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CASE REPORT

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Congenital lobar emphysema: A diagnostic dilemma with coexistent congenital heart defects

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Key Clinical Message

Clinicians should think beyond pneumonia and left-to-right shunts when young children have persistent respiratory distress. Congenital lung anomalies, including congenital lobar emphysema, should be considered differential diagnoses. Chest X-ray and CT imaging should be conducted to gain insight regarding and establish diagnosis, respectively.

Abstract

Congenital lobar emphysema (CLE) is a rare and life-threatening congenital lung anomaly that often poses a diagnostic dilemma. The issue can be further confused with coexistent congenital heart defects (CHDs) with left-to-right shunts. The clinical presentation of CLE during infancy is similar to that of CHD, with CHD being more common entity, gets detected early. The presence of underlying CLE may only be suspected after CHD repair. In our case, a 2-month-old infant presented with respiratory distress. On evaluation, a large ventricular septal defect and patent ductus arteriosus were detected. After successful cardiac defect repair, the infant continued to experience cough, respiratory distress, and failure to thrive. Furthermore, on radiological reanalysis, CLE was suspected on X-ray and confirmed via computed tomography. CLE can be detected on chest X-rays; however, at an early stage, it is often confusing and misleading.

KEYWORDS

congenital heart disease, failure to thrive, lobectomy, respiratory distress

1 | INTRODUCTION

Congenital lobar emphysema (CLE) is a rare and life-threatening lung anomaly characterized by the hyperinflation of usually one or sometimes more lung lobes. Its incidence is ~1 in 20,000–30,000 live births with a 3:1 male preponderance.[1,2] The hyperinflated lobe compresses the surrounding normal lung tissue causing atelectasis, leading to ventilation/perfusion mismatch and hypoxia. In early infancy, it usually manifests with respiratory distress.[3,4] The most common lobe involvement is that of the left upper lobe (43%), followed by the right middle lobe (32%) and right upper lobe (21%). Congenital heart defects (CHDs) can occur in approximately 14%–20% of patients

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with CLE, and ventricular septal defect (VSD) is the most commonly associated anomaly.[1,4] The clinical presentation of CLE and most CHDs are similar and overlapping in the infantile age, with CHDs often being detected early as they are more common. CLE diagnosis is often overlooked in the presence of CHDs.[5,6] Herein, we reported a case where the presence of CLE was established following a corrective repair of a large perimembranous VSD with moderate patent ductus arteriosus (PDA).

2 | CASE PRESENTATION

A 54-day-old male child presented with cough, tachypnea, and chest retraction for 2 days before admission. The child did not have fever or any other complaints on arrival.

The child's birth and antenatal histories were unremarkable. He was born full term to a primi mother via spontaneous vaginal delivery, cried immediately, and required routine newborn care only. His birth weight was 2.6 kg. The child was brought to another hospital for the aforementioned complaints. Blood examinations and chest X-ray were performed, and the infant was diagnosed with right upper lobe pneumonia. He was started on an injectable antibiotic; however, because of worsening tachypnea and chest retraction, the infant was referred to our hospital. On arrival, the child's vital signs were as follows: temperature: 98.2°F; pulse rate: 154/min, palpable with warm periphery; capillary refill time: <2 s; respiratory rate: 72/min with subcostal and intercostal retractions; and oxygen saturation: 91% at room air and 97% with 2 L of oxygen via nasal prong. The child did not present with any external congenital deformity or facial dysmorphism. No adventitious sound on respiratory auscultation or hepatosplenomegaly were noted. However, grade 2/6 pan-systolic murmur was observed at the left parasternal region at the third intercostal space with a loud S2. The three-generation family history did not indicate the presence of any congenital heart or lung anomalies. Immunizations at birth were administered. The child's weight and height on admission were 3.5 kg and 52 cm, respectively, and both parameters were <3rd percentile. His birth weight was 2.6 kg, suggesting poor weight gain. The child was on mixed feeding (breast milk + cow milk) from 1 month of age. The chest X-ray suggested right upper lobe consolidation (Figure 1).

