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CASE REPORT

PULMONARY AGENESIS WITH UNILATERAL RENAL AGENESIS AND POLYSPLENIA IN A YOUNG PATIENT: A RARE CASE REPORT

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ABSTRACT

Introduction: Pulmonary agenesis is a rare congenital anomalythat is due to developmental failure of primitive lung buds. It is often associated with other congenital anomalies like cardiovascular, central nervous, gastrointestinal, genitourinary and skeletal systems.[1]The combination of pulmonary agenesis with renal agenesis and polysplenia makes it a first-of-its-kind case. **Case report:** A25-year-old female presented with complaints of low-grade fever, chest pain and shortness of breath on exertion for 2 months. On examination, she hadan asymmetrical chest with reduced chest expansion, vocal fremitus, vocal resonance and breath sounds on the left hemithorax. X-ray chest showed homogenous opacity in the middle and lower part of the left chest. So provisionally we kept the diagnosis as collapse or loculated pleural effusion.The patient underwent further radiological investigations and was diagnosed as a case of left pulmonary agenesis with left renal agenesis and polysplenia. **Discussion:** Bilateral pulmonary agenesis is not compatible with life whereas patients with unilateral agenesis can present at any age with variable respiratory symptoms and recurrent chest infections. The condition mimics other pathologies like collapse, pleural effusion and consolidation on clinical examination and radiograph. **Conclusion**: Pulmonary agenesis is arare condition that is generally diagnosed in childhood and its presentation in adulthood is extremely rare. Once diagnosed, the patients need further evaluation for other associated congenital anomalies which can be treated potentially before getting symptomatic.

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INTRODUCTION

Pulmonary agenesis is an intriguing congenital anomaly characterised by the absence or underdevelopment of one or both lungs during embryonic development. It is an exceptionally rare condition, the incidence of which is not precisely known but various reports have suggested it to be from 0.0034 to 0.0097%.[2]Although the main aetiology of the disease is unknown, lack of vitamin A during pregnancy, viral agents, and genetic as well as iatrogenic factors have been mentioned as possible causes.[3]It can occur in isolation, however, when combined with other congenital anomalies of cardiovascular, central nervous, gastrointestinal, genitourinary and skeletal systems, these cases present a perplexing medical enigma that demands immediate attention. Pulmonary agenesis is classified into three types, 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.[4] In our case it is type one pulmonary agenesis with multisystem involvement.

CASE REPORT

A 25-year-old female, unmarried, student, pursuing her higher education, was admitted tothe medicine ward of Maharana Bhupal Government Hospital, RNT Medical College, Udaipur, Rajasthan with complaints of fever, chest pain and shortness of breath for the last 2 months. The fever was low grade, continuous, with no diurnal variations, relieved on medications, and not associated with cough, coryza, headache, palpitations or night sweats. She also had insidious onset and non-progressive shortness of breath for the last two months, only on exertion, although she can carry out her routine daily activities with ease. She isthe third child to her parents and was bornat full term by normal vaginal delivery at home. According to her mother, she was a low-birth-weight baby. Apart from being operated for appendectomy 2 years back, she has never been admitted to the hospital.

Parents and all the siblings are healthy and never soughtmedical attention for a similar illness. The patient's menarche was at the age of 13 years and has been continuous with normal flow since then. The patient was vitally stable at the time of admission. On general physical examination, the patient was poorly built with a BMI of 17, pallor present, and no icterus, cyanosis, clubbing, lymphadenopathy or oedema. On detailed respiratory examination, the chest was asymmetrical, depressed on the left side, intercostal spacesretracted, tactile vocal fremitus and vocal resonance reduced, dull note on percussion and air entry reduced in the left side in left interscapular, infra-axillary, inframammary and subscapular regions. Physical examination findings along with the chest radiograph, the patient was provisionally diagnosed withleft lung collapse or loculated pleural effusion as another possibility.

