CASE REPORT

Left Pulmonary Agenesis: Report of a Case

Sudhir Singh*, Jiledar Rawat, Gurmeet Singh

Department of Pediatric Surgery, King George’s Medical University, Lucknow, Uttar Pradesh, India


ABSTRACT

Pulmonary agenesis is a very rare condition with poor prognosis. Although the condition is a congenital malformations, delayed presentation is also seen. A female neonate with left pulmonary agenesis presented to us with severe respiratory distress. She was initially misdiagnosed as a case diaphragmatic hernia at another hospital and referred to us for further management. Clinical examination and further investigations confirmed the diagnosis of left pulmonary agenesis. The child improved on medical management and is asymptomatic up to 6 months of follow-up.

Key words: Neonate; Pulmonary agenesis; Respiratory distress

INTRODUCTION

Pulmonary agenesis is a very rare condition with an incidence of 1:15,000 autopsies [1]. Lung agenesis or aplasia can be divided into three types: Type 1 with complete absence of lung, bronchus, and vessels (agenesis), Type 2 has a rudimentary bronchus with complete absence of lung parenchyma (aplasia), and Type 3 is manifested by the presence of variable amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature (hypoplasia) [2]. Bilateral condition is incompatible with life. Unilateral pulmonary agenesis is symptomatic in early infancy and more than 50% of babies die before their fifth birthday; however, incidentally diagnosed adult cases have also been available in the literature [2].

CASE REPORT

A female newborn weighing 2.2 kg, born vaginally in a peripheral hospital to a 28-year-old para 2 mother, was referred to our department with a diagnosis of left congenital diaphragmatic hernia. She had severe dyspnea, tachypnea, and peripheral cyanosis at arrival. There was nothing significant in antenatal history other than the history of prolonged labor. Two antenatal ultrasonograms in second and third trimester had failed to pick up any congenital anomaly. On examination, there was no external chest wall deformity. Air entry was absent on the left side, while the right side of chest had mild coarse crepitations; apex beat of the heart was shifted toward left. Plain X-ray chest showed scoliosis, homogenously opaque left hemithorax with mediastinal shift, and hyperlucency of right lung field. The child improved on medical management and is asymptomatic up to 6 months of follow-up.

DISCUSSION

Pulmonary agenesis occurs due to the failure of the respiratory bud to divide equally between the two lung buds at 4th week of intrauterine life. This leads to normal development of one side of the lung, while
the other does not develop (agenesis) or undergoes only limited development (aplasia/hypoplasia) [3]. The precise etiology is not exactly known, but hereditary factors, viral agents, and dietary insufficiency of Vitamin A throughout pregnancy have been incriminated [3].

The heart and the opposite lung occupy the whole of the thorax. Mediastinal shift with heart rotation and kinking and compression of trachea by the displaced aortic arch and truncus arteriosus may be present. The presentations in these cases are usually chest infections during infancy and cardiopulmonary insufficiency or may be due to associated congenital anomalies [4].

Surgery is only indicated in symptomatic cases; prophylactic interventions should not be done in these subjects. Diagnostic or therapeutic bronchoscopy is needed to confirm the condition and in selected cases to suck the bronchial secretions to relieve the respiratory distress [5]. Aortopexy can be done in cases where findings are suggestive of compression on trachea by great vessels in the thorax. Diaphragmatic translocation is indicated in cases where patients had severe degrees of mediastinal shift and heart rotation, tracheal kinking, and compression of the aortic arch and innominate artery or emphysema of a single lung. In this procedure, the diaphragm is detached along the costal margin, from the sternum to the spine. The diaphragm is fixed at the level of the third rib laterally, fourth rib anteriorly, and to the adjacent ribs posteriorly to achieve tension-free attachment of the diaphragm [6]. Krivchenya et al. [7] reported 10 patients (9 with lung aplasia and 1 with lung agenesis) and found that all patients had different degrees of mediastinal shift and heart rotation, tracheal kinking and compression of the aortic arch, and innominate artery or emphysema of a single lung; only 2 of their patients required operation.

REFERENCES