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Right Pulmonary Agenesis with Associated Skeletal Malformation

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Abstract **Case Report**

Pulmonary agenesis is a rare congenital anomaly defined by the total absence of pulmonary parenchyma, bronchi and vessels. It is often associated with other malformations. We report the case of a late revelation of right pulmonary agenesis in a 3 year old patient, presenting with recurrent episodes of respiratory infection. Chest radiography showed an opaque right hemithorax. The diagnosis of right lung agenesis was confirmed by chest CT scan with contrast enhancement. In our case pulmonary agenesis was associated with ipsilateral rib synostosis. CT scan is the main imaging modality for establishing a diagnosis of pulmonary agenesis, as well as demonstrating other associated malformations.

Keywords: pulmonary parenchyma, malformations, respiratory infection.

INTRODUCTION

Pulmonary agenesis is a rare congenital entity in which there is complete absence of one or both lungs, including the bronchi, bronchioles, vasculature, and respiratory parenchyma. It can be unilateral or bilateral. Bilateral pulmonary agenesis is extremely rare and is incompatible with extra uterine life.

In 50% of cases, lung agenesis accompanies other congenital anomalies like cardiovascular, digestive, genitourinary, and central nervous system anomalies [1, 2].

Patients can be asymptomatic or present with respiratory complaints such as dyspnea and respiratory distress or recurrent chest infections. The symptoms may occur as early as in neonatal period or later on during childhood and even adult life. Multidetector computed tomography (MDCT) is the preferred imaging modality for definitive diagnosis [1, 3, 4].

Here we present the case of a 6 year old patient with pulmonary agenesis and associated costal malformation.

CASE REPORT

A 3 year old boy was admitted to our hospital with a complaint of dyspnea. On admission, the patient was eupneic, afebrile. He has a history of recurrent respiratory infection. General physical examination revealed no anomaly. The respiratory system examination revealed no breathing sounds on the right side and normal breathing sounds on the left side. Heart sounds were displaced, heard from the right hemithorax.

A posterior anterior (PA) chest radiograph revealed homogenous opacity of the right hemithorax. Right tracheal, mediastinal and cardiac shift was noted (Fig. 1).

There was also rib synostosis across the right 8th intercostal space, connecting the medial aspects of the ribs (Fig. 1).

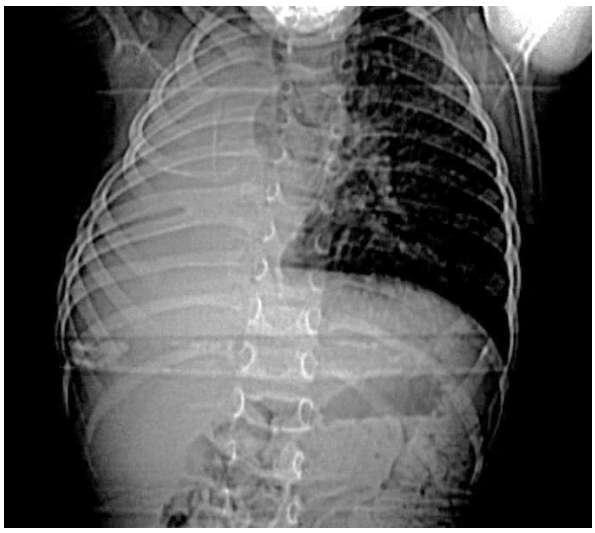

Figure 1: Opaque right hemithorax with mediastinal shift and rib synostisis

Upon these findings, a MDCT scan of the chest was performed, with contrast administration.

It revealed the absence of the right lung parenchyma, right main bronchus, and right pulmonary vasculature. The left lung had normal pulmonary vasculature, was hyper inflated, and partially extended to the right hemithorax (Fig. 2a, b).

The heart size was normal.

