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A rare variant of right sided pulmonary agenesis presenting in adulthood: 1st reported case from Bangladesh

S.M.Tajdit Rahman [a,*], Tasnim Shahriar [b], Kazi Munzerin Akhter [c], Adiba Tarannum [d], Mosharraf Hossain [a]

[a] Department of Thoracic Surgery, National Institute of Diseases of the Chest & Hospital, Dhaka, Bangladesh
[b] Khulna Medical College, Khulna, Bangladesh
[c] Thoracic Surgery, Department of Thoracic Surgery, National Institute of Diseases of the Chest & Hospital, Dhaka, Bangladesh
[d] Green Life Hospital, Dhaka, Bangladesh

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ABSTRACT

Pulmonary agenesis is a rare disorder, and the right-sided one is much rarer. Most of the cases are diagnosed during early life. Because of rarity, it can be misdiagnosed and even more challenging to diagnose when presented during adult life.

However, we report a rare late manifestation of right-sided unilateral lung agenesis in a 22-year-old female patient who was treated for pneumonia several times, the first reported case from Bangladesh. We also highlighted the diagnostic approach of the case in low-resource settings.

1. Introduction

Pulmonary Agenesis (PA) is the absence of a lung and its supporting vasculature, with or without the absence or hypoplastic changes of the main bronchi [1]. Congenital PA is a rare ailment with a death rate of up to 50% by the age of five, originally reported by Depozze while performing an autopsy in 1673 [2,3]. It could be both bilateral and unilateral where bilateral PA is extremely rare and incompatible with life which was first described by Morgagni in 1955 [4]. Unilateral PA presents with a variety of severity of symptoms from patient to patient, depending on the affecting side. Because of its rarity, it can be misdiagnosed with similar clinical presentations like pneumonia with parapneumonic effusion, and so forth [2]. It is even more challenging to diagnose when presented during adult life. PA is often associated with other congenital anomalies, most commonly the skeletal, cardiovascular, genitourinary systems, and gastrointestinal systems [5]. Left PA which is less rare than the right one and has a better prognosis and higher life expectancy than right PA [5].

However, we report a rare late manifestation of right-sided unilateral lung agenesis in a 22-year-old female patient who was treated for pneumonia several times. This is the first reported adult case of right-sided pulmonary agenesis from Bangladesh that we are aware of.

2. Case

A twenty-two-years old lady was referred to our hospital from a rural city of Bangladesh with occasional dry cough and a history of

Abbreviations: PA, Pulmonary agenesis; CXR, Chest X-ray; CT, Computed Tomography.

* Corresponding author.
E-mail address: tanims@ymail.com (S.M.Tajdit Rahman).

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repeated respiratory tract infection since adolescence. In the meantime, she has been treated with several courses of antibiotics and empirical anti-tubercular drugs two years back due to close contact with an infected tubercular patient. Our patient had no history of high-grade fever, wheeze, chest pain, anorexia, or weight loss. She is the first issue of non-consanguineous parents and belongs to a middle-class family. There is no history of any congenital diseases in her family, and she was immunized according to the EPI (Expanded Program on Immunization) schedule of Bangladesh.

On general examination, she was found to be slightly anaemic and had an average body build with less muscle mass. There was no pallor, clubbing, icterus, engorged neck veins, or lymphadenopathy. Her vitals are within the standard limit during the examination. Respiratory system examination revealed right-sided restricted chest movement with asymmetry than the left with no scar or visible abnormality over the chest. The trachea was shifted to the right side, and the apex beat was heard over the right border of the sternum in the fourth intercostal space. On the right side, percussion was resonant in the upper chest, but impaired from the third intercostal space to downward anteriorly and along the scapular line posteriorly. It was resonant in the left side of the chest, but no cardiac dullness was noted. Breath sound was vesicular over the left chest and upper part of the right chest. However, no breath sound was heard in the rest of the right chest. Heart sound was more prominent on the left side of the chest.

Her routine blood test showed normal values except haemoglobin concentration 9.8 gm/dl. Chest x-ray demonstrated homogenous opacity in the right middle and lower chest with gross mediastinal shifting to the right and hyperinflation of the left lung (Fig. 1). Computed tomography of the chest revealed the complete absence of the right lung with the absence of right main bronchus and a compensatory increase in the left lung volume with right sided herniation (Fig. 2). Flexible bronchoscopy revealed no opening of right main bronchus with the absence of carina (Fig. 3). Left bronchial tree seemed to be normal.

She was diagnosed as a case of agenesis of the right lung and advised for regular follow up.

3. Discussion

Pulmonary agenesis (PA) is a rare condition; right-sided PA is significantly more uncommon [6]. The prevalence ratio of PA among male-female is similar, and the left lung affects more frequently compared to the right one and has a greater life expectancy [6].

