection (Safian and Mandeville, 1959).

Case report

A 25-year-old man presented in March 1974 with a productive cough and moderate haemoptysis. His history revealed that 7 years earlier he had undergone surgical drainage for left maxillary sinusitis. At the age of 21 he had had a left middle lobectomy carried out elsewhere for the treatment of bronchiectasis.

The functional postoperative recovery had been satisfactory, but recurrent respiratory infections had persisted. An angiopneumograph showed the total reversal of the vascular pulmonary tree. A cardiogram did not reveal any abnormality.

On clinical examination there were some coarse râles and wheezing, which were most severe over both lung bases posteriorly, a moderate pectus excavatum and dextrocardia, which was confirmed by a chest X-ray (*Fig.* 1). The rest of the clinical examination was normal. *Haemophilus* sp., *Streptococcus viridans* and *Neisseria* sp. (which were all sensitive to ampicillin) were cultivated from the sputum.

Bronchoscopy was performed and revealed yellowish green sputum in the right basal segments. A bilateral bronchograph showed the residual left middle lobar bronchial stump—the result of the previous operation. It also revealed tubular and bilateral moniliform bronchiectasis ('dead tree' appearance), which was predominantly in the right lower lobe (*Fig.* 2).

X-rays of the face showed aplasia of the frontal sinuses and hypoplasia of the spheno-ethmoïdal sinuses (*Fig.* 3).

Radiological investigation of the upper digestive tract revealed reversed anatomy at the level of the œsophagus, gastric dextroposition (*Fig.* 4) and a stricture of the third part of the duodenum (*Fig.* 5) caused by the mesenteric pedicle. This resulted in slow evacuation of the duodenum.

In view of the recurrent pulmonary infections intensive preoperative treatment was instituted, including antibiotics (based on the data from the sputum cultures), inhalants, postural drainage and chest physiotherapy. Because of a small functional deficiency which was solely restrictive and not

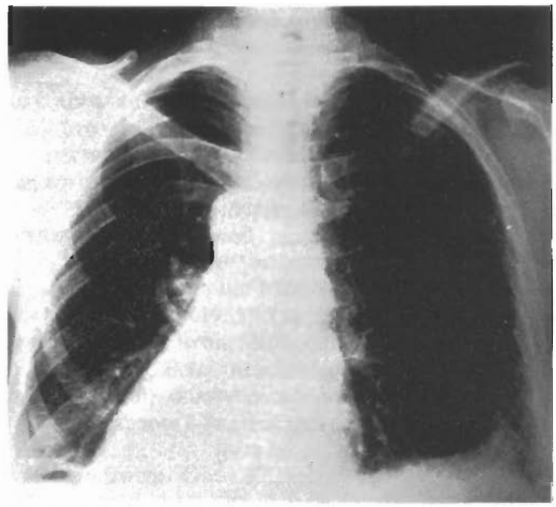

Fig. 1. Postero-anterior thoracic X-ray showing situs inversus.

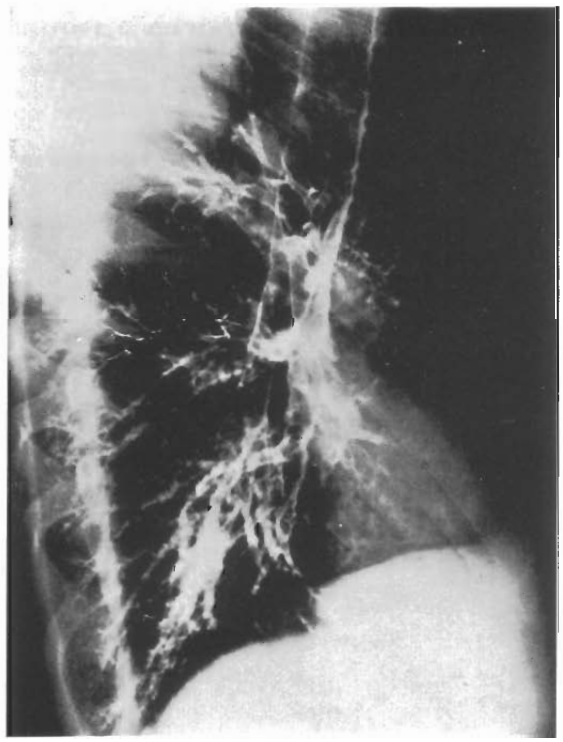

Fig. 2. Right anterior oblique bronchogram demonstrates considerable bronchiectasis at the level of the right base.

