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Case Report

Rare presentation of a rare disease: Bilateral congenital lobar overinflation ☆

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ABSTRACT

Congenital lobar overinflation is a rare but well-recognized congenital cause of neonatal and infantile respiratory distress. At times, the condition can mimic other congenital or acquired diseases and have atypical distribution and imaging patterns. Lobectomy of the involved lobe(s) is curative. We present our experience with 3 surgically confirmed cases of congenital lobar overinflation. Referral papers, patient's charts, including operation notes, and radiographic records were reviewed. All of them were initially misdiagnosed or underdiagnosed based on the initial radiographic examination alone. All 3 were referred to our center with respiratory distress, and the first 2 were treated with antibiotics prior to the settlement of their diagnosis. Chest computed tomography was key in diagnosing all 3 cases. The first patient was a 10-day-old neonate diagnosed with bilateral congenital lobar overinflation. The second patient was a 2-month-old infant diagnosed with right middle lobe disease. In these 2 cases, the initial assessment of the vascularity was atypically excessive in the affected lobe(s). Eventually, correlation with typical concurrent imaging features and the clinical condition of the patients led to the correct diagnosis. The third case was a 4-month-old infant with left upper lobe congenital lobar overinflation. All cases underwent successful surgical treatment. Congenital lobar overinflation is a rare anomaly, and multiple-lobe involvement is even rarer. Vascularity within the affected lobes is a subjective assessment that can be overestimated, leading to confusion, and a feature that needs correlation with other common imaging features and the clinical course of patients.

Introduction

Congenital lobar overinflation (CLO) is a rare congenital lung condition that causes selective hyperexpansion, usually of 1 segment or lobe [1–3]. It occurs between 1 in 20,000 to 30,000 live births [4]. The time of presentation, bilaterality