Routine investigation showed hemoglobin–10.8 g/dL, total leukocyte count (TLC) –5510/cu mm, platelet count –2.13 lakh/cu mm, urea –23.3 mg/dL, creatinine - 0.6 mg/dL, total bilirubin –0.17 mg/dL, conjugated bilirubin –0.09 mg/dL, serum-glutamic oxaloacetic transaminase (SGOT)/serum glutamate pyruvate transaminase (SGPT)- 20/08 U/L. Serum electrolytes were sodium 139mEq/L, potassium 4.1mEq/L and chloride 107 mEq/L. Her viral markers (HBV, HCV, HIV) were also negative. Her hormone assays showed triiodothyronine (T3) - 2 nmol/L (1.3-3.1), thyroxine (T4) – 90.6 nmol/L (66-181),thyroid-stimulating hormone (TSH) - 3.3 uIU/mL (0.27-4.20). Chest X-Ray PA View reveals trachea shifted to the left side with a rounded homogenous radiolucent shadow in the left upper zone with bronchopulmonary markings seen in the shadow and also homogenous opacity in the left middle and lower zone with CP angle obliteration. The above findings are suggestive of collapse-consolidation/ fibrothorax/ loculated pleural effusion.

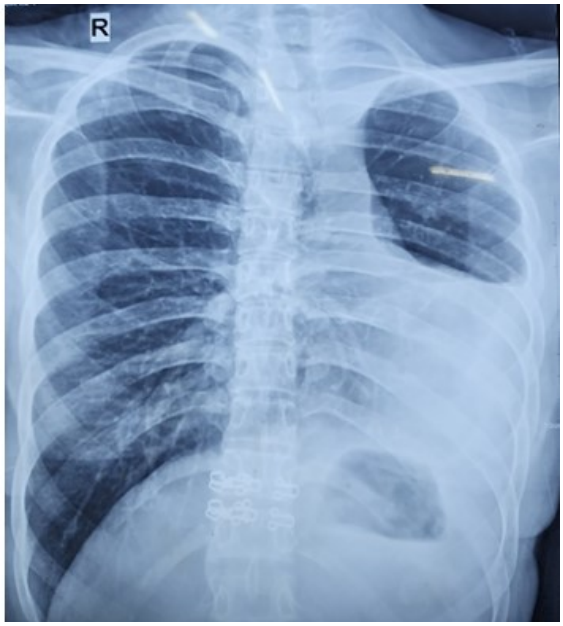

Figure 1. Chest X-Ray PA View

CECT Thorax reveals non-visualization of the left lung parenchyma, left main bronchus, left pulmonary vasculature with a gross mediastinal shift towards the left side and hyperinflation of the right lung resulting in part of the right upper lobe and right middle lobe occupying left upper thoracic space.

It also reveals an absent kidney in the left renal fossa and a spleneculi in the right side (30*27)mm adjacent to the upper pole of the spleen. Figure 2, 3 and 4