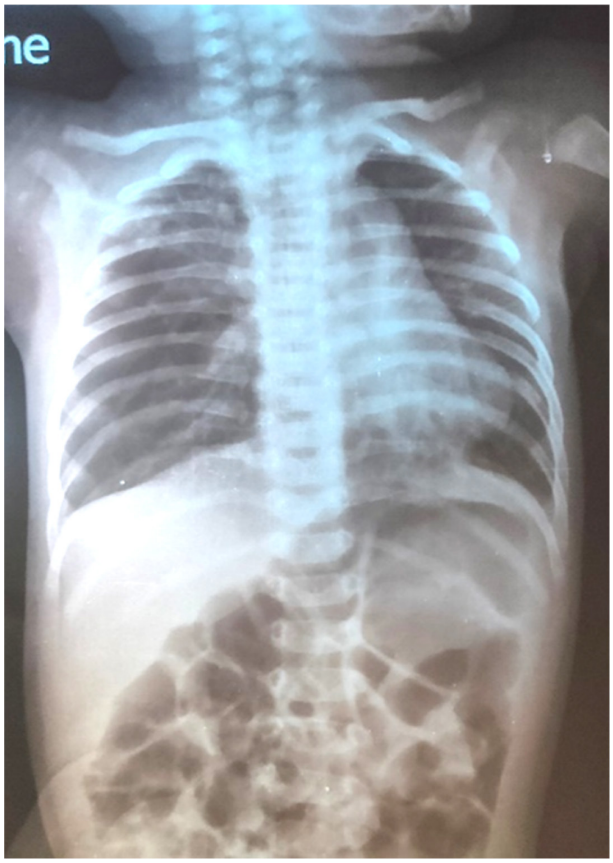

FIGURE 1 Posteroanterior view chest X-ray at 2 months of age interpreted as right upper zone consolidation.

The child was kept on low-flow oxygen, and an empirical antibiotic was initiated. The investigations indicated arterial hypoxia (paO_2: 61 mm Hg) and minimal respiratory acidosis (pCO_2: 48 mm Hg) with compensatory metabolic alkalosis (HCO_3: 29 mEq/L). The blood counts and C-reactive protein (CRP) levels were within the normal ranges. Moreover, serum electrolyte, serum creatinine, liver enzymes, and blood glucose were within the normal limits.

Blood culture was reported as sterile on the third day. Two dimensional (2D) echocardiography revealed large perimembranous VSD with moderate PDA and severe pulmonary arterial hypertension. Anti-heart failure medications (furosemide and spironolactone) were administered, and definitive repair was planned after sufficient weight gain. Oxygen was weaned off, and the child was discharged after 7 days on exclusive breastfeeding.

On regular subsequent follow-ups, cough and tachypnea were persistent and growth failure was also noted. Thus, definite cardiac surgery was performed at 3 months and 7 days of age. The patient underwent glutaraldehyde-treated pericardial patch closure of VSD with PDA ligation. The surgical procedure was uneventful.

Following the definitive cardiac surgery, cough, tachypnea, occasional retraction, and growth failure continued. During the subsequent follow-ups, the child was thoroughly examined, and the presence of an ongoing infection was considered unlikely owing to the absence of fever with repeat normal blood counts and CRP levels. The establishment of successful breastfeeding was ensured on follow-up; however, as the child continued to show

growth failure, a supplementary diet was recommended with appropriate micronutrient supplements. Without hepatosplenomegaly or neurological manifestations, the likelihood of a metabolic disease was low. Even repeat examinations demonstrated normal blood glucose, electrolyte, liver function, renal function, and acid–base balance. The child had no complaints of vomiting or abdominal distension. No features suggestive of persistent diarrhea or malabsorption were noted. The clinical examination did not suggest congestive cardiac failure. Serial echocardiography revealed no residual cardiac lesions alongside normal biventricular systolic function.

Thereafter, follow-up was lost for almost 3 months as the parents shifted to another geographic location. Follow-up was resumed at 7 months and 15 days of age with persistence of the above complaints. He had several admissions at another healthcare facility. He continued to grow poorly with a weight of 5.1 kg and a height of 62 cm, and both parameters were <3rd percentile. On repeat X-ray chest evaluation, right middle lobe hyperlucency and crowding of the right upper and lower lobe were noted (Figure 2).

Computed tomography (CT) confirmed the presence of CLE involving the right middle lobe (Figures 3 and 4).

(Image Credits: Dr. Viral B. Patel).

The lobectomy was planned after weighing the benefit–risk of surgical resection with a thorough literature review. Owing to the unavailability of double-lumen endobronchial tubes in this age group, a review of literature highlighted the use of single-lumen 3.5 Fr. endobronchial tube assisted with placement of a blocker in the form of 3 Fr. Fogarty catheter in the right intermediate bronchus after the take-off of right upper lobe bronchus facilitated using a 2.8 mm. ultra-thin fiber-optic bronchoscope. Right middle lobectomy was performed with baby in left lateral position. The right upper lobe and lower lobe surfaces were adhered together with two interrupted 5/0 polypropylene pledgetted sutures to obliterate the space post surgery. The specimen was sent for histopathological examination which was consistent with the radiological diagnosis of CLE (Figure 5A, B).