It is estimated that nearly half of the PA cases are accompanied by cardiovascular, musculoskeletal, gastrointestinal and genito-urinary system anomalies [7]. Majority of the time, left PA is associated with other congenital anomalies like pulmonary artery atresia, patent ductus arteriosus, tracheoesophageal fistula, cardiac malformation and horse-shoe type kidney. However, several cases suggest right-sided agenesis is more associated with other congenital anomalies, and persons with right-sided agenesis mostly die within the first year of their life due to associated cardiac malformation [8].

Certain factors are considered responsible for the development of PA during the embryogenesis period. Though the actual cause is unknown, it is believed that autosomal recessive chromosomal abnormalities, consanguineous marriage, vitamin A deficiency, mother's use of salicylates, intrauterine infections, environmental factors are associated with PA [9].

Clinical presentation of agenesis lung is marked by its variety from recurrent childhood respiratory infection resulting from imperfect drainage of lung secretions or the spillover of pooled secretions from a blind bronchial stump into initially normal lung tissue, frequent hemoptysis due to bronchiectasis of remaining lung to major organ malformation leading the patient to succumb in early life. Patients may reach adulthood or may even live their entire lives without developing any symptoms [8,10]. Similar clinical presentations like collapse, thickening of pleura, pneumonectomy, destroyed lung, scoliosis with pleural effusion, diaphragmatic

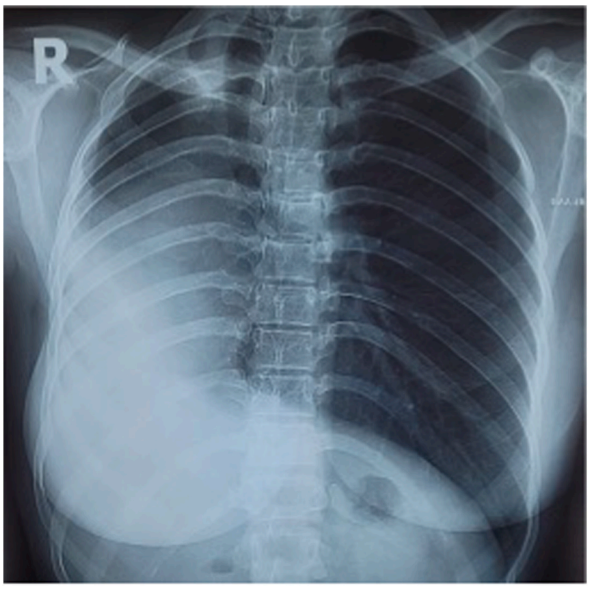

Fig. 1. Chest Xray shows Hyperinflated left lung with extreme mediastinal shift to the right side.

