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INTRATHORACIC DUPLICATION OF THE FOREGUT

BY

B. T. LE ROUX

From the Department of Thoracic Surgery, Royal Infirmary, Edinburgh

Intrathoracic foregut duplications are amongst the less common cysts and tumours which occur in the mediastinum. The incidence of mediastinal lesions found in a relatively static population of 1,500,000 in the south-east region of Scotland (le Roux, 1959; 1960a and b; 1961a and b) is shown in Table I. In addition to the 17 lesions indisputably of thymic origin, in 19 patients lymphomata were thought also to be of thymic origin. The 105 cysts and tumours were encountered over a ten-year period when 3,000 patients with bronchial carcinoma and 40 patients with bronchial adenoma (Zellos, 1962) were managed in the same thoracic surgical unit.

TABLE I

MEDIASTINAL LESIONS

Neural tumours	Of nerve sheath origin	20	30
	Others	10	
Teratomata			21
Pericardial coelomic cysts			20
Thymic lesions	Cysts	4	17
	Benign tumours	7	
	Malignant tumours	6	
Foregut duplications	Tracheobronchial	9	14
	Posterior mediastinal	5	
Heterotopic mediastinal thyroid			3
			105

Details relating to the 14 examples of foregut duplication are shown in Table II. Of nine tracheobronchial cysts (Cases 1 to 9), seven were chance findings at mass radiography and two patients presented with stridor. All the cysts were right-sided; six were attached to the trachea and three to the right stem bronchus. In the two patients with stridor (Cases 6 and 9) the cysts were attached to the trachea.

Posterior mediastinal cysts in five patients (Cases 10 to 14) were regarded as examples of foregut duplication because of their position, their association with vertebral deformities in three patients, a lining of ciliated columnar epithelium

TABLE II

Case No.	Age	Sex	How Discovered	Side	Size (cm.)	Symptoms	Related to	Lining	Vertebral Deformity	
1	47	F	Mass radiography	R	5	—	Trachea	Ciliated columnar epithelium	—	
2	65	M	,, ,,	R	4	—	,,	,, ,,	—	
3	55	F	,, ,,	R	4	—	Right main bronchus	,, ,,	—	
4	50	M	,, ,,	R	5	—	,, ,, ,,	Stratified squamous epithelium	—	Well-developed muscle wall to cyst
5	24	F	,, ,,	R	7	—	Trachea	Ciliated columnar epithelium	—	
6	56	F	Respiratory symptoms	R	12	Stridor	,,	,, ,,	—	
7	20	F	Mass radiography	R	5	—	,,	Chronic inflammatory tissue	—	
8	24	M	,, ,,	R	7	—	Intermediate bronchus	Ciliated columnar epithelium	—	
9	1	F	Respiratory symptoms	R	7	Stridor	Trachea	,, ,,	—	Well-developed muscle wall and large mucus glands
10	17	F	,,	L	2	Cough and sputum	6th thoracic vertebra	Gastric mucosa	—	Cyst found during left lower lobectomy for bronchiectasis
11	32	F	Mass radiography	R	12	—	Vertebrae	Chronic inflammatory tissue	Cervico-thoracic scoliosis convex to the left	
12	29	M	,, ,,	R	7	—	Oesophagus and upper thoracic vertebra	Ciliated columnar epithelium	—	
13	41	F	At thoracotomy	L	4	—	6th thoracic vertebra	,,	Wedged 6th thoracic vertebra	Cyst found at thoracotomy for mitral stenosis
	54	F	Mass radiography	L	3	—	10th and 11th thoracic vertebrae	,, ,,	Wedged 9th thoracic vertebra	