The clinical course was uneventful after surgery. The patient demonstrated a rapid recovery during the postoperative period and remained asymptomatic, following which the tachypnea and retraction resolved. Weight gain was noted during the subsequent follow-up. X-ray of the lungs revealed good expansion of the right lung with the upper and lower lobe well expanded, filling the gap of the middle lobe resection (Figures 6 and 7).

High-resolution computed tomography (HRCT) conducted on follow-up at 5 years of age revealed normally aerated lungs (Figures 8 and 9).

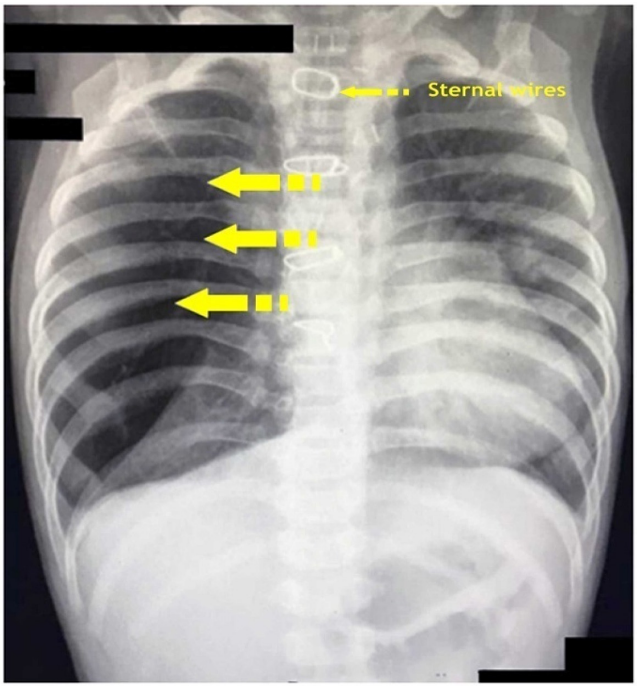

FIGURE 2 Chest X-ray Posteroanterior view demonstrating clear hyperlucency of the right middle lobe with crowding of the right upper and lower lobe with mediastinal shift. Sternal wires from the previous heart surgery can be observed.

3 | DISCUSSION

The association between CHDs and CLE is not unknown, and the large left-to-right shunts are commonly associated with CHD. However, tetralogy of Fallot and other right-to-left shunts are occasionally observed.[6] Cough, tachypnea, and respiratory distress in early infancy are the common clinical presentation in both the conditions: CLE and left-to-right shunts. In clinical practice, it is possible that CHD, which is considerably more common, is suspected and diagnosed first. Then, the entire clinical constellation is attributed to the CHD and the presence of CLE is only detected following the correction of the CHD.[6,7]

In almost half of the cases, the etiology of CLE remains unknown. Absent or dysplastic bronchial cartilage is evident in one-fourth of the cases. In the remaining cases, internal obstruction or external compression of the lobar bronchus due to various causes is postulated. These defects cause a ball valve effect and air trapping during expiration with the progressive hyperinflation of the affected lobe. The hyperinflated lobe may compress the surrounding normal lobes. In extreme cases, the hyperinflated lobe can herniate to the surrounding lobes and to the opposite thoracic cavity with tracheal and mediastinal shifts. This reduces the respiratory reserve

