A term male baby, vaginally delivered, with birth weight of 2.8 kg presented on D3 of life with provisional diagnosis of congenital pneumonia in view of opaque right hemithorax. There was no significant antenatal event. On presentation, there was marked respiratory distress and excessive frothing from mouth. The orogastric (OG) tube could not be passed beyond 10 cm. There was no audible breath sound over right hemithorax. Chest X-ray showed opacification of right hemithorax with hyperinflated left lung and curved OG tube was visible. (Figure 1) CECT chest was done which showed similar findings. Baby underwent surgical repair where diagnosis of absent right lung and type c tracheoesophageal fistula (TEF) were confirmed. With right posterolateral thoracotomy approach, fistula was ligated and divided. Esophago-esophageal anastomosis was performed. Baby was kept on mechanical ventilation, but could not be weaned off and succumbed on D6 of life.

Figure 1: Chest X-ray showing opacification of right hemithorax with hyperinflated left lung and curved orogastric tube

The association of unilateral lung agenesis with esophageal atresia and tracheoesophageal fistula is extremely rare with a very lethal outcome. (1)

Agenesis of lung is believed to be caused by failure to maintain development balance between the two lung buds. Congenital underdevelopment of the lungs was classified by Schneider. (2) Combination of TEF with pulmonary agenesis had been universally fatal until 1985 when first documented case of long term survival was reported. (3) Since then around 16 cases have been reported to survive beyond neonatal period. Verma et al reported a case who is surviving till 10 yrs of age for the same condition after surgery. (4)

Conflict of Interest : None

References :