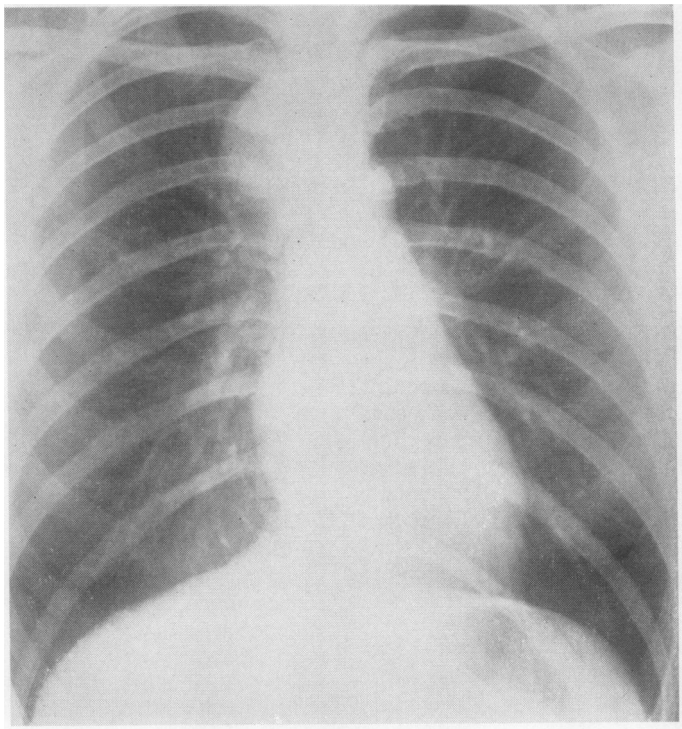

FIG. 1a

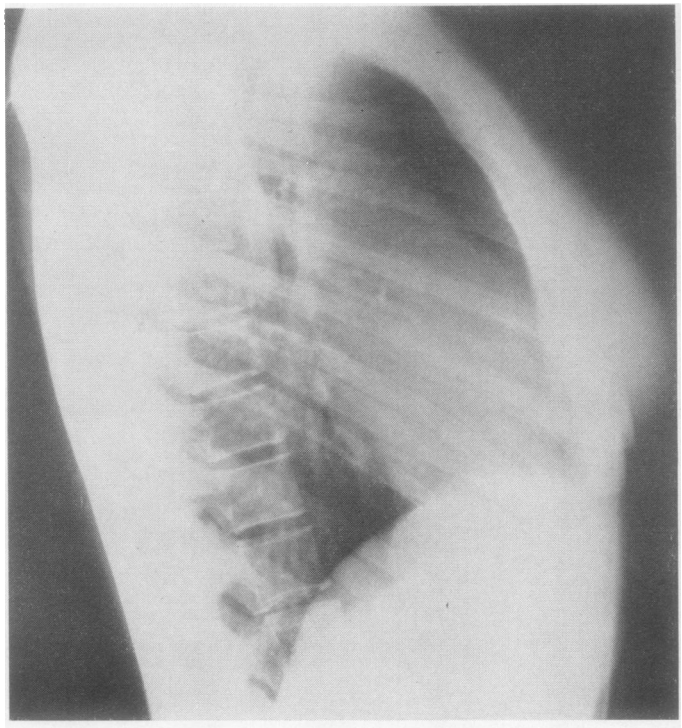

FIG. 1b

FIG. 1. (a) *Postero-anterior and* (b) *lateral radiographs showing a foregut duplication found at mass radiography in a symptomless woman* (*Case* 5).

