Rare association of pulmonary artery sling with tracheo-oesophageal fistula and patent ductus arteriosus

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Abstract

Congenital tracheo-oesophageal fistula (TEF) is a life-threatening complication caused by communication between the oesophagus and the tracheobronchial tree within the neck or the thorax. TEF without oesophageal atresia is commonly known as 'H'-type TEF, and it is extremely rare in infants, accounting for ~2–4% of all congenital tracheo-oesophageal malformations. Here, we report a rare case of a pulmonary artery sling with TEF and patent ductus arteriosus.

Keywords: Pulmonary artery sling • Patent ductus arteriosus • Tracheo-oesophageal fistula

Congenital tracheo-oesophageal fistula (TEF) is a life-threatening complication caused by communication between the oesophagus and the tracheobronchial tree within the neck or the thorax. TEF without oesophageal atresia is commonly known as 'H'-type TEF, and it is extremely rare in infants, accounting for ~2–4% of all congenital tracheo-oesophageal malformations [1, 2]. Here, we report a rare case of a pulmonary artery sling with TEF and patent ductus arteriosus (PDA).

An 8-month old boy was admitted to our department because of stridor for 1 month, repeated dyspnoea for 2 weeks and fever for 2 days. On admission, his body temperature was 39.2°C, pulse rate was 168 bpm, respiratory rate was 45 breaths/min and blood pressure was 95/62 mmHg. Physical examination revealed wheezes. On auscultation, rales were heard in both the lungs and a continuous murmur was heard in the precordial region. Chest radiography showed pulmonary oedema, and transthoracic echocardiography showed the left pulmonary artery (LPA) coursing posterior to the trachea and the presence of PDA. Therefore, multidetector computed tomography was performed for diagnosis (Fig. 1). TEF (Fig. 2) was discovered by chance on the left posterolateral wall.

General anaesthesia was induced. Cardiopulmonary bypass was established using a single venous cannula with a perfusate temperature of ~32°C. After ligation of the PDA, LPA was cut away from the right pulmonary artery, and the defect in the latter was closed with two rows of 5-0 polypropylene sutures. The LPA was then pulled out into the left pleural space. A large window was made in the pericardium behind the phrenic nerve and alongside the pulmonary trunk, and the LPA was brought into the pericardial space through it. An incision was made in the left lateral aspect of the pulmonary trunk, and its proximal end was anastomosed to the side of the pulmonary trunk with continuous 6-0 polypropylene sutures. Then, TEF was ligated. The surgery was successful; the patient was extubated 2 days after the surgery. The patients was successfully discharged to his home 10 days after the surgery. The follow-up period was uneventful.

H-type TEF accounts for 4–5% of all congenital tracheo-oesophageal malformations. The clinical features are variable; however, the common presentations include recurrent respiratory symptoms, aspiration during feeding with cyanosis and abdominal distension. Early diagnosis of this disorder is difficult, and some patients may remain undiagnosed until late infancy or childhood. The first surgical repair of such a defect was reported by Imperatori in 1939 [3, 4].

A pulmonary artery sling is frequently associated with the presence of complete cartilaginous rings in the distal trachea. Cardiovascular anomalies have been reported in 60–80% of patients with the ring–sling complex [5]. However, the association of the pulmonary artery sling with TEF and PDA is rare and special.

Figure 1: Multidetector computed tomography image showing the left pulmonary artery (the red arrow) passing posterior to the trachea.
The LPA sling passing posterior to the trachea worsens the patient’s lung ventilation condition. Adding to the patient’s woes, the existing PDA results in a left-to-right shunt that draws a considerable quantity of blood into the lungs, rendering the patient more prone to infection. TEF may present as chronic lung disease of unknown origin, because repeated aspirations can lead to recurrent lung infections and bronchiectasis. All the three entities encountered in this patient compromise the respiratory system. To the best of our knowledge, this is the first report of a pulmonary artery sling with associated TEF and PDA.

Conflict of interest: none declared.

REFERENCES