

## Case Report

# Left Lung Agenesis in an Adult Patient: Diagnosis by CT Thorax and CT Pulmonary Angiography

Sarita Jilowa, Yashvant Singh, R.K. Mishra and Rani Devi Sharma

Department of Radio-diagnosis, Postgraduate Institute of Medical Education and Research and Associated Dr Ram Manohar Lohia Hospital (PGIMER - Dr RMLH), New Delhi, India

### Abstract

Agenesis of lung is a rare congenital anomaly. It is usually detected in early childhood as a result of pulmonary symptoms and associated cardiovascular, gastrointestinal or genitourinary anomalies. Diagnosis may be late in adulthood or later in life in asymptomatic patients. Patients often present with repeated chest infections. We present a case of left lung agenesis in a 40-year-old female patient, who presented with progressive breathlessness due to severe bronchiectatic changes in contralateral right lung using computed tomography (CT) of the thorax and CT pulmonary angiography. [Indian J Chest Dis Allied Sci 2017;59:139-142]

**Key words:** Lung agenesis, CT pulmonary angiography, Bronchography, Bronchiectasis.

### Introduction

Pulmonary agenesis refers to complete absence of the lung and its bronchi.<sup>1,2</sup> The diagnostic difficulty arises because there are no characteristic symptoms or clinical signs of pulmonary agenesis. The chest wall is normally developed and bilaterally symmetrical. The serious associated defects are less common in patients of left lung agenesis, hence these patients live longer. We report the occurrence of left lung agenesis in a 40-year-old female patient, who presented with progressive breathlessness due to severe bronchiectatic changes in contralateral right lung using computed tomography (CT) of the thorax and CT pulmonary angiography.

### Case Report

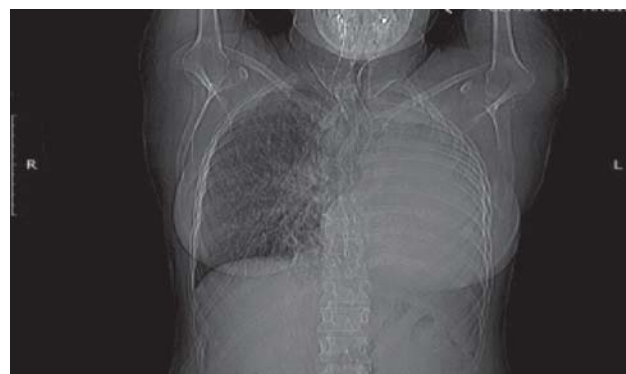
A 40-year-old female presented with progressive breathlessness for one year and cough with mucopurulent expectoration for two months. She also gave a history of chest pain, occasional palpitations, anorexia and weight loss. There was no history of previous hospitalisation or surgery.

On examination there was no spine deformity, pulse was 90/minute, blood pressure was 90/60 mmHg. Oxygen saturation on pulse oximetry was 70%. On auscultation, bronchial breath sounds, crepts in the inter-scapular and mammary area were evident in the right lung. However, there were absent breath sounds and impaired percussion note on the left side of the chest. On cardiovascular system examination, first and second heart sounds were normal. There

was a grade-II, non-radiating systolic murmur at the apex. Abdominal examination was normal.

On electrocardiography (ECG) normal sinus rhythm was noted. On two-dimensional echocardiography right atrium and right ventricle were dilated with normal shaped left ventricle cavity. Left ventricular ejection fraction was 55%. Mild tricuspid regurgitation and mild pulmonary artery hypertension were noted. Patient could not perform spirometry.

On computed tomography (CT) scanogram there was complete white-out of the lung field [Figure 1]. There was shift of mediastinal structures to the left side of hemithorax. The right lung showed non-homogeneous opacities with bronchiectatic changes. Diagnosis of total left lung collapse, aplasia or agenesis were considered.



**Figure 1.** Computed tomography scanogram showing left opaque hemithorax with crowding of upper ribs with shifting of mediastinal structures to left side with normally developed right lung.

[Received: November 9, 2016; accepted after revision: April 10, 2017]

**Correspondence and reprint requests:** Dr Yashvant Singh, Senior Radiologist, Department of Radio-diagnosis, Postgraduate Institute of Medical Education and Research and Associated Dr Ram Manohar Lohia Hospital (PGIMER - Dr RMLH), New Delhi-110 001, India; E-mail: y\_ashvant@yahoo.com

On bronchoscopy right main bronchus was visualised completely but the bronchi were found to be dilated and distorted. The left main bronchus was completely absent. Contrast-enhanced CT (CECT) of the thorax (mediastinal window) showed absent left lung. Left hemithorax was occupied by mediastinal structures with dilated and distorted bronchi of the right lung (Figure 2A). Mild compensatory expansion of the right lung was seen with shifting of mediastinal structures towards the left side. Computed tomography-pulmonary angiography showed absent left pulmonary artery. The pulmonary trunk and the right pulmonary artery measured 12.5cm in length. Antero-posterior diameter of pulmonary trunk was 3.5cm (Figure 2B).