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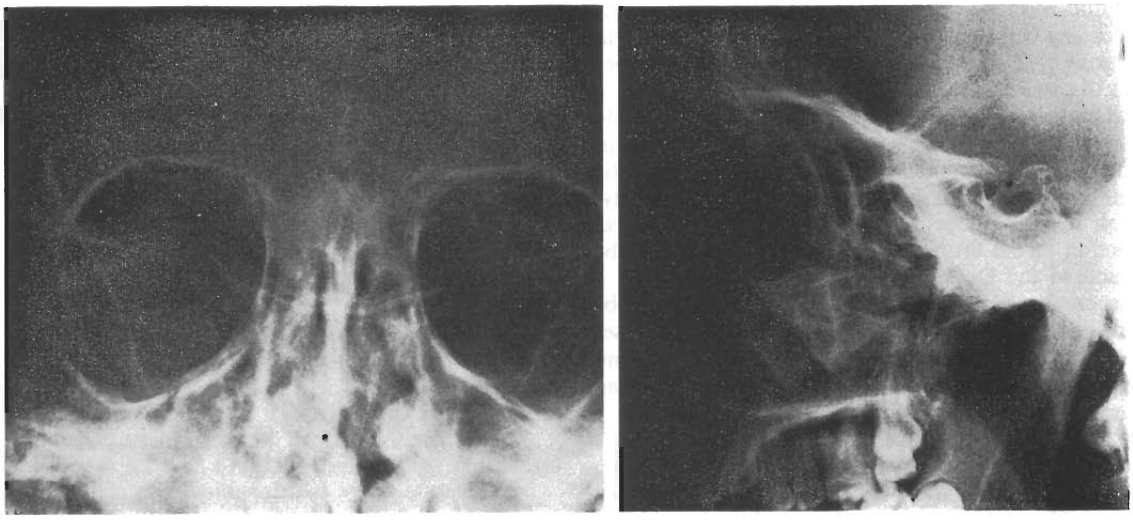

a *b*

Fig. 3. Postero-anterior (*a*) and lateral (*b*) skull X-rays showing aplasia of the frontal sinuses and hypoplasia of spheno-ethmoïdal sinuses.

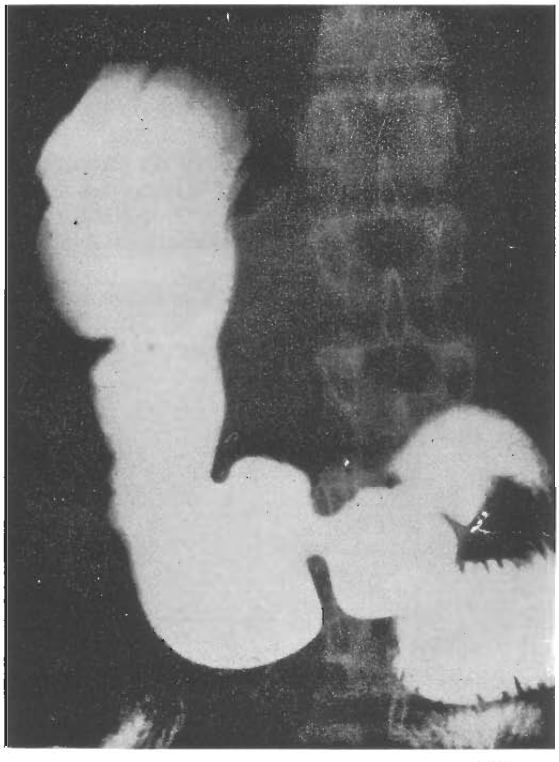

Fig. 4. X-ray of the stomach showing its dextroposition.

affecting gas exchange (vital capacity 86 per cent, forced expiratory volume in the first second (FEV_1) 88 per cent, normal Tiffeneau index), limited surgery was undertaken. This consisted of a right basal resection, leaving the apical segment of the right inferior lobe, as well as lingulectomy (right side in Kartagener's syndrome).