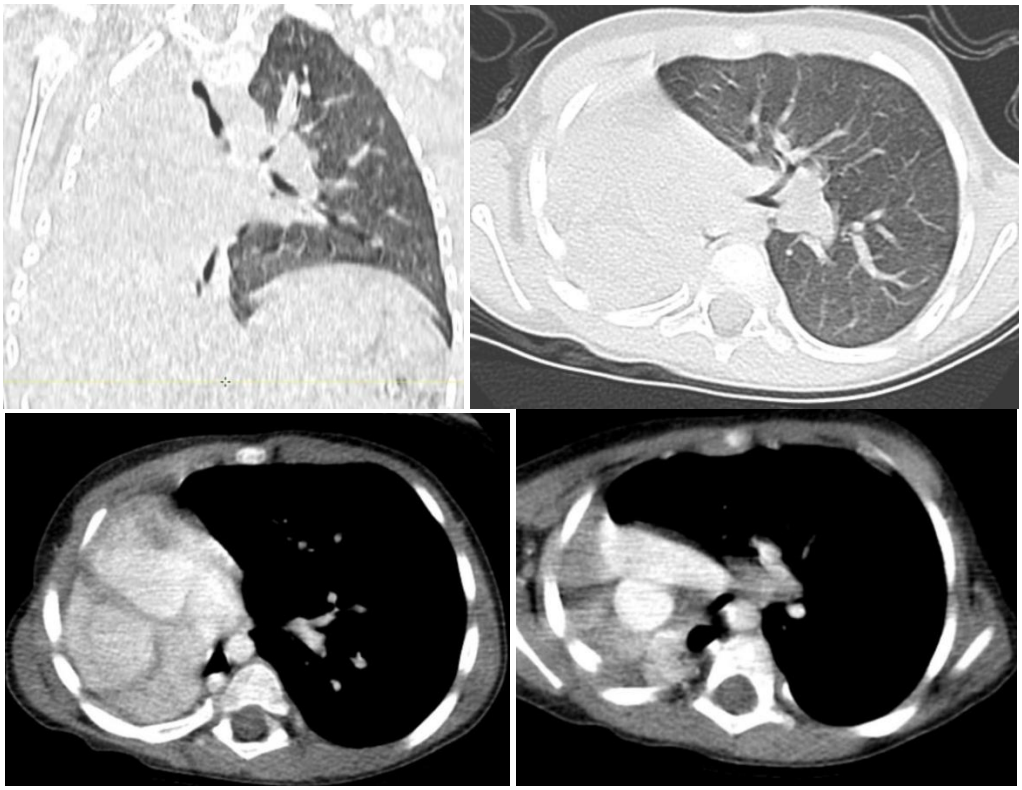

Figure 2: Right pulmonary agenesis: a) ***Study in parenchymal window shows the absence of right pulmonary parenchyma, as well as the absence of bronchial structures, best seen on the coronal reformatted image*****. b)** ***Study in mediastinal window shows the absence of a right pulmonary artery, as well as the right mediastinal shift***

CT also showed the rib synostosis of the posterior ends of 8th and 9th ribs (Fig. 3).

There were no vertebral abnormalities on the MDCT image. No additional congenital anomalies were detected. On the basis of these finding, the patient was diagnosed with type 1 right pulmonary agenesis.

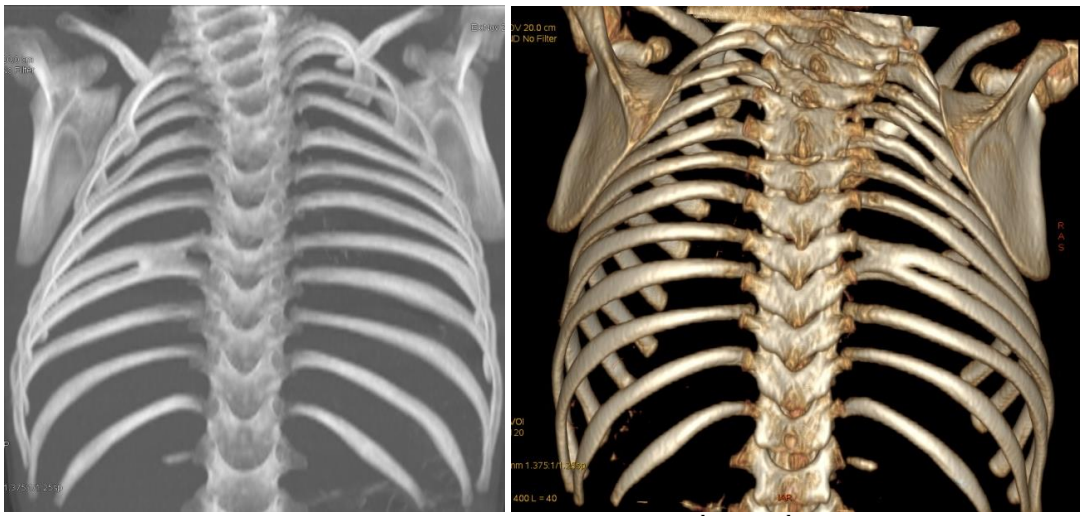

Figure 3: Right rib synostosis of the posterior segments of the 8th and 9th ribs showed on VR and 3D reconstructions

DISCUSSION

Pulmonary agenesis and aplasia represent two different forms of arrested lung development that result in the absence of the distal lung parenchyma , with and incidence ranging from 1 to 2 per 10,000 live births [5].