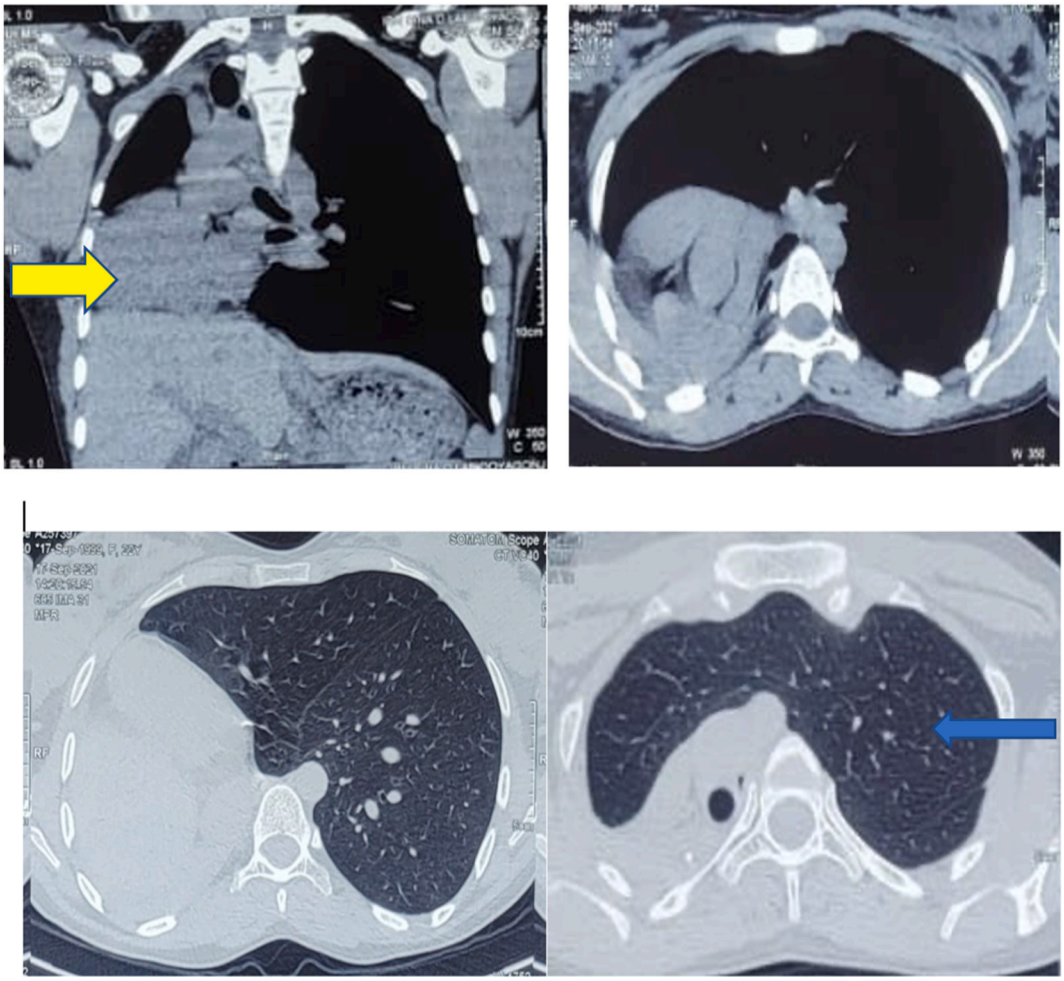

Fig. 2. CT chest demonstrates total absence of right lung along with compensatory overinflation of left lung extending to right hemithorax (blue arrow). Mediastinum and heart is grossly shifted to the right (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

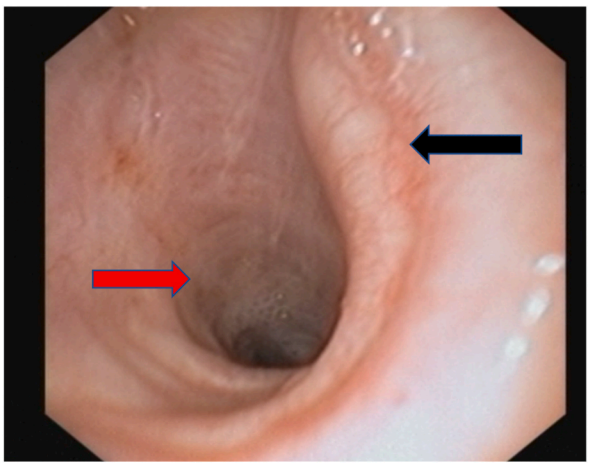

Fig. 3. Bronchoscopy reveals absence of opening of the right principal bronchus with presence of dimpling (black arrow). Red arrow indicates opening of the left principal bronchus. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

hernia, adenomatoid cystic malformation and sequestrations often lead to misdiagnosis of PA, and during adult life, it becomes more challenging [10].

Initial chest radiograph helps in the diagnosis of PA. Chest X-ray can present as an opaque hemithorax with ipsilateral mediastinal shift & contra-lateral lung hyperinflation [11]. And computed tomography of the chest provides detailed information about the bronchial tree, parenchyma and vasculature of the lung and is considered as the gold standard investigation to diagnose a case of PA [12]. Bronchoscopy also aids in the diagnosis, which demonstrates rudimentary bronchus of the affected side.

In asymptomatic cases, no treatment is required if there are no additional anomalies. Treatment is necessary for recurrent chest infections. Patients having bronchial stumps may require surgical removal if postural drainage and antibiotics fail to resolve the infection. Surgical intervention may be needed in cases with other associated congenital anomalies [10,13].

4. Conclusion

Pulmonary agenesis is a rare manifestation. Despite its rarity, lung agenesis should be considered as a differential diagnosis in a patient with repeated respiratory tract infection and unilateral lung opacification. A high index of suspicion and the early confirmatory investigation leads to early diagnosis and prevents further complications and any unnecessary treatment approach.

Ethics approval and consent to participate

Ethics approval has been taken from the ethical board of National Institute of Diseases of the Chest and Hospital, Dhaka, Bangladesh. Parents of the patient have given consent for the study.

Consent for publication

Consent for publication has been obtained from parent of the patient and copy has been stored.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

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Authors' contributions

Mosharraf Hossain was the chief of the unit, treating this patient and supervised the report writing. S M Tajdit Rahman (TRT) and Kazi Munzerin Akhter carried out the patient diagnosis. TRT contributed to regular follow-up of the patient, data collection from the in-hospital admission period. Tasnim Shahriar made the preliminary draft for the case report. TRT and Adiba Tarannum gave it a final form. All authors were involved in the final reviewing and approving of the final manuscript.

Declaration of competing interest

We do not have any financial and non-financial competing interests.

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