

Unilateral lung agenesis: a case report and literature review

Rahul Tyagi, G.S. Chowdhary

Pulmonary agenesis is an extremely rare malformation of lung with an estimated prevalence of only 34 per million live births. It is even more uncommon in females and on the right side. We report a case of this rare entity.

Egypt J Bronchol 2018 12:486–488

© 2018 Egyptian Journal of Bronchology

Egyptian Journal of Bronchology 2018 12:486–488

Keywords: congenital malformation, lung agenesis, mediastinal shift

Introduction

Since its first description in 1673 by De Pozze [1], numerous cases of pulmonary agenesis have been reported. This extremely rare malformation is estimated to occur at a prevalence of 34 per million live births [2]. It is seen more commonly in male infants and affects left lung in almost 70% of cases [3]. We report a case of right-sided lung agenesis in a 16-year-old girl.

Case report

A 16-year-old girl presented with history of dry cough and breathlessness on exertion of 1-week duration. There was no history of fever, expectoration, hemoptysis, chest pain, wheeze, or weight loss. She was a full-term normal delivery, born of a nonconsanguineous marriage, and achieved her developmental milestones normally. Her general physical examination was within normal limits. Respiratory system examination revealed tracheal shift to right with relative flattening of right hemithorax. Chest movements were markedly reduced on the right side on both inspection and palpation. Percussion note was dull over all areas of chest on right side except suprascapular and infrascapular areas. Breath sounds were absent over right hemithorax. Breath sounds over left hemithorax were normal. Apex beat was shifted to right. Rest of the systemic examination did not reveal any abnormality.

Chest radiograph was done, which revealed a homogenous opacity in right middle and lower radiological lung zone with shift of trachea and heart to right (Figs 1 and 2). Lateral chest radiograph additionally showed an increased translucency in retrosternal area. Computed tomography of the chest revealed complete absence of lung on right with ipsilateral shift of mediastinum and compensatory hyperinflation and herniation of left lung to opposite side (Fig. 3). A small right main bronchus was seen

Department of Medicine, INHS Asvini, Mumbai, Maharashtra, India

Correspondence to Rahul Tyagi, MD, DNB Pulmonary Medicine, Department of Medicine, INHS Asvini, Near RC Church, Colaba, Mumbai 400005, Maharashtra, India. Tel: 22029413; e-mail: raw_haul4960@rediffmail.com

Received 18 February 2018 **Accepted** 24 June 2018

abruptly ending soon after tracheal bifurcation (Fig. 4). Her workup for associated congenital anomalies included ultrasonography of abdomen and echocardiography, where the results of both were normal. She was managed symptomatically and is under regular follow-up at our center. The nature of illness and possible complications were discussed in detail with the parents and the patient. She was also given influenza and pneumococcal vaccines.

Discussion

Pulmonary agenesis may be unilateral or bilateral depending on whether there has been an arrest of development of lung bud (bilateral agenesis) or developmental imbalance between two lung buds (unilateral agenesis) [1]. Pulmonary agenesis was initially classified by Schneider and Schawatbe and later modified by Boyden into three variants based on stage of development of lung bud [4]:

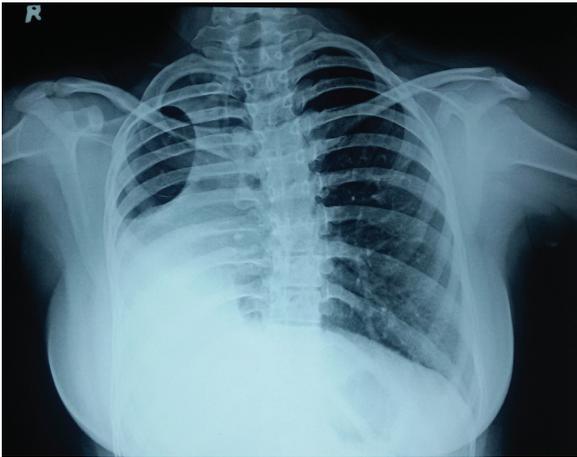
- (1) Type I: absence of lung parenchyma, bronchus, and blood supply to affected side (Agenesis).
- (2) Type II: absence of lung parenchyma with presence of rudimentary bronchus only. (Aplasia).
- (3) Type III: variable amount of lung parenchyma, bronchial tree, and vasculature (Hypoplasia).

Our patient will be classified as type II owing to presence of rudimentary bronchus on the affected side.

Although the exact etiology is not known, consanguinity, drugs, environmental agents, vitamin A deficiency, and intrauterine infections have been considered responsible for congenital lung

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Figure 1



Chest radiograph showing homogenous opacity in right middle and lower zone with ipsilateral shift of trachea and heart.