Histological examination of the resected segment showed layers of purulent bronchiectasis, atelectasis and segmental emphysema.

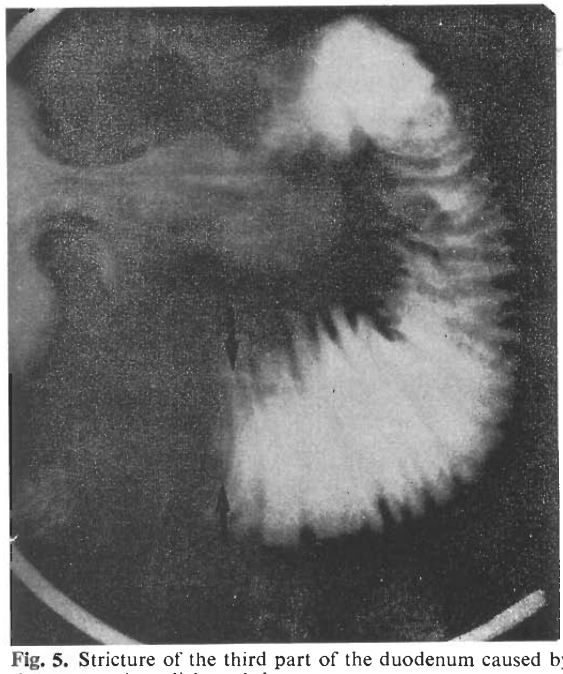

Fig. 5. Stricture of the third part of the duodenum caused by the mesenteric pedicle and the aorta.

The postoperative recovery was short and uncomplicated. At the present time the patient shows no further signs of purulent sputum or a haemoptysis. X-rays and functional tests are satisfactory.

Discussion

The patient reported here had suffered recurrent respiratory infections, sinusitis and bouts of haemoptysis of varying degrees since childhood. Although antibiotics and postural drainage considerably reduce the frequency and severity of infectious complications

of bronchiectasis, surgery is sometimes necessary. According to Logan et al. (1965) and Miller and Divertie (1972), surgery is required in about 30 per cent of cases of Kartagener's syndrome.

In almost all the reported series the commonest site of bronchiectasis is the left lower lobe. The left main bronchus has a smaller diameter compared with the right and crosses the mediastinum more transversely. These factors are probably responsible for the predilection of the bronchiectasis for the left lower lobe (Bradford and DeCamp, 1966).

In confirmed bronchiectasis of the lower left lobe the lingula is also involved in 80 per cent of cases (Perry and King, 1940). According to Bradford and DeCamp (1966), the right middle lobe is diseased almost as frequently.

Remembering that in a case of Kartagener's syndrome the right bronchial anatomy is situated on the left and inversely, in the present patient the most important bronchiectasis was found at the level of the transposed middle left lobe (operation in 1971) and of the transposed inferior right lobe (operation in 1974).

According to Perry and King (1940) bronchiectasis is an illness of the young adult. The symptoms appear in two-thirds of cases before the age of 10 years (Borrie and Lichter, 1965). The bronchiectasis has usually reached its full extent and distribution by the time diagnosis is made (Churchill and Belsey, 1939).

In cases with haemoptysis and extensive bronchial suppuration, which occurs especially in bronchiectasis of the inferior lobes, a curative resection can be envisaged, but only after evaluating the lesions by bilateral bronchography. The extent of the pulmonary resection must be based upon the results of pulmonary function tests (both ventilatory and gas exchange factors). Formerly, palliative interventions, resecting only the most affected area, produced rather unsatisfactory results. At the present time, repeated surgical resections of the more severely diseased segments give better results. This implies preparation for the operation by all the modern methods of bronchial toilet and postural drainage. The good functional result following bilateral partial pulmonary resection for bronchiectasis in the present case of Kartagener's syndrome seems to confirm the value of this approach.

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