Esophageal Atresia with Tracheoesophageal Fistula Associated with Situs Inversus Totalis

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Abstract Esophageal atresia with tracheoesophageal fistula with situs inversus totalis is an extremely rare association. We are presenting a case of a preterm neonate suffering from similar condition. Fistula ligation with primary repair was done via left thoracotomy to avoid technical difficulties. Pre-operative echocardiography should be done to confirm dextrocardia and disposition of the aortic arch.

Keywords Esophageal atresia · Situs inversus totalis · Preterm neonate · Left thoracotomy

Description

A 6-day-old female preterm baby with a body weight of 1.8 kg was admitted with features of pneumonia and frothing from the mouth since birth. The attending pediatrician first suspected it to be a case of pneumonia and so treated the baby with intravenously (IV) administered antibiotics. But due to the presence of persistent vomiting with frothing from the mouth and respiratory distress, the pediatrician suspected the diagnosis of esophageal atresia (EA), and so the baby was referred to our institute. X-ray of the chest and abdomen with a 10-French red rubber catheter showed arrest of the tube in the esophagus at the level of the upper border of the T5 vertebra. The presence of gas shadows in the abdomen suggested the presence of tracheoesophageal fistula (TEF). X-ray picture also showed dextrocardia with stomach bubble on the right (S) and liver shadow (L) on the left side of the abdomen

and liver shadow on the left side, suggesting the presence of situs inversus totalis (SIT, Fig. 1). After stabilizing the patient with IV administered antibiotics and nebulization, the baby was scheduled for open left thoracotomy via the fourth intercostal space. On exploration, ligation and division of the distal

Fig. 1 Radiograph of the chest and abdomen. Note the arrest of the red rubber catheter at the upper border of the T5 level (arrow) with dextrocardia and the stomach bubble on the right (S) and liver shadow (L) on the left side of the abdomen

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TEF with primary repair was done with placement of an intercostal chest drain.

More than 50 % of children with EA have other congenital anomalies [1]. EA with TEF associated with SIT is an extremely rare association [2, 3]. The incidence of SIT reported in the literature is 0.01 % [4]. In such cases, left thoracotomy is preferable to avoid the technical problems during primary repair after confirming dextrocardia and disposition of the aortic arch preoperatively by doing echocardiography. The presence of other associated anomalies should also be looked into. Finally, for knowing the long-term outcome, clinicians must be aware that SIT occurs randomly in 50 % of patients with primary ciliary dyskinesia (PCD). Delayed complications of EA can potentially exaggerate PCD and Kartagener's syndrome [5].

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