

Pulmonary agenesis: A rare entity

Sir,

Pulmonary agenesis results from abnormal development of the first and second aortic arches and is a rare entity. We report here a 17-year-old female patient with the complaints of gradually progressive dyspnea on exertion; the dyspnea was Grade 1 MMRC along with right-sided chest discomfort, progressing over the past 14 years associated with cough, and mucopurulent expectoration for the past 6 months. She had no complaints of fever, anorexia, and palpitations. The pain was diffuse in character involving the right anterior and posterior chest wall and was associated with cough impulse and exertion. She was born of a nonconsanguineous marriage with uneventful antenatal, natal, and postnatal history.

On general clinical examination, she was poorly built with body mass index of 18. She maintained saturation of 99% of oxygen. On respiratory examination, the trachea was shifted to the right with the right-sided sternocleidomastoid prominence. There were decreased breath sounds on the right side, and apex beat was situated in the right fifth intercostal space, 5-cm lateral to the sternum. Her chest X-ray [Figure 1] showed trachea shifted to the right side with volume loss; there was hyperinflation of the left lung. Her computed tomography (CT) thorax [Figure 2] showed the right pulmonary agenesis with left lung hyperinflation and herniation to the right side. The main pulmonary artery was normal in size with the hypoplastic right pulmonary artery. Fiber-optic bronchoscopy [Figure 3] was done which revealed complete stenosis of the right main bronchus. Pulmonary angiography for assessment



Figure 1: Chest X-ray showed deviation of trachea to right side with hyperinflation of left lung

of the pulmonary artery could not be done as the patient refused for it.

De Pozze^[1] first discovered the condition accidentally at the autopsy of an adult female in 1673. From India, the first case was reported by Muhamed^[2] in 1923, of a left-sided pulmonary agenesis in a medicolegal autopsy. The incidence of this condition is not precisely known, but various reports have suggested it to be from 0.0034% to 0.0097%.^[3] This is a rare malformation may be seen in isolation or associated with other anomalies involving cardiovascular, skeletal, and gastrointestinal systems.^[4] Most patients present with recurrent respiratory infection in childhood and half of them die within first 5 years of life itself.

Pulmonary agenesis is classified into three types,^[5] which is as follows:

- Type 1 (agenesis) – Complete absence of the lung and bronchus and no vascular supply to the affected side
- Type 2 (aplasia) – Rudimentary bronchus with the complete absence of pulmonary parenchyma
- Type 3 (hypoplasia) – Presence of variable amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature.

Our patient would classify as Type 2 pulmonary agenesis.

Autosomal recessive chromosomal aberration, associated with consanguineous marriage, deficiency of Vitamin A, intrauterine infections, and environmental factors have been held responsible for the etiology of congenital lung malformations.^[3] Pulmonary agenesis or aplasia occurs perhaps due to the failure of the bronchial analogue to divide equally between the two lung buds.

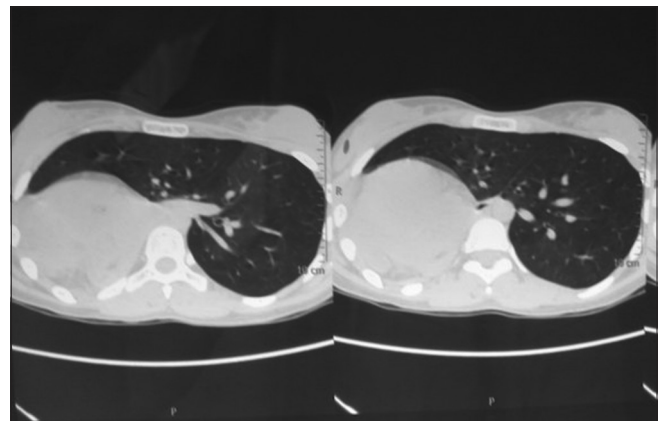


Figure 2: Computed tomography thorax-lung window showed hyperinflation of left lung with herniation to the right side

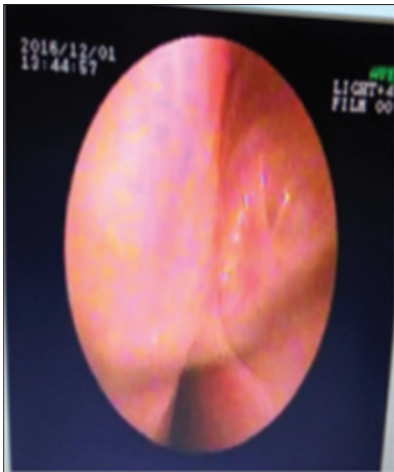


Figure 3: Bronchoscopy image showing complete stenosis of right main bronchus at the carina

In adults, unilateral agenesis of the lung may mimic collapse, thickening of pleura, destroyed lung, pneumonectomy, and scoliosis with pleural effusion, diaphragmatic hernia, adenomatoid cystic malformations, and sequestrations.

CT chest, which provides a detailed description of bronchial tree, parenchyma, and vasculature is considered to be the most definitive investigation to diagnose agenesis when chest radiograph is not diagnostic.^[6] Bronchography is almost obsolete now, but bronchoscopy is useful to demonstrate rudimentary bronchus.

Pulmonary angiography or magnetic resonance imaging angiography is considered to show the absence of ipsilateral pulmonary vessel.

No treatment is required in asymptomatic cases. Treatment is necessary for chest infections. Patients having stumps (hypoplastic bud) may require surgical removal if postural drainage and antibiotics fail to resolve the infection.^[6] Overall, prognosis depends on two factors. First, the severity of associated congenital anomalies, and second, involvement of the normal lung in any disease process.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Access this article online	
Quick Response Code: 	Website: www.lungindia.com
	DOI: 10.4103/lungindia.lungindia_294_17

How to cite this article: Vyas S, Mathew T, Advani M, Meena D. Pulmonary agenesis: A rare entity. *Lung India* 2018;35:275-6.

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