Right lung agenesis

B. Kumar, D. K. Kandpal, C. Sharma, D. D. Sinha

ABSTRACT

Congenital pulmonary agenesis or aplasia is extremely rare. Although more than fifty percent of patients die before first five years of age, some individuals may remain asymptomatic throughout their life. A three-month-old female child with right pulmonary agenesis presented to us with severe respiratory distress. She was misdiagnosed as a case of foreign body bronchus at the peripheral health centre. Bronchoscopy confirmed the diagnosis and relieved the symptoms. It is recommended that invasive diagnostic procedures and prophylactic surgery should not be done in asymptomatic cases.

Key words: Foreign body bronchus, lung aplasia, pulmonary agenesis

INTRODUCTION

Lung aplasia is often associated with acute respiratory distress and has a high mortality rate.[1] Fifty percent born with pulmonary aplasia are stillborn or die within the first five years of life.[2] Bilateral congenital pulmonary agenesis is a rare lethal anomaly, first described by Morgagni.[2] In unilateral lung agenesis, the trachea continues directly into the main bronchus of the normally developed lung, and respiratory distress usually occurs due to retention of bronchial secretions and inflammations.[1] We describe a case of right lung agenesis presenting with respiratory distress due to aspiration and misdiagnosed as a case of foreign body bronchus at the peripheral centre.

CASE REPORT

A three-month-old female child, weighing 6 kg, presented with severe respiratory distress and peripheral cyanosis. Plain X-ray chest showed homogenously opaque right hemithorax with mediastinal shift and hyperlucency of left lung field [Figure 1]. She was misdiagnosed as a case of foreign body bronchus by the paediatrician at the periphery who referred the baby to us. The baby's mother gave a history of breast-feeding about 2 hours previously. There was sudden appearance of cough and difficulty in breathing a while after the feed. On examination, the child had cough, dyspnea, and tachypnea with peripheral cyanosis. Air entry was absent on right side while harsh breathing with wheezing was noted on left side of chest. She had bilaterally symmetrical chest wall without any skeletal deformity. Emergency bronchoscopy was performed. There were thick bronchial secretions with mucosal oedema and hyperemia. Bronchoscopy revealed that trachea continued as left main bronchus without any stenosis or compression with absent right bronchus remnant. Tracheobronchial suction was performed. The child improved dramatically in postoperative period. There was no past history of similar episode. Perinatal history was normal. Contrast enhanced CT-scan thorax was done which showed absence of right bronchus and pulmonary parenchyma with normal hyper-inflated left lung, extending anteriorly across midline to right and slightly posterior deviation of trachea. Thoracic cage appeared normal and the right hemithorax was occupied by vascular and cardiac chambers [Figure 2]. Echocardiography was within normal limits. The baby was discharged in improved condition. She is doing well. She had only one episode of cough and fever in the last ten months, which was managed successfully without hospitalisation.

DISCUSSION

Pulmonary agenesis is defined as complete absence of bronchus, parenchyma and vessels. In case of complete unilateral agenesis, no pleural cavity can be found on the affected side. 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.[1-3] Lung agenesis or aplasia is very rare and the exact incidence is not known. It appears to be in order of 1:15000 autopsies with a slight preponderance of females. Both lungs are equally affected but patients

Department of Paediatric Surgery, Sir Padampat Mother and Child Health Institute (JayKayLon Hospital) S.M.S. Medical College, Jaipur - 302004, Rajasthan, India

Correspondence:
Dr. Basant Kumar,
Room No. - 189, Resident Doctors` Hostel, JLN Marg,
S.M.S. Medical College, Jaipur - 302 004. Rajasthan, India.
E-mail: drbkpg75@rediffmail.com

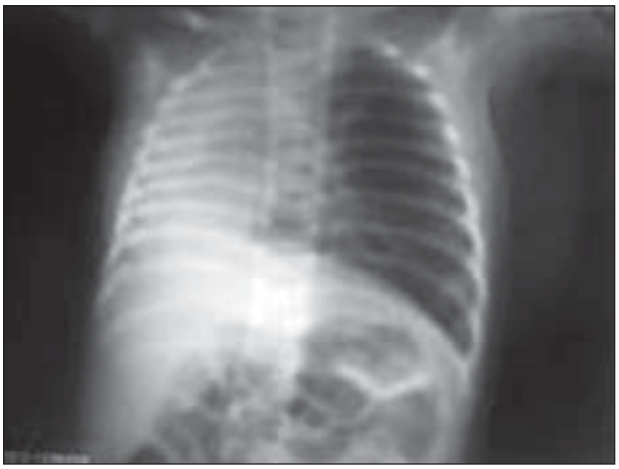


Figure 1: CECT- Thorax showing right pulmonary agenesis with mediastinal shift and hyper inflated left lung. Right hemithorax is occupied by vascular and cardiac chambers



Figure 2: X-ray chest showing opaque right hemithorax with mediastinal shift and over inflation of left lung

with aplasia of left lung have better prognosis.[2] Bilateral pulmonary aplasia or agenesis is incompatible with life.[4] Dextrocardia with situs solitus and normally related great arteries without intracardiac malformation may be present, secondary to agenesis or hypoplasia of right lung.[3]

