CASE REPORT

LEFT LUNG AGENESIS IN AN ADULT – A RARE ANOMALY.

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Abstract

Pulmonary agenesis is a rare congenital anomaly and only a few cases are reported in the literature. Adult cases present with a heterogeneous spectrum of manifestations and radiological findings. Historically, the diagnosis was made during autopsy, but with modern imaging technology, the diagnosis can be confidently made based on radiological findings.

We present a 72 year old lady with left lung agenesis, diagnosed at the age of 40 years old. This is the first case to be reported in Malaysia.

Key words: Agenesis, lung, Congenital, Incidental, Opaque lung

Introduction

Lung agenesis is a rare condition with a prevalence of approximately 1 in 100,000 births[1]. Bilateral lung agenesis is extremely rare and not compatible with life while unilateral agenesis is also very uncommon in adults. In childhood, it is usually accompanied by other system anomalies.[2,3] The oldest patient reported by Oyamada *et al*. was 72 years old and younger patients reported were 19 and 24 years old respectively.[4,5] We report a case of left lung agenesis in a 72 year old lady who had remained in reasonably good health despite this condition being diagnosed when she was 40 years old.

Case report

A 72-year-old lady with no co-morbid had been diagnosed with left lung agenesis at the age of 40 years when she presented with non-productive cough and wheezing. She had no significant childhood history of recurrent chest infections and no symptoms suggestive of reflux esophagitis or hiatus hernia. Physical examination of the chest revealed reduced breath sounds and dullness on the left side with right sided expiratory wheeze. Examination of the cardiovascular system was unremarkable. Lumbar scoliosis was noted.

Chest radiograph revealed a homogenous opacity of the left hemi thorax (Figure 1). Thoracic computed tomography (CT) demonstrated a hyper expanded right lung with, marked tracheal and midline shift to the left and a non- aerated left lung accompanied by small pulmonary vessels. (Figure 2, 3).

She was not able to complete the pulmonary function studies. Other laboratory evaluation, including tuberculosis screening was unremarkable. She was empirically treated for obstructive lung disease with budesonide inhaler.

Discussion

Lung agenesis is a rare congenital anomaly due to failure of pulmonary system development during embryogenesis [6, 7] and is usually associated with other cardiovascular anomalies.

The exact aetiology is still unknown although genetic, and teratogenic factors have been implicated[8] including viral agents, and dietary deficiency of Vitamin A during pregnancy. Agenesis of the lung was originally classified into three groups by Schneider and was subsequently modified by Boyden. Lung agenesis is classified by Boyden[9] into three categories.

Type 1 (Agenesis) - Complete absence of lung bronchus and no vascular supply to the affected side.

Type 2 (Aplasia) - Rudimentary bronchus with complete absence of pulmonary parenchyma.

Type 3 (Hypoplasia) - Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

Our patient will be classified as type 3 as a small pulmonary artery is present within a non-aerated lung. There was a case report in Malaysia of an isolated pulmonary artery agenesis in a young man who underwent pneumonectomy.[10] The incidence and prevalence of lung agenesis was found to be similar in both genders. Right lung agenesis has a shorter life expectancy compared to the left anomaly.[11] Our patient was only diagnosed at the age of 40 when she presented with respiratory symptoms.

The clinical manifestations vary from being asymptomatic to severe respiratory distress depending on the extent of malformations and presence of comorbidities. Recurrent chest infection is the most common presentation leading to the diagnosis and usually begins in childhood. Clinical findings may pose a diagnostic challenge especially in adults since there are other diagnosis to be considered such as lung collapse, pleural thickening, hiatus hernia and pleural effusion.

The CT scan is an important diagnostic tool for this rare entity as it will demonstrate the pulmonary architecture in great detail.

Lung agenesis is often associated with anomalies in the musculoskeletal, cardiovascular and

gastrointestinal systems.[12] Our patient presented with sciatica which is attributed to her lumbar scoliosis.

Conclusion

This case highlights the diagnostic challenge for clinicians, and one must have a high index of suspicion in patients presenting with repeated unexplained respiratory symptoms. CT or MRI/MRA are usually diagnostic. Treatment and intervention is usually indicated only for symptomatic patients.