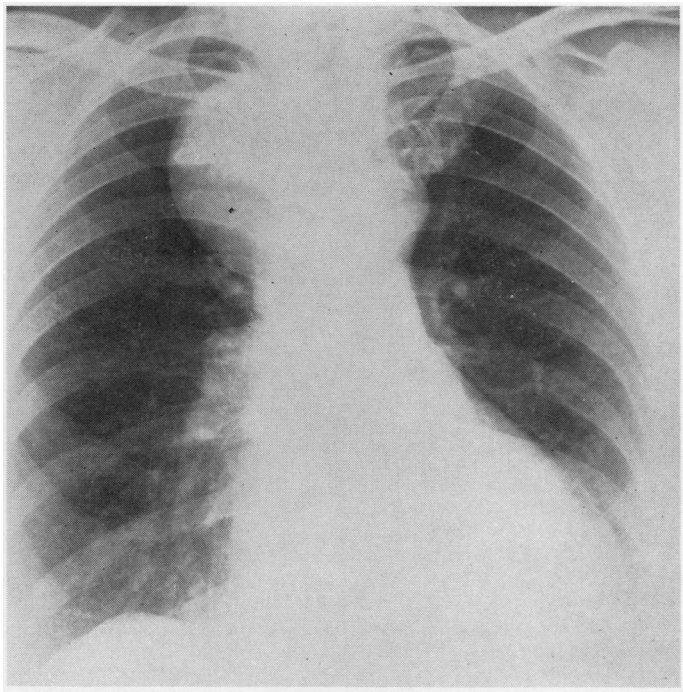

FIG. 2a

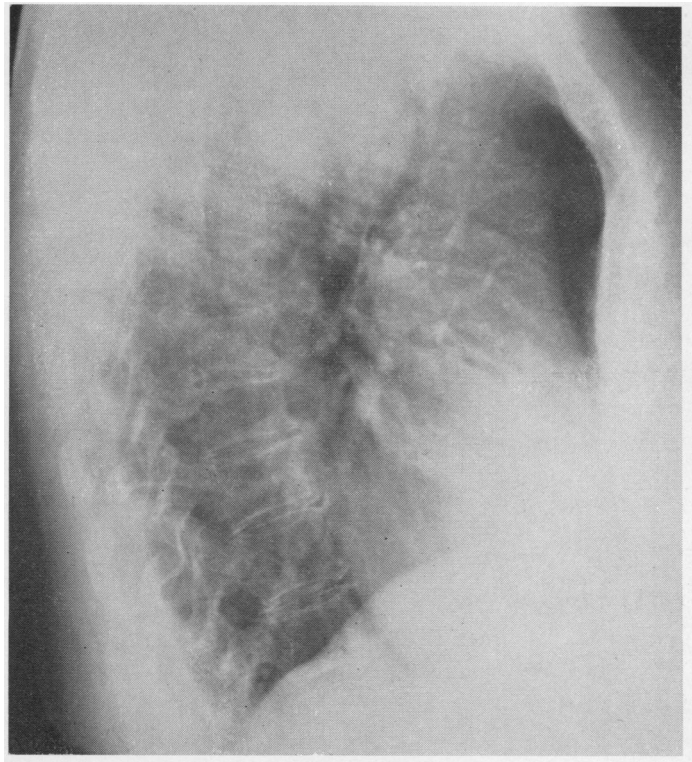

FIG. 2b

FIG. 2. (a) *Postero-anterior and* (b) *lateral radiographs showing a foregut duplication in a woman of* 56 (*Case* 6) *who presented because of stridor.*