In normal development, the heart shifts to the left in the 4th week of foetal life and simultaneously the trachea develops as a ventral diverticulum arising from the foregut.[3] Pulmonary agenesis or aplasia occurs perhaps due to the failure of the bronchial analogue to divide equally between the two lung buds. If this balance is not established, one side will develop normally while the other will fail completely (agenesis / aplasia) or undergo only limited development (dysplasia or hypoplasia).[2] A disturbance at 4th week of gestation could involve both the shift of the heart and the appearance of the right main bronchus.[3] More than 50% of patients with pulmonary agenesis have other associated anomalies including vascular, cardiac or non-cardiac like oesophageal, diaphragmatic, renal etc.[2,3] If left lung is present, it is frequently trilobed.[2] Patients usually present with respiratory distress, cough, tachypnea, strider, wheezing and cyanosis or with chronic unproductive cough.[1-4] More than 50% of children with lung aplasia die within five years of birth but some individuals live a normal life span with their deformity. Respiratory tract infections are the greatest danger in these individuals.[2]

Koseoglu *et al*.[5] described a 30-year-old man diagnosed with right lung agenesis and left pulmonary bronchiectasis, after diagnostic workup for dyspnea, developed during haemodialysis for chronic renal failure.[5] Musleh *et al.* [6] reported a 55-year-old man with left lung agenesis discovered during mitral valve repair.[6] Krivchenya *et al*.[7] on the basis of their series of 10 patients (9 patients with lung aplasia and 1 patient with lung agenesis), suggested that these patients had different degree of mediastinal shift and heart rotation. Tracheal kinking and compression by the aortic arch and innominate artery or emphysema of single lung may be present, secondary to lung aplasia.[7] In their series of 10 patients, only two patients required surgery.[7]

In our case, the child was asymptomatic up to three months of age then presented with sudden appearance of severe respiratory distress due to aspiration. Early intervention removed the distress and saved the child. There was no previous history of respiratory insufficiency. Perinatal history was insignificant. Child was full term, normal, vaginally delivered at home. Asymmetry of the thorax, with or without scoliosis, is a common but by no means universal finding.[2] In our case, scoliosis was absent and child had bilateral symmetrical chest wall without any skeletal deformity. In the last ten months, she had only one episode of cough and fever, which was managed successfully without hospitalisation. So, we decided against intervention. Patients with persistent or recurrent symptoms like chronic unproductive cough or signs of respiratory insufficiency, such as tachypnea, dyspnea, harsh breath sounds and cyanosis are indicated for surgery.[1-3,7] Most often, signs of respiratory insufficiency are due to kinking and compression of the trachea caused by heart rotation, mediastinal shift, and posterior curvature of trachea due to pressure by dislocated aortic arch and truncus arteriosus.[1,3,7] As evidenced by the literature, there is no specific therapy for this condition. Few cases are reported in literature that were treated successfully by the technique of aortopexy or diaphragmatic translocation.[1,3,7] These procedures provide recovery from respiratory distress by reducing

heart rotation, mediastinal shift and relieving kink and compression on trachea as well as hyperinflation of lung parenchyma.[1,3,7] Prophylactic surgical intervention is not recommended.[3,4]

It is concluded that congenital pulmonary agenesis is an extremely rare anomaly and may be asymptomatic throughout life. It may present acutely with severe respiratory distress and may need urgent intervention. Close follow-up is needed but invasive diagnostic procedures and prophylactic surgery are not required in asymptomatic patients.

REFERENCES

1. Krivchenya DU, Dubrovin AG, Krivchenya TD, Khursin VN, Lysak CV. Aplasia of the right lung in a 4-year-old child: Surgical stabilization of the mediastinum by diaphragm translocation leading to complete recovery from respiratory distress syndrome. J Pediatr Surg 2000;35:1499-502.
2. Skandalakis JE, Gray SW, Symbas P: The trachea and the lungs. In: Skandalakis JE, Gray SW, editors. Embryology for Surgeons. 2nd ed. Baltimore, MD: Williams and Wilkins; 1994. p 429-32.
3. Dohlemann C, Mantel K, Schneider K, Guntner M, Kreuzer E, Hecker WC. Deviated trachea in hypoplasia and aplasia of the right lung: Airway obstruction and its release by aortopexy. J Pediatr Surg 1990;25:290-3.
4. Maltz DL, Nadas AS. Agenesis of the lung: Presentation of eight new cases and review of the literature. Pediatrics 1968;42:175-88.
5. Koseoglu N, Ucan ES, Cavdar C. Right lung agenesis and left lung bronchiectasis. Respir Med Extra 2005;1:110-2.
6. Musleh GS, Fernandez P, Jha PK, Hasan R. Mitral valve repair in a 55 year old man with left lung agenesis. Ann Thoac Surg 2004;77:1810-1.
7. Krivchenya DU, Rudenko EO, Lysak SV, Dubrovin AG, Khursin VN, Krivchenya TD. Lung aplasia: Anatomy, history, diagnosis and surgical management. Eur J Pediatr Surg 2007;17:244-50.

Source of Support: Nil, **Conflict of Interest:** None declared.