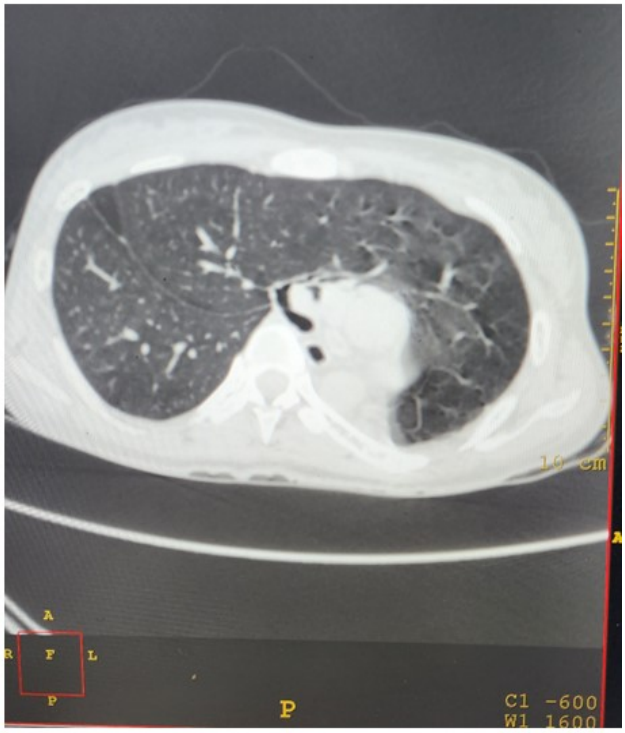

Figure 2. CECT Thorax axial view showing compensatory hypertrophy of left lung

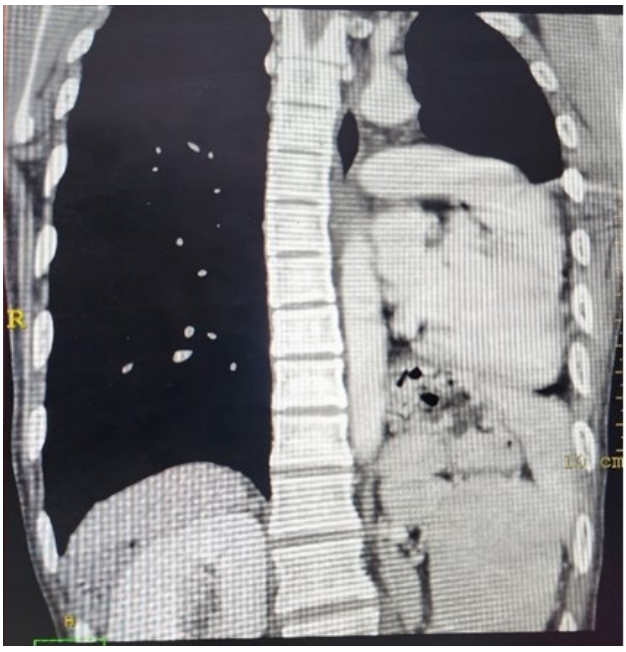

Figure 3 CECT Thorax coronal view showing transverse heart, one large and one small splenucli, and absent left kidney

USG abdomen shows two spleens and an absent left kidney. The right kidney measured (100*41)mm. Other organs revealed no abnormality.

2D Echocardiography: Viscero-Atrial Situs Solitus, Levocardia {base (right) to apex (left)}, transverse heart, left pulmonary artery could not be visualized. After all the relevant investigations, the patient was diagnosed as a case of left pulmonary agenesis. The patient was asymptomatic for any

gastrointestinal conditions and had no complaints of decreased urination or any complaints related to urinary tract infections. for diagnosing other system involvement, we wanted the patient to undergo CECT abdomen but as the patient did not give consent for further investigations and treatment, she was treated symptomatically and discharged.

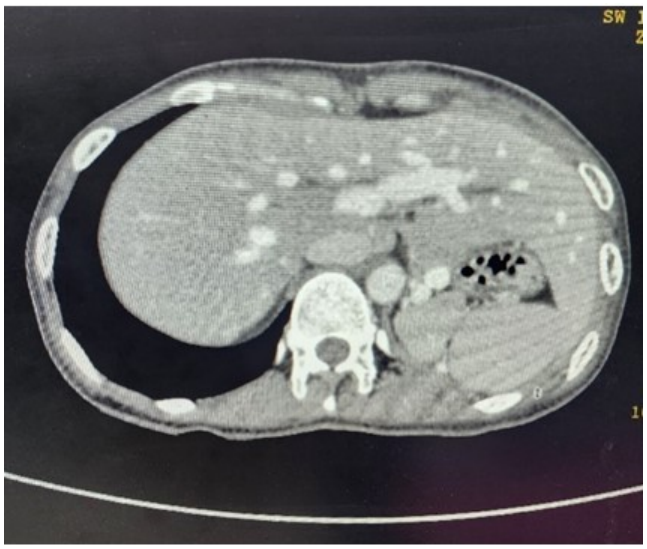

Figure 4. CECT Thorax Axial view revealing two spleens

DISCUSSION

Pulmonary agenesis is a rare disease associated with other systems involvement; hence, in most cases diagnosed in childhood when symptomatic. In other cases, it can present as an isolated condition and be picked up at any age later on. On physical examination and radiograph, the disease resembles closely with collapse, pleural effusion and consolidation. Further radiological investigations like contrast-enhanced CT scan or pulmonary angiography for differentiating among the three types of pulmonary agenesis, 2D-echocardiography to find out any associated cardiac pathology, CECT or USG whole abdomen for pathologies of the genitourinary tract and gastrointestinal system should always be done. Any pathology detected should be given a multimodal approach and referred to differentspecialities.

The significance of understanding and managing pulmonary agenesis cannot be overstated. Not only does this condition present intricate diagnostic challenges for clinicians, but it also poses immense physical, emotional and societal burdens for affected individuals and their families. By conducting a meticulous analysis of the existing literature, we can pave the way for future research end eavours, facilitate improved clinical management practices and ultimately enhance the quality of life for individuals living with this rare congenital anomaly.

CONCLUSION

In conclusion, through this paper publication, we aim to consolidate the current knowledge surrounding pulmonary agenesis with multiple system involvement, drawing attention to its variable clinical presentation and potential interventions. By addressing the gaps in our understanding and exploring novel insights that may impact this rare condition's diagnosis and management, we hope to facilitate a collective effort towards providing comprehensive care and support for affected individuals.

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