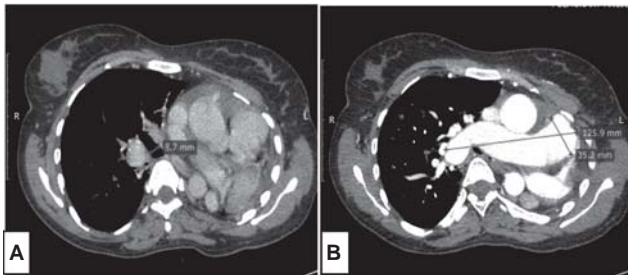


Figure 2. Axial computed tomography scan of the thorax (mediastinal window) and pulmonary angiography showing (A) absent left lung with left hemithorax occupied by mediastinal structures with dilated right bronchi and (B) long and stretched out pulmonary artery. Note the absence of left lung and left pulmonary artery.

On lung window, sub-pleural ill-defined nodular lesions and ground-glass haziness was scattered in upper lobe of the right lung with occasional bullae (Figure 3A). Cystic bronchiectatic changes with few cysts showing air-fluid levels in all basal segments of the right lower lobe and both segments of the right middle lobe were seen (Figure 3B). Mild herniation of the right lung was also noted across the midline (Figure 3C). Absent bronchus of the left lung with distorted bronchi of the right lung were confirmed

in coronal and sagittal sections. The patient was diagnosed to have left lung agenesis with superadded infection in right lung. Patient was managed symptomatically for chest infection and was advised to come for follow-up.

## Discussion

Agenesis of the lung refers to complete absence of the lung or lobe and its bronchi along with absence of supporting pulmonary and bronchial vasculature. Unilateral lung agenesis is not very common. Bilateral agenesis is not compatible with extra uterine life. Agenesis of the lung was first described in 1673 by de Pozze in an adult female in an autopsy case.<sup>3</sup> Munchmare first described it clinically in 1885.<sup>4</sup> It was concluded by Schmit in 1893 that formation of thoracic cage is independent of normal lung growth, when he found normal chest wall in an infant at necropsy who was born with agenesis of both lungs.<sup>5</sup> It is detected usually in childhood as a result of pulmonary symptoms or associated congenital heart disease (e.g., ventricular septal defect, atrial septal defect, Tetralogy of Fallot), gastrointestinal (oesophageal atresia, imperforate anus), genitourinary (e.g., renal agenesis or polycystic kidney disease) or musculoskeletal anomalies (e.g., hemivertebrae, absent ribs, absent radius) or other anomalies (e.g., hypoplastic trachea, ear deformities). More than half of the patients with unilateral lung agenesis die at birth or within five years of the life. The patients usually present with cough, tachypnoea, stridor, wheezing and at times with cyanosis. Morbidity and mortality are related to complications due to associated cardiac anomalies. Death has been attributed to respiratory failure.

Agenesis of the lung itself does not give rise to symptoms unless complicated by bronchopulmonary distress. Most patients present with repeated chest infections. The time of onset of symptoms is variable from patient to patient. Some patients are in great



Figure 3. Axial computed tomography of thorax (lung window) showing (A) distorted bronchi of the right lung with mosaic attenuation and bullae formation (arrow), (B) cystic bronchiectatic changes in the right lung with air fluid level (arrow) and (C) herniation of the right lung to the left. Left bronchus or left lung tissue not seen.

difficulty at birth, survive only one or two gasping respirations. On the other hand, case reports are available of even older patients. If not symptomatic, it may not be detected until adult life. The presenting complaints have been repeated attacks of lower respiratory tract infections, such as bronchitis, asthmatic bronchitis or pneumonia. Our patient was asymptomatic until adulthood and had no other significant disability.

The prognosis differs between agenesis of the right and the left lung. The average age at death is lower in right-sided lesions. Many earlier authors documented that average age of death for the right sided agenesis is six years and for left sided lesions it is 16 years. Over the years many authors concluded that right sided agenesis is associated with more severe congenital anomalies, like trachaeo-oesophageal fistula which leads to earlier death than the left sided agenesis. No gender predilection has been found.

Originally Schneider and Schwalbe classified agenesis into three groups which was later modified by Boyden depending upon stage of development of the primitive lung buds.<sup>6,7</sup> In Type I (agenesis) complete absence of lung and bronchus or bronchi with no vascular supply to the affected side is noted. In Type II (aplasia), rudimentary bronchus (short blind ending) with complete absence of pulmonary parenchyma occurs. In Type III (hypoplasia), presence of variable amount of bronchial tree, pulmonary parenchyma and supporting vasculature is described. Our patient was categorised to have type I (agenesis).