Informed Consent

Written informed consent for the paper to be published (including images, case history and data) was obtained from the patient.

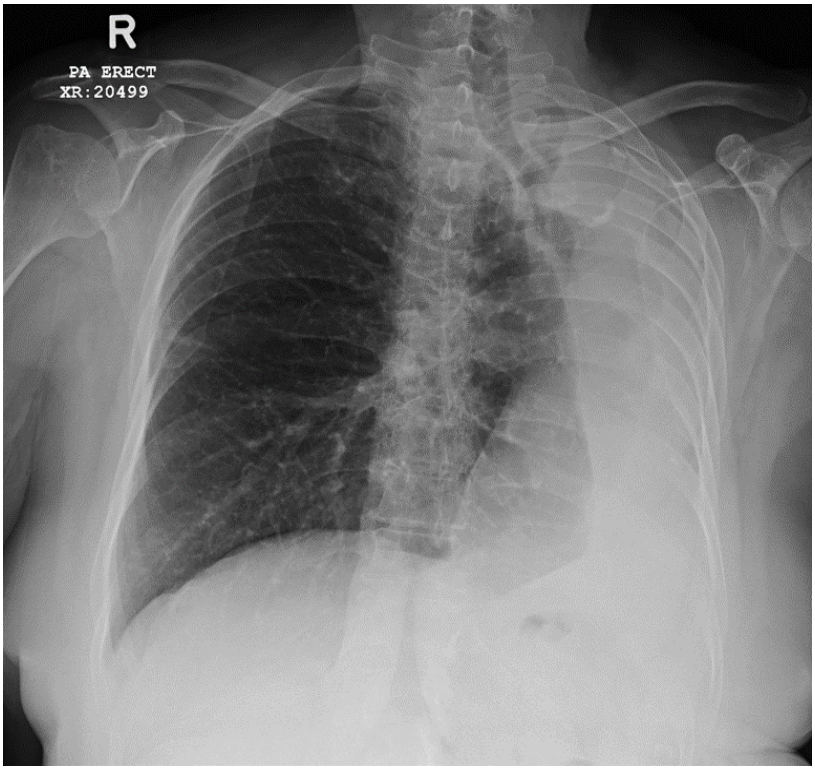

Figure 1: Plain Chest radiograph showing hyperinflation of the right lung with herniation towards the left thorax. There is gross tracheal and mediastinal shift to the left and crowding of ribs of the left thorax.

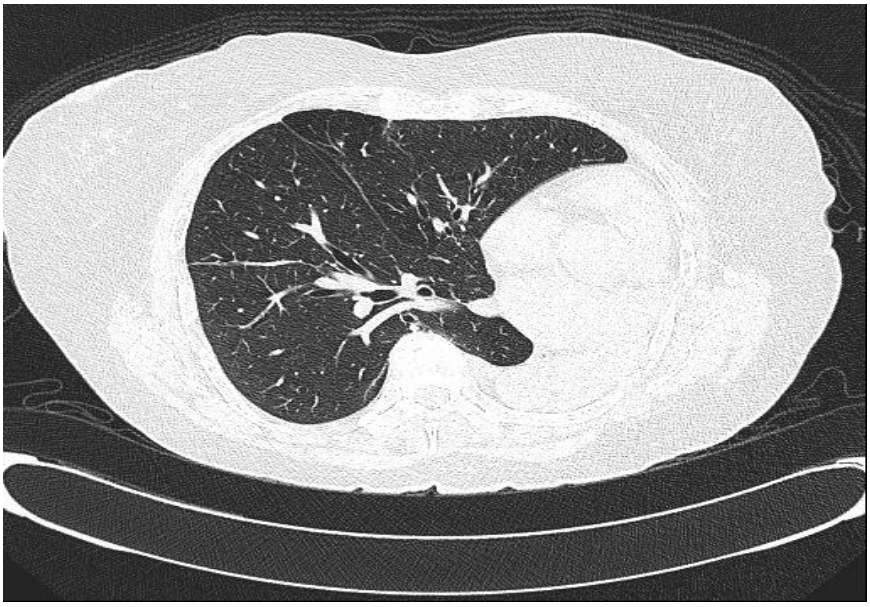

Figure 2. Computed axial tomography scan of the thorax showing the mediastinal shift to the left with hyper expansion of the right lung. Smaller left hemi thorax is present.