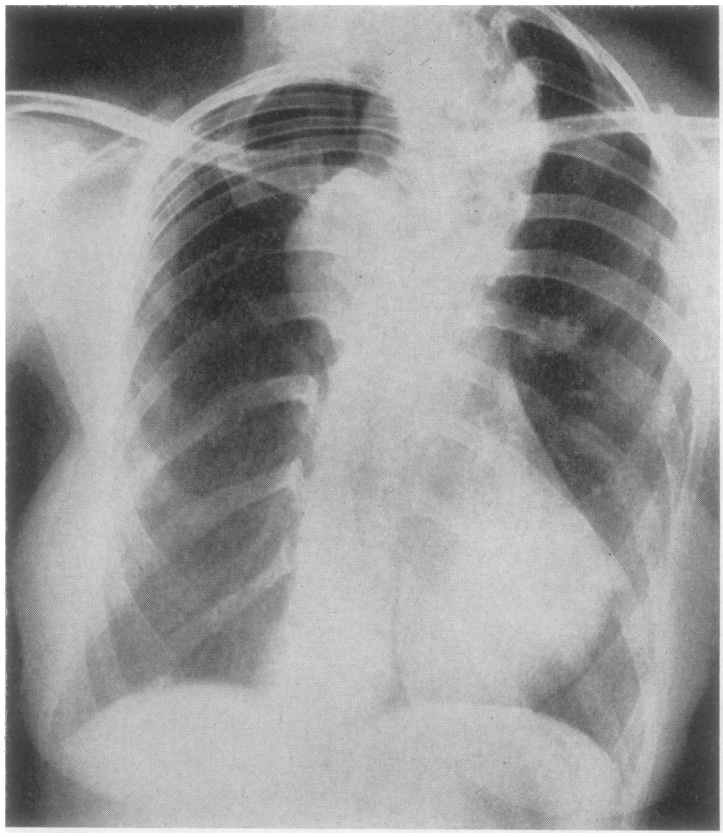

FIG. 3. *A foregut duplication associated with vertebral deformity.*

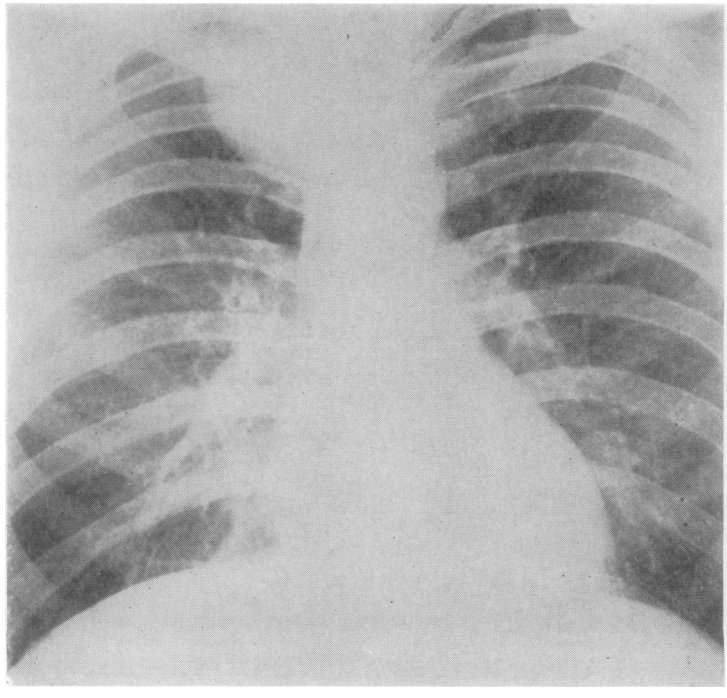

FIG. 4. *A foregut duplication without vertebral deformity.*

in one, and in one a lining of gastric mucosa. This last-mentioned patient was the only one in the series with symptoms; the other four cysts were chance findings, at mass radiography in three and at thoracotomy in one. The patient with symptoms (Case 10) was shown to have left lower bronchiectasis as the cause of respiratory symptoms of many years' duration. In infancy she had had an episode of haemorrhage, either haematemesis or haemoptysis, for which a cause was not found and after which she had recurring respiratory infections. At left thoracotomy a shrunken lower lobe was adherent to a 2 cm. cyst (not recognized on pre-operative radiographs) in the posterior mediastinum. The cyst was densely adherent to the sixth thoracic vertebra and was lined by gastric mucosa. This is believed to be an example of peptic ulceration in a foregut duplication with pulmonary erosion in infancy producing haemorrhage and subsequent pulmonary destruction from infection and possibly from digestion.

Examples of tracheobronchial cysts, one (Fig. 1) a chance radiographic finding and one (Fig. 2) in a woman of 56 with stridor, and of posterior mediastinal cysts of foregut origin with (Fig. 3) and without (Fig. 4) vertebral deformity are illustrated.