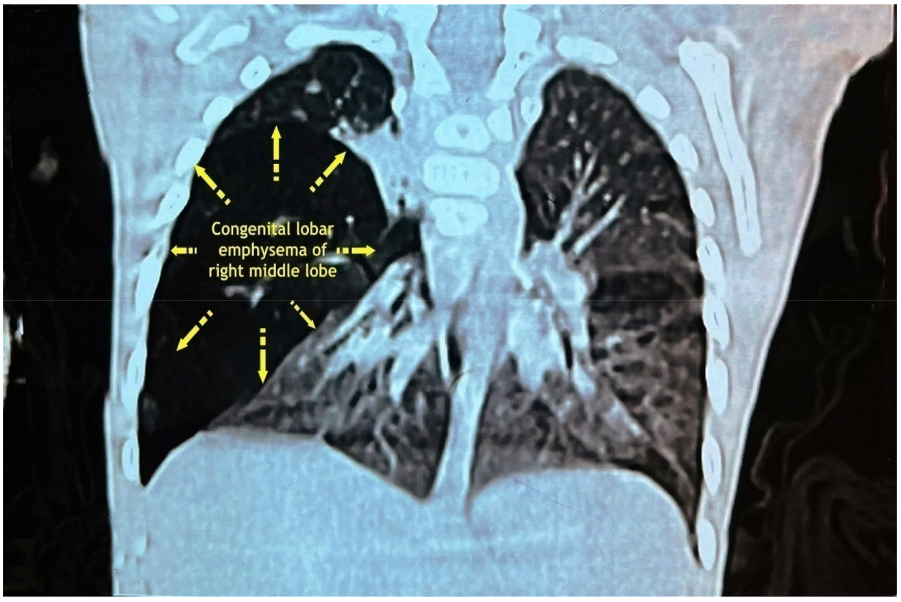

FIGURE 3 Preoperative coronal reformatted high-resolution computed tomography scan shows congenital lobar emphysema of the right middle lobe.

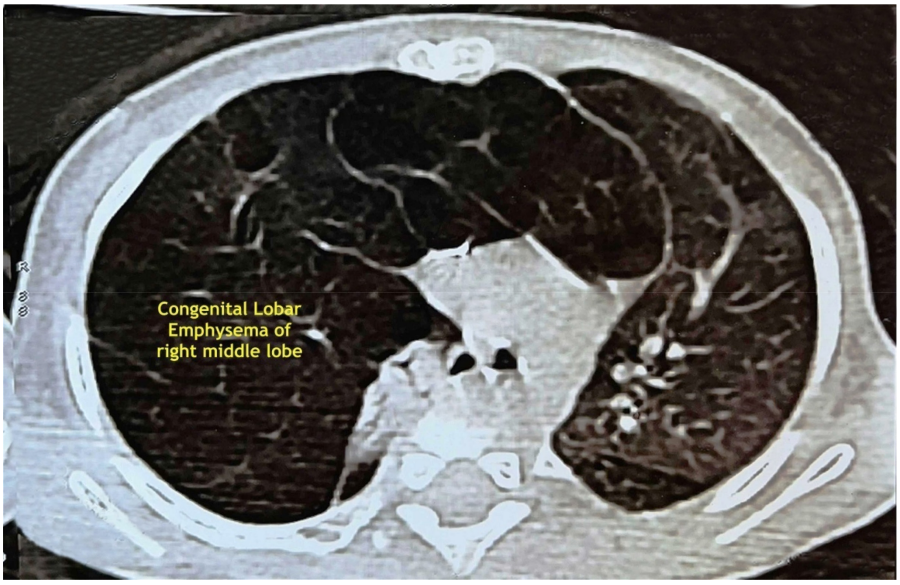

FIGURE 4 Axial view of the high-resolution computed tomography lung image confirming the presence of congenital lobar emphysema of the right middle lobe.

and causes ventilation / perfusion mismatch with resultant hypoxia.[1,3]

In the embryo, bronchial cartilage development occurs during the fourth to sixth intrauterine weeks. This time coincides with an important developmental stage of cardiac chamber septation. Thus, a defect during this stage can affect both heart and lungs.[1] Many a time, CLE is secondary to bronchial compression due to aberrant or dilated pulmonary vessels owing to pulmonary hypertension in the presence of a large left-to-right shunt.[5,6]

CLE continues to pose a diagnostic dilemma. The first diagnostic modality is the chest X-ray. The affected lung lobe is hyperlucent with atelectasis of the adjacent lung lobes secondary to compression. Furthermore, a mediastinal shift can be observed. However, all the features are not evident on X-ray at an early age. X-ray findings are often misreported as pneumonia owing to the crowding of the surrounding lobes, and the affected lobe hyperlucency may be reported as a pneumothorax. CT scan of the lung is confirmatory and delineates the anatomical details. Bronchoscopy may aid in diagnosis when the internal bronchial obstruction is suspected.[1–4]

The treatment for CLE involves the surgical resection of the affected lobe. Conservative management is