The lungs normally develop from the foregut during the 4th and 5th weeks of gestation.

Hypoplasia, aplasia and agenesis of unilateral pulmonary parenchyma result from the failure of bronchial analogue to divide equally between two lungs with possible abnormal blood flow in dorsal aortic arch.

There is concomitant increased alveoli production in the contralateral lung in compensation. During this period of time the migration of heart also occurs, and therefore congenital heart anomalies may coexist [1].

Pulmonary agenesis is the complete absence of one or both lungs, including bronchi, bronchioles, vasculature, and respiratory parenchyma. In pulmonary aplasia, only rudimentary bronchi are present, each of which ends in a blind pouch, with no pulmonary vessels or respiratory parenchyma [5].

The classification of pulmonary agenesis was modified by Boyden [6]; depending upon the stage of development of the primitive bud, agenesis of the lung is divided into three variants:

- **Type I: Agenesis:** Absence of the lung parenchyma, bronchus, and blood supply in the affected side.
- **Type II: Aplasia:** Absence of the lung parenchyma and the presence of only a rudimentary bronchus.
- **Type III: Hypoplasia:** A variable amount of the lung parenchyma, bronchial tree, and vasculature.

Postero anterior chest radiography is the first imaging modality used for the diagnosis of pulmonary agenesis. It shows diffuse opacification and decreased size of the affected hemithorax, compensatory hyperinflation of the contralateral hemithorax and elevation of the ipsilateral hemidiaphragm.

It also demonstrates absent ipsilateral pulmonary artery shadow and ipsilateral mediastinal shift [3].

MDCT is used for the definitive diagnosis of pulmonary agenesis. It shows the absence of bronchi in case of agenesis or the presence of a rudimentary pouch on the affected side in case of aplasia.

Differential diagnosis include total collapse of the lung, diaphragmatic hernia, pneumonitis, pleural effusion and pulmonary hypoplasia [3].

Prenatal diagnosis is possible , suggested by a prenatal ultrasound showing hyperechoic hemithorax and can be confirmed by Fetal MRI [1].

Pulmonary agenesis is frequently associated with other congenital anomalies of the lung, such as tracheal stenosis, trachea esophageal fistula, and bronchogenic cysts, as well as cardiac defects including patent ductus arteriosus, tetralogy of Fallot, and anomalies of the great vessels [5, 7].

CONCLUSION

Pulmonary agenesis is a rare congenital malformation, which can present at birth with neonatal respiratory distress or later in life with recurrent infectious episodes being the main symptom. MDCT with contrast enhancement is the key for a correct diagnosis. The prognosis depends essentially on the associated malformations and the date of onset of symptoms.

REFERENCES

1. Sadiqi, J., & Hamidi, H. (2018). CT features of lung agenesis – a case series (6 cases). *BMC Medical Imaging*, *18*(1), 37.
2. Kané, B., Camara, M. A., Togo, S., Touré, B. M., Traoré, M. M., Ouattara, M. A., & Togo, B. (2021). Agénésie pulmonaire droite: rapport de 2 cas dans le Service de Pédiatrie de l´ Hôpital du Mali de 2015-2019. *PAMJ-Clinical Medicine*, *5*(16). Disponible sur: https://www.clinical-medicine.panafrican-med-journal.com/content/article/5/16/full
3. Tanrivermis Sayit, A., & Elmali, M. (2020). An adult patient presenting with right unilateral pulmonary agenesis: a case report and literature review. *Surg Radiol Anat*, *42*(11), 1299-301.
4. Nakoulima, A. D., Niang, T., Fall, K., Diallo, M., Fall, M., Goumba, A. I., & Seye, M. N. (2021). Agénésie pulmonaire de révélation tardive. *Journal de Pediatrie et de Puericulture*, *34*(6), 323-326.
5. Wert, S. (2017). Normal and Abnormal Structural Development of the Lung. In p. 627-641.e3.
6. Boyden, E. A. (1955). Developmental anomalies of the lungs. *Am J Surg*, *89*(1), 79-89.
7. Berrocal, T., Madrid, C., Novo, S., Gutiérrez, J., Arjonilla, A., & Gómez-León, N. (2004). Congenital Anomalies of the Tracheobronchial Tree, Lung, and Mediastinum: Embryology, Radiology, and Pathology. *RadioGraphics*, *24*(1), e17-e17.