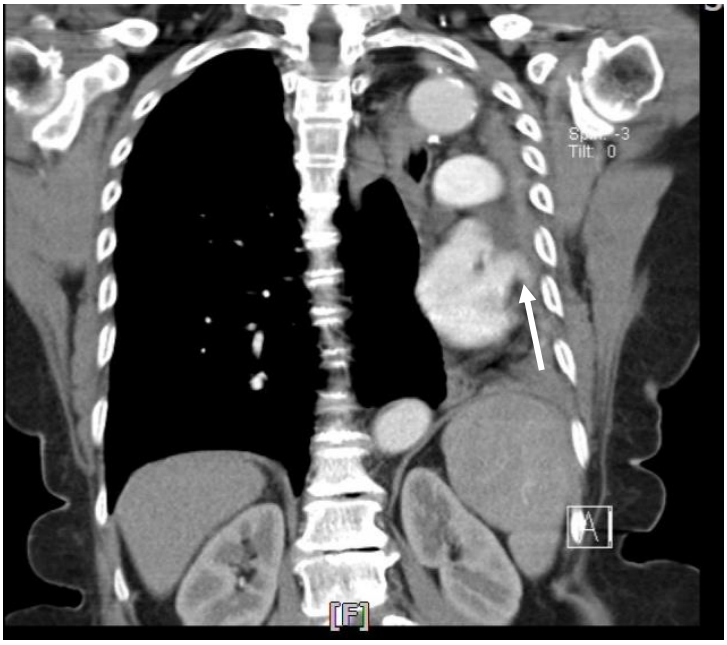

Figure 3. Contrast enhanced coronal CT Thorax showing the tracheal and mediastinal shift to the left with absent of the left lung parenchyma. Small left pulmonary artery seen (*Arrow*)

References

1. Conway K, Gibson RL, Perkins J, Cunningham ML. Pulmonary agenesis: Expansion of the VCFS phenotype. Am J Med Genet 2002; 113:89-92

2. Razaque MA, Singh S, Singh T. Pulmonary agenesis. Indian J Chest Dis Allied Sci 1980; 22:174-8

3. 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.

4. Oyamada A, Gasul BM, Holinger PH. Agenesis of the lung. Am J Dis Child 1953; 85(2):182-201.

5. Omer Kaya, Bozkurt Gulek, Cengiz Yilmaz, Gokhan Soker, Kaan Esen, Mehmet Ali et at. Adult presentation of symptomatic left lung agenesis. Radiology Case Report 201; 12:25 – 28.

6. Rogue AS, Burton EM, Boedy RF, Falls G, Bhatia JS. Unilateral pulmonary agenesis without mediastinal displacement. South Med J 1997; 90(3): 335-337.

7. Gupta R, Gupta A, Singh A. Congenital agenesis of lung – A case report and literature review. J Pediatr Dis 2016; 1(1): http://dx.doi.org/10.18282/jpedd.v1.i1.38.

8. Sbokos CB, McMillan IRK. Agenesis of the lung. Brit J Dis chest 1977; 3:183-197.

9. Boyden E. Developmental anomalies. AM J Surg 1955; 89:79-88

10. Britton J, Sachithanandan A, Srinivasan L, Ghosh S. Pneumonectomy for Congenital Isolated Unilateral Pulmonary Artery Agenesis. Med J Malaysia Vol 66 No 4 October 2011

11. Black PR, Welch KJ. Pulmonary agenesis (aplasia), oesophageal atresia, and tracheoesophageal fistula: A different treatment strategy. J Pediatr Surg 1986; 21:936-8.

12. Booth JB, Berry CL. Unilateral Pulmonary Agenesis. Arch Dis Child 1967; 42(224):361-74

13. Kaya O, Gulek B, Yilmaz C, et al. Adult presentation of symptomatic left lung agenesis. Radiol Case Rep: 2017: 25-8.