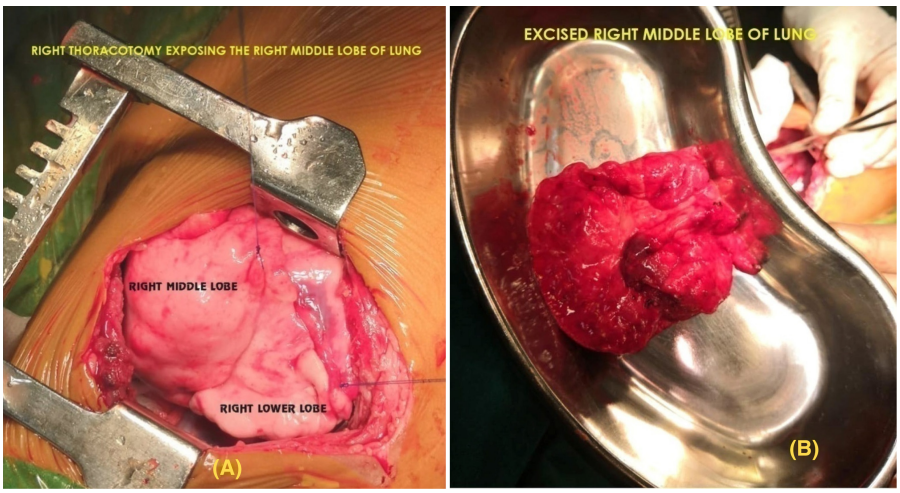

FIGURE 5 (A) Intraoperative image showing expanded right middle lobe; (B) excised right middle lung lobe.

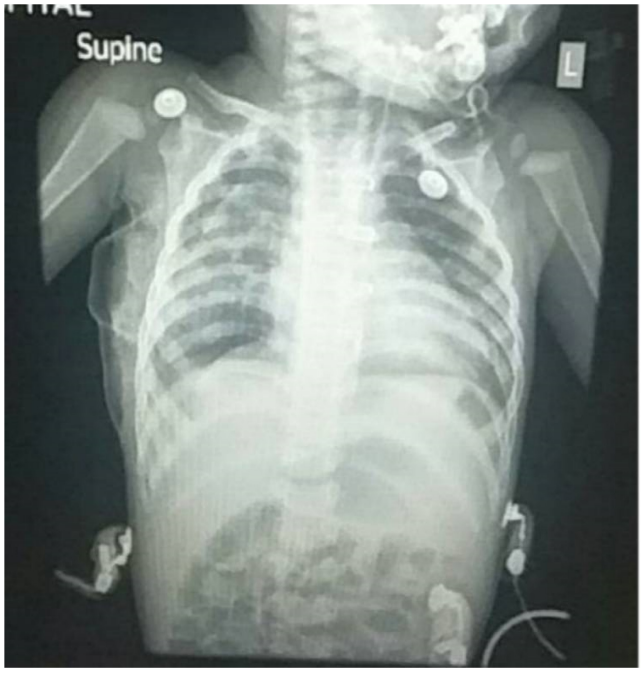

FIGURE 6 Immediate postoperative chest X-ray showing good expansion of the right lung and filling of the gap of the right middle lobe resection with right upper and lower lobes in the immediate postoperative period.

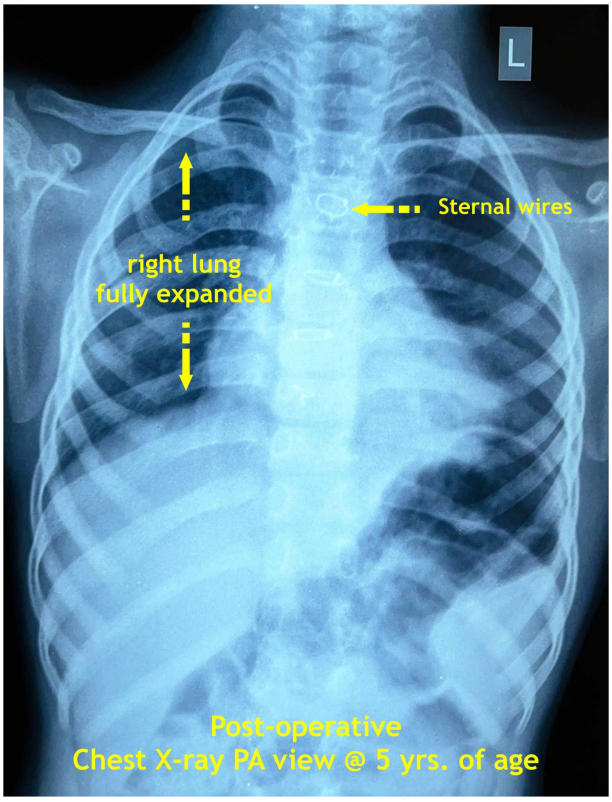

FIGURE 7 X-ray chest showing good expansion of the right lung and filling of the gap of the right middle lobe resection with the right upper and lower lobes at 5 years of age on follow-up.