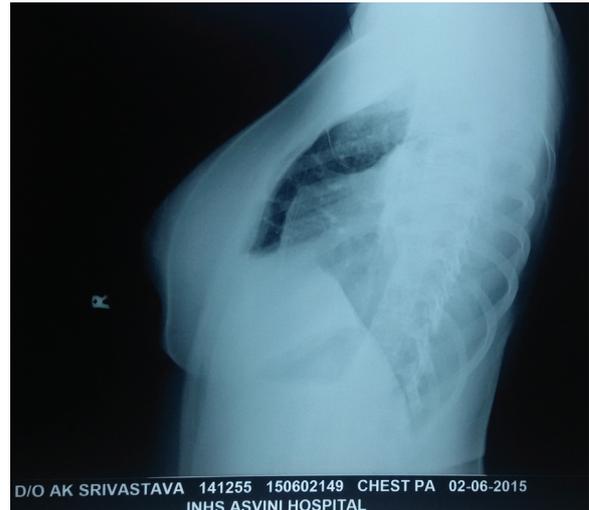
Figure 2



Computed tomography of the chest showing complete absence of lung on right with ipsilateral shift of mediastinum and compensatory hyperinflation and herniation of left lung to opposite side.

malformations [5]. Although commonly it is detected in childhood owing to repeated pulmonary infections or during assessment of associated malformation [2], there are reports of cases being diagnosed in adolescence without any symptoms till time of diagnosis [3,4], pneumothorax leading to diagnosis [2] and patient presenting with upper respiratory tract infection and being diagnosed as lung agenesis during evaluation [1]. Oldest patient with unilateral agenesis was diagnosed at 72 years of age [1]. Chest radiograph in lung agenesis may mimic pleural effusion, consolidation, collapse, or diaphragmatic hernia [2]. There are reports of pleural aspiration being tried [3] or patient being managed as bacterial pneumonia

Figure 3



Lateral chest radiograph showing increased translucency in retrosternal area.

Figure 4



Contrast-enhanced computed tomography showing small right main bronchus with gross ipsilateral mediastinal shift.

based on chest radiography [6]. Presently, contrast-enhanced computed tomography forms the standard investigation for diagnosis of lung agenesis, and pulmonary angiography and bronchography are rarely required [1,3]. Bronchoscopy for visualization of rudimentary or absent bronchus may be done but is not essential [1]. Prognosis depends on the side affected, status of remaining lung tissue and associated anomalies [4]. Right lung agenesis is associated with worse prognosis owing to carinal malrotation, distortion of bronchi and vascular structures, and deviation of heart and mediastinum [6]. Lung agenesis may be associated with other malformations in up to 50% of cases [3].

Associated anomalies may include patent ductus arteriosus, ventricular septal defect, atrial septal defect, narrowed trachea, and tracheoesophageal fistula [7]. In our patient, search for associated anomaly was done with help of echocardiography and ultrasonography abdomen, both of which were normal. Patients who are asymptomatic require no regular treatment [5]. Chest infections need to be treated aggressively, and patient with bronchial stump may benefit from postural drainage [5]. As mediastinal deviation can lead to air trapping in normal lung and tracheal abnormality may lead to respiratory distress, various surgical corrections have been attempted. Placement of inflatable prosthesis, aortopexy, and diaphragmatic translocation as surgical management for lung agenesis are reported [8].

Conclusion

Lung agenesis is an extremely rare congenital anomaly of the lung. As it is common on left side and in males, our report of this condition with right lung agenesis in a female makes it even more uncommon. Correct recognition of this condition is a must to avoid management as effusion, collapse or consolidation, and contrast-enhanced computed tomography is the most effective diagnostic

tool at present. Treatment consists of observation and aggressive management of chest infection.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

- 1 Singh U, Jhim D, Kumar S, Mittal V, Singh N, Gour H, et al. Unilateral agenesis of the lung: a rare entity. *Am J Case Rep* 2015; **16**:69–72.
- 2 Hentati A, Neifar C, Abid W, M'saad S. Left lung agenesis discovered by a spontaneous pneumothorax in a 20 year old girl. *Lung India* 2016; **33**:205–207.
- 3 Mohan A, Guleria R, Sharma R, Das C. Unilateral pulmonary agenesis: an uncommon cause of lower zone lung opacity. *Indian J Chest Dis Allied Sci* 2005; **47**:53–56.
- 4 Ta RK, Banerjee SN, Chatterjee K. Agenesis of left lung in an adult female: a rare case report. *Muller J Med Sci Res* 2015; **6**:172–174.
- 5 Roy PP, Datta S, Sarkar A, Das A, Das S. Unilateral pulmonary agenesis presenting in adulthood. *Respir Med Case Rep* 2012; **5**:81–83.
- 6 Malcon MC, Malcon CM, Cavada MN, Caruso PEM, Real LF. Unilateral pulmonary agenesis. *J Bras Pneumol* 2012; **38**:526–529.
- 7 Khurram MSA, Rao SP, Vamshipriya A. Pulmonary agenesis: a case report with review of literature. *Qatar Med J* 2013; **2013**:38–40.
- 8 Kim DH, Choi SH. Diaphragm translocation as surgical treatment for agenesis of the right lung and secondary tracheal compression. *Korean J Thorac Cardiovasc Surg* 2016; **49**:59–62.