Discussion

Alimentary duplications are of two types, those which result from a localized aberration during relatively late development of the gut wall (Forshall, 1961) and those which develop early as part of a more extensive abnormality which has been called the split notochord syndrome (Feller and Sternberg, 1929). The respiratory system develops as an outgrowth from the ventral surface of the primitive foregut; intrathoracic foregut duplications of the first type may be therefore in the wall either of the oesophagus or of the trachea and major bronchi. Bremer (1944) has explained the frequency of cysts at the tracheobronchial junction on the ground of the development of the trachea as a separate entity from the primitive oesophagus rather than as part of the original lung bud, an area of differentiation being always a more common site of congenital malformation. Maier (1948) has classified tracheobronchial cysts as paratracheal, carinal, hilar, para-oesophageal, and miscellaneous, a classification made without reference to the two varieties of foregut duplication and therefore probably untenable. In the present series there were nine tracheobronchial cysts but none of this category within the wall of the oesophagus. The cysts may be either intra- or extra-luminal, are lined by entodermal epithelium, and appear to have developed within the circular muscle coat of the parent tube. The lining is usually ciliated columnar epithelium but may occasionally be stratified squamous epithelium (Morrison, 1958), and mucous glands and hyaline cartilage may also be found (Schlumberger, 1951). The occasional occurrence of heterotopic epithelium is explained by earlier initiation of the abnormality at a time when the cells of the primitive entodermal tube have the potential to develop into any type of intestinal or respiratory epithelium. Cysts in the oesophageal wall may occasionally communicate with the parent organ (Higgins, 1959), and some examples of cylindroma (Bergmann and Charnas, 1958) and oat cell carcinoma (McKeown, 1952) in the oesophagus are believed to have their inception in rests which have differentiated as respiratory epithelium.

Duplications which are part of the split notochord syndrome are commonly associated with vertebral abnormalities and may in addition be associated with abnormalities of the spinal cord and of the overlying dorsal covering including the skin. Fallon, Gordon, and Lendrum (1954), Beardmore and Wiglesworth (1958), Bentley and Smith (1960), and Forshall (1961) have described in detail the mechanisms of the development of this syndrome. The abnormalities are determined at the blastocyst stage of development. They are the result of varying degrees of adhesion between the entoderm of the yolk sac and the ectoderm of the future neural plate, which will prevent development of the notochord in its normal midline position. The ectodermal cells from which the notochord develops are displaced to either side or split so that two notochordal centres result. This is the developmental basis of the congenital scoliosis or hemivertebra so commonly associated with duplications of this type. The duplications all show a similar structure. They are thick-walled with two muscle coats, the fibres of which lie at right angles to each other, muscularis mucosae, Meissner's and Auerbach's plexuses, and a lining of epithelium of entodermal origin. Adhesion to ectoderm results in a diverticulum of entoderm from the parent layer. This may develop as a cyst in close relation to the associated vertebral anomaly; the cyst may be drawn caudally with growth and descent of the foregut and come to lie at a level in the mediastinum some distance caudal to the vertebral anomaly; the vertebral anomaly may be extensive and the cyst may communicate through a spina bifida with the dorsal skin surface; a

thoraco-abdominal duplication may result when the caudal attachment of the diverticulum is more distal with the resulting duplication traversing the diaphragm or passing through the same gap as the oesophagus, a tubular rather than a spherical duplication (Smith, 1960): and posterior mediastinal cysts may occur in association with mesenteric duplications and may be multiple.

Posterior mediastinal cysts are seldom attached to the oesophagus and frequently attached to the vertebral body, and the vertebral deformity is usually upper dorsal or cervico-dorsal. Veeneklaas (1952) has suggested that caudal traction by the cyst on the vertebra to which it is attached is responsible for the vertebral deformity, but this is unlikely since there are many examples of a posterior mediastinal cyst directly applied to the anomalous vertebra. A strand occasionally runs from the cyst dorsally between hemivertebrae. Peptic ulceration with perforation into the lung or the mediastinum may occur.

It has been suggested (Fallon *et al.*, 1954) that when the split notochord syndrome is more generally recognized a careful search for vertebral anomalies in patients with intrathoracic foregut duplications will probably reveal vertebral abnormality with a greater frequency than has so far been reported. Certainly when a child is found at laparotomy to have a duplication of the gut in the mesentery, either spherical or tubular, it is necessary to search radiographically for a vertebral anomaly and for mediastinal foregut duplications. It is notoriously difficult in an infant to differentiate between haemoptysis and haematemesis, and mediastinal ectopic gastric mucosa may be a source of bleeding in infancy. A child with a congenital vertebral anomaly of any sort belongs to a group in which an abnormality of the foregut either in the mediastinum or in the abdomen is likely.

SUMMARY

Fourteen previously unrecorded examples of foregut duplication in the mediastinum are presented. The embryological facts relating to this abnormality are briefly discussed and the relationship between both mediastinal and intra-abdominal duplications of the foregut and the split notochord syndrome is emphasized.

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