Case report

Right unilateral lung agenesis with dextroposition; the first case in Saudi Arabia

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A B S T R A C T

In 1673, the first case of lung agenesis was reported during an autopsy. Since then many more similar cases were reported and a classification of these cases developed for better medical management. Those cases ranged from hypoplasia to aplasia or agenesis of the lung tissue, with a survival rate of less than 50% by the age of 5 years in cases of agenesis. More commonly affecting the left side than the right, here we present a case of a late presentation of right lung agenesis in a young 24-year-old female. Who presented with multiple episodes of shortness of breath over a year. When examined she had an asymmetric chest and dull right sided auscultation. The x-ray done showed a homogeneous opacity in the right upper, middle and lower zone with shift of the mediastinum to the right side, making the heart in dextroposition. Watchfulness need to be maintained when encountering patients with simple complaint, as sometimes, it is not just the normal anatomy.

1. Introduction

Congenital lung agenesis is a rare condition with a mortality rate as high as 50% by the age of 5 [1]. Lung agenesis has been first reported by Depozze in 1673 while doing an autopsy on a woman [2]. It could be bilateral - in which life is incompatible - or unilateral where the severity of symptoms and clinical presentation vary from patient to another depending on the affected side.

Lung agenesis has been further divided into 3 major types: Agenesis, aplasia, and hypoplasia.

In agenesis (Type 1 agenesis), lung parenchyma, bronchus and pulmonary artery are all absent. On the other hand, aplasia (Type 2 agenesis) has no lung tissue but a rudimentary bronchial stump is present. In the last type, hypoplasia (Type 3 agenesis), the bronchi are present but the lung tissue is hypoplastic. Pulmonary agenesis in comparison to pulmonary hypoplasia, would show no lung tissue on radiological imaging or bronchus on bronchoscopy [3]. Pulmonary agenesis is associated with a variety of other congenital anomalies involving skeletal, cardiac, gastrointestinal and genitourinary systems [4]. Left pulmonary agenesis, which is the more common type, has been shown to have better outcomes and a longer life expectancy than right sided agenesis [5]. Both left and right pulmonary agenesis are associated with mediastinal shifts. Unilateral lung agenesis, specifically the right sided one, has been associated with dextroposition. Here, we describe a case of a rare late presentation of a unilateral lung agenesis.

2. Case report

A 24 years old female presented with shortness of breath for one month that became worse in a period of three days and accompanied with dry cough. There was no history of fever, wheeze, chest pain, anorexia, or weight loss. The patient gave history of experiencing multiple episodes of shortness of breath for one year, since then she has had frequent hospital visits due to her recurrent symptoms. Her perinatal history was insignificant and there was no history of similar complaints in any of her siblings. She was the outcome of a non-consanguineous marriage.

On general physical examination, she was of average build and moderately nourished, with no pallor, icterus, clubbing, engorged neck veins, or lymph-adenopathy. The examination of the abdominal, musculoskeletal, and neurological systems was unremarkable. On examination of her chest it was asymmetrical with the right side smaller and moving less than the left side. The trachea was deviated to the right side and the apex beat was heard at the 4th intercostal area mid axillary line on the right side. On the left side, percussion was resonant and no cardiac dullness were noted. On the right side it was resonant in the clavicular area, but impaired from the 4th intercostal space downward anteriorly and in a scapular line posteriorly. On auscultation, vesicular
breathing was heard on the left side and in the clavicular area on the right side. In the rest of the right lung breath sounds were absent. Investigation revealed hemoglobin of 12.2 g/dl. 

Chest radiograph showed right-sided homogenous opacity and hyperlucency of left lung with a mediastinal shift to the right side Fig. 1. Thoracic computed tomography examination revealed total absence of the right lung. A compensatory increase in the left lung volume was noted, which partially extended to the right hemithorax. The right pulmonary artery was absent, together with the right pulmonary veins. Along with small subpleural nonspecific pulmonary nodule Fig. 2.

3. Discussion

The respiratory bud develops at the caudal end of the laryngotracheal diverticulum during the 4th week of embryological development. This bud then gives two outpouchings, the primary bronchial buds, which then gives rise to the bronchi and the entire lung tissue [6]. Even though the underlying mechanism of how pulmonary agenesis occurs isn’t clearly understood, it has been suggested that vitamin A or folic acid deficiency or the mother’s use of salicylates may be one of the causes [7]. The loss of volume that accompanies pulmonary agenesis causes mediastinal shift in the direction of the affected side. Thus, with right pulmonary agenesis, the mediastinal shift causes the heart to be displaced. Three terms have been used to describe this abnormal shifting of the heart’s position. These terms are: Dextrocardia, Dextroversion and Dextroposition. However, there is still a matter of debate on which term is the most accurate and whether these terms can be used interchangeably or not. We believe that “Dextrocardia” has been divided into 3 subcategories. The first one - the most common type - is the “mirror...
image dextrocardia", where the parts of the heart are normal but the right to-left orientation is disrupted and is reversed. Abdominal situs inversus seems to be associated more commonly with this subcategory of dextrocardia. “Dextroversion” is the second type, and it is when the heart appears to be rotated into the right hemithorax, in relation to its normal position, while maintaining normal positioning of the great arteries. The third and final type of dextrocardia is “Dextroposition”, where a heart that was normal during embryological development is displaced and shifted into the right hemithorax by means of extracardiac causes such as right pulmonary agenesis, like in the case of our patient. Other causes of dextroposition include right pulmonary fibrosis and right pneumonectomy [8]. It has been shown that patients with left sided agenesis have better prognosis and longer life expectancy [9]. In regards to the reason behind this, it was hypothesized that increased morbidity and mortality of patients with right pulmonary agenesis in comparison to left sided ones is a result of a greater distortion in the tracheo-bronchial tree that occurs due to the mediastinal shifting [10]. However, a more recent review has showed that the most probable reason is related to the number of anatomical lobes and the ability of the right lung with its three lobes to adapt and meet the oxygen demand required by the body in a better way than the left lung would in cases of agenesis [11].

4. Conclusion

Caution and vigilance need to be taken in patient with similar symptoms and imaging profile as any chest infections can be life threatening. Prompt recognition and appropriate treatment will lead to good prognosis in patients with a solitary lung.

Declaration of competing interest

The authors declare no conflicts of interest in association with the present study.

CRediT authorship contribution statement

Elhaitham K. Ahmed: Conceptualization, Resources, Writing - original draft, Writing - review & editing, Visualization. Abdullah Tajeddin: Resources, Writing - original draft. Aya Farfour: Writing - original draft. Mubarak Alshammary: Supervision. Khaled AlKattan: Supervision.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.100994.

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