Trachea develops as a ventral diverticulum arising from foregut. Pulmonary agenesis/aplasia develops due to failure of bronchial analogue to divide equally into two lung buds. Abnormal blood flow in the dorsal aortic arch during fourth week of the gestation has been hypothesised as the cause of pulmonary agenesis. The contra-lateral lung may develop as much as twice more alveoli in response to pulmonary agenesis/hypoplasia. Biyyaam *et al*<sup>8</sup> reported a case with pulmonary agenesis in partial trisomy of 2p and 21q pointing towards the teratogenic factors. Chromosomal factors, vitamin A deficiency, intrauterine infection and environmental factors have been held responsible for aetiology.<sup>9</sup>

Majority of patients with this condition do not reach reproductive age. Prognosis depends upon severity of associated congenital anomalies and involvement of normal lung in any disease process.<sup>10</sup>

Sometimes, cases are picked up on routine chest radiography done for some other ailment. Chest radiograph reveals normal bony symmetry. However, opaque hemithorax with mediastinal shift and

herniation of the contralateral lung to the affected side points towards the diagnosis of lung agenesis.<sup>11</sup> Other differentials to the entity are pulmonary hypoplasia, complete lung atelectasis due to endobronchial occlusion (due to congenital/fibrosing stricture) and post pneumonectomy status.

Lung agenesis can also be detected on prenatal ultrasound screening which is seen as hyperechoic hemithorax.<sup>12</sup> The diagnosis is difficult to make but mediastinal shift is striking. The recognition of associated congenital malformation is of critical importance to the parents and the physician to predict the life expectancy. Conventional CT, CT pulmonary angiography, magnetic resonance imaging (MRI) and magnetic resonance (MR) angiography provide important diagnostic information showing the absence of lung parenchyma, bronchial tree, pulmonary vessel on the affected side. CT angiography and MR angiography are currently the imaging modalities of choice in diagnosis of this entity.<sup>13,14</sup> Conventional pulmonary angiography is used only in selective cases. With the advent of CT and CT angiography, other diagnostic tools, like bronchoscopy which are invasive, can be avoided. Bronchography is obsolete these days. Our case was diagnosed on CT thorax and CT angiography where trachea was seen continuously directing into right main bronchus of normally developed lung with absence of left main bronchus and left pulmonary artery and left lung parenchyma.

Asymptomatic patients with pulmonary agenesis do not require any treatment. Recurrent chest infections need symptomatic management. Patients with rudimentary bronchus (lung aplasia) require surgical correction postural drainage and antibiotics fails to treat the condition. Associated congenital anomalies need surgical correction. Bronchoscopy is useful in clearing of obstruction in cases of rudimentary bronchus of lung aplasia/hypoplasia. A long term survival is expected in patients with absence of associated congenital malformations and functional integrity of solitary lung.

## References

1. Smith RA, Bech AO. Agenesis of lung. *Thorax* 1958;13:28.
2. Sadashiv S, Culver GJ, Pirson HS. Agenesis of lung in an adult. *AJR Am J Roentgenol* 1979;133:755-7.
3. Pozze S, Brescia MA, Amerman EE, Sharma KK. Agenesis of left lung. *Arch Paediatr* 1960;77:485-90.
4. Kisku KH, Panigarhi MK, Sudhakar R, Nagarajan A, Ravikumar R, Daniel JR. Agenesis of lung: a report of two cases. *Lung India* 2008;25:28-30.
5. Yetim TD, Bayarogullari H, Yelcin HP, Arica V. Congenital agenesis of the left lung: a rare case. *J Imaging Sci* 2011;1:4711.
6. Dinamarco PVV, Ponce CC. Pulmonary agenesis and respiratory failure in childhood. *Autops Case Rep* 2015;5:29-32.

7. Espinosa L, Agarwal P. Adult presentation of right lung agenesis and left pulmonary artery sling. *Acta Radiologica* 2008;49:41–4.
8. Biyyam DR, Chapman T, Ferguson MR, Deutsch G, Dighe MK. Congenital lung abnormalities: embryologic features, prenatal diagnosis and postnatal radiologic-pathologic correlation. *Radiographics* 2010;30:1721–38.
9. Roy PP, Datta S, Sarkar A, Das A, Das S. Unilateral pulmonary agenesis presenting in adult hood. *Respir Med Case Rep* 2012;5:81–3.
10. Tsunozuka Y, Oda M, Ohta Y, Watanabe G. Congenital absence of right upper lobe of the lung. *Ann Thorac Surg* 2002;74:571–3.
11. Sunam G, Ceran S. Pulmonary artery agenesis and lung hypoplasia. *Eur J Gen Med* 2009;6:265–7.
12. Meller CH, Morris RK, Desai T, Kilby MD. Prenatal diagnosis of isolated right pulmonary agenesis using sonography alone: a case study and systemic literature review. *J Ultrasound Med* 2012;31:2017–23.
13. Johnson TRC, Thieme SF, Deutsch MA, Hinterseer M, Reiser MF, Becker CR, et al. Unilateral pulmonary artery agenesis: noninvasive diagnosis with dual source computed tomography. *Circulation* 2009;119:1158–60.
14. Prabhakar Rajiah. Pulmonary hypoplasia imaging. *Medscape* 2016