Total oesophageal duplication associated with dextrocardia and situs inversus

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ABSTRACT A case of complete oesophageal duplication in association with dextrocardia and situs inversus is presented.

Enteric duplications are rare. Oesophageal duplications are the second most common but their variable presentation may make the diagnosis difficult before surgery. We describe a case of this condition in association with situs inversus and discuss its embryology, incidence, pathology, and treatment.

Case report

A 21 year old man presented with a 10 day history of left sided pleuritic chest pain and purulent sputum. He had vague abdominal pain and had been vomiting for three days. His only significant past medical history was the recognition of dextrocardia and situs inversus during an admission for an appendicectomy.

On examination he was pale, restless, febrile (38°C), and tachypnoeic but not cyanosed. Examination of the chest showed dextrocardia and signs of consolidation at the left base. The white cell count was $16.8 \times 10^9/l$ (normal range $4.0-11.0 \times 10^9/l$). A diagnosis of pneumonia was made and he was treated with antibiotics. A chest radiograph showed a fluid level in the posterior mediastinum on both anteroposterior and lateral films, patchy shadowing at the left base and wedging of the seventh thoracic vertebra. A diagnosis of achalasia was considered. A barium swallow showed a normal oesophagus with the fluid level lying posterior to and separate from the oesophagus (fig 1). Computed tomography showed dextrocardia, situs inversus, and a large air containing space posterior to the trachea (fig 2), suggesting the possibility of a neuroenteric cyst or oesophageal duplication.

The patient was transferred to the regional cardiothoracic unit, where he was noticed to have developed a hoarse voice, suggesting recurrent laryngeal nerve compression. After oesophagoscopy and bronchoscopy he underwent surgery. The chest was opened via a left posterolateral thoracotomy and entered through the seventh intercostal space. A complete oesophageal duplication was found to enter the stomach separately through the anterior part of the hiatus. This was divided and mobilised as far as the left sided arch of the azygos vein. A further third space thoracotomy allowed the duplication to be traced to the junction of the crico-

pharyngeus and inferior pharyngeal constrictor, where it was transected and resected. The postoperative period was uneventful and he was discharged home on the 10th day.

Histopathological examination showed the specimen to be 19 cm in length with squamous epithelium superiorly and columnar epithelium inferiorly. There were two sets of smooth muscle arranged in a circular and longitudinal manner.

Discussion

Oesophageal duplications are the second most common form of enteric duplication. They are tubular or cystic structures of variable size, which arise from the posterior mediastinum and lie in a retropleural location.

The embryology of these duplications is still not understood and though many theories have been put forward none has fully explained the various anatomical patterns and associations. In 1944 Bremmer suggested that during the sixth to seventh week of intrauterine life the intestine goes through a diverticulum and in association with dextrocardia and situs inversus this diverticulum could be a neurenteric cyst or oesophageal duplication.
through a “solid stage,” during which the lumen is obliterated by an accumulation of epithelial cells. Vacuoles then appear in this solid cell mass and coalesce to form longitudinal rows parallel to the long axis of the gut. The walls of these rows finally break down to establish the definitive bowel lumen. Bremmer suggested that abnormal persistence of some vacuoles and failure to coalesce in a transverse manner may lead to a new channel parallel to the original lumen, which then separates from the latter through the union of intestinal layers. As the duplication develops within the intestine the outer wall of the duplicate portion contains all the tissue layers of the gut.

Another commonly accepted hypothesis, the notochordal theory of Veeneklass for mediastinal lesions, provides an explanation of the common association of mediastinal duplications with vertebral anomalies. It suggests that the duplication is formed by herniating of the primitive notochord through the endoderm around the third week of intrauterine life. This causes traction on the primitive gut so that a pouch of gut is drawn towards the vertebral bodies. Further loss of continuity between the pouch and the primitive gut results in a duplication.

Various associated abnormalities have been described, such as gastric heterotopia and vertebral abnormalities, including anterior spina bifida, hemivertebrae, and vertebral wedging. But to our knowledge no case has been described in association with dextrocardia and situs inversus. Malignant change has been reported on several occasions with enteric duplications, especially with those of the large bowel, but not as yet with oesophageal duplication.7

Seventy-five per cent of oesophageal duplications present in early childhood or infancy.8 Their presentation is highly variable, making diagnosis difficult. Presenting symptoms include dysphagia, dyspnoea, superior vena cava obstruction, hoarseness, and cardiac arrhythmias, all of which may be attributed to dilatation of the duplication. Gross dilatation may lead to rupture and mediastinitis. Other forms of presentation include chest pain, respiratory infections, haematemesis, and melaena, resulting from infection and haemorrhage within the duplication.

The criteria for diagnosing an enteric duplication are: (1) the duplication must be connected to the digestive tract; (2) it must have a mucus membrane of digestive tract type; (3) it must have a wall of smooth muscle running in two separate directions. Most duplications are located in the middle third of the oesophagus (60%), the rest being divided between the upper and lower thirds.1

The differential diagnosis on presentation includes achalasia, neuroenteric cysts, and other mediastinal tumours. Surgery may not confirm the diagnosis because many are unrecognisable and confirmation may be by histology alone. Computed tomography is probably the most useful investigation for confirming the diagnosis, supplemented by barium studies and ultrasound. Technetium pertechnetate scanning may also be used where gastric heterotopia is suspected.8

Surgery is undertaken because of the possible complications of bleeding, perforation, respiratory obstruction, and difficulty in differentiating the mass from a posterior mediastinal malignancy.9 It used to consist of marsupialisation and mucosal stripping, but this resulted in infection, fibrosis, scoliosis, and even death. Today complete excision through a transthoracic approach is the procedure of choice.10

This case is unusual in that the association with dextrocardia and situs inversus has not been described before; it suggests the possibility of a common embryological abnormality.

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