proposed for older children with no or minimal symptoms. However, infants with persistent distress are considered ideal candidates for surgery. Surgery has been well-tolerated with low mortality and favorable outcomes to date.[2,8] In the case of concomitant CLE and CHD, there are diverging views on what defects to be repaired first or whether a combined lung and cardiac repair is the ideal approach. An individualized approach has been suggested. Many a time, CLE gets corrected with the relaxation of the vascular compression on the repair of the large left-to-right shunt and reduction of pulmonary hypertension.[5,9] Lobectomy can lead to drastic improvement and is well tolerated.[10]

4 | CONCLUSION

CLE is a diagnostic challenge, and the confusion can be further increased by the presence of an obviously large

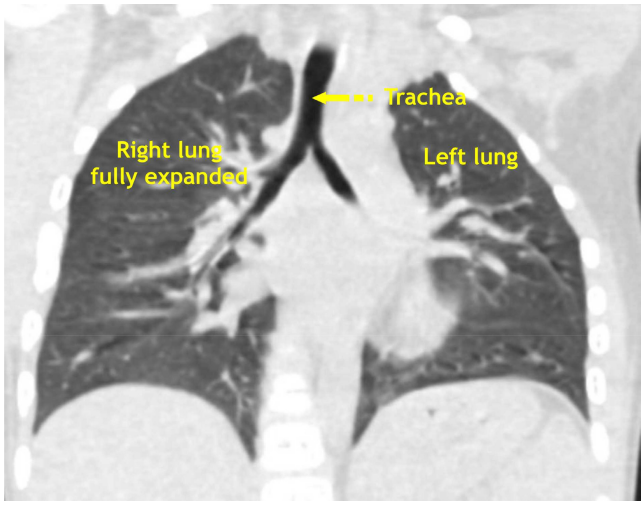

FIGURE 8 Postoperative coronal computed tomography image showing the normal lung parenchyma. (Image Credits: Dr. Viral B. Patel)

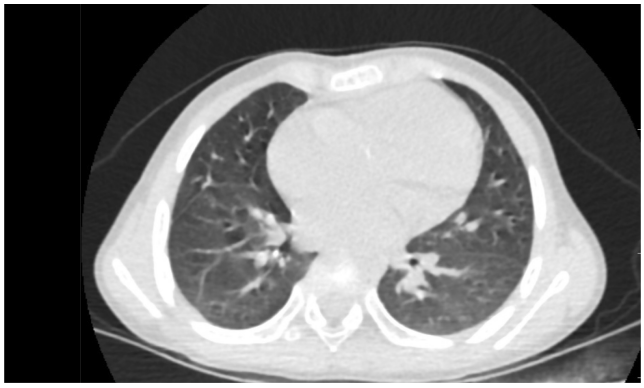

FIGURE 9 Postoperative high-resolution computed tomography axial image showing normally aerated lungs. (Image Credits: Dr. Viral B. Patel)

CHD. A more optimal chest X-ray observation may provide guidance; however, CT is the confirmatory imaging modality. Regular follow-up and a high index of suspicion are necessary for early diagnosis. Resection of the affected lobe may be a surgical treatment of choice.

AUTHOR CONTRIBUTIONS

Jigar Prabhulal Thacker: Conceptualization; data curation; formal analysis; funding acquisition; investigation; methodology; project administration; resources; software; supervision; validation; visualization; writing – original draft; writing – review and editing. **Vishal Vinayak Bhende:** Conceptualization; data curation; formal analysis; funding acquisition; investigation; methodology; project administration; resources; software; supervision; validation; visualization; writing – original draft; writing – review and editing. **Tanishq Shashikant Sharma:** Funding acquisition; project administration.

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CONFLICT OF INTEREST STATEMENT

All authors have declared that they have no financial relationships at present or within the previous 3 years with any organizations that might have an interest in the submitted work. All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are openly available in DOI: 10.22541/au.168261292.28643687/v1.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

TRANSPARENCY STATEMENT

The corresponding author confirms that manuscript is an honest, accurate, and transparent account of the study being reported; that no important aspects of the study have been omitted; and that any discrepancies from the study as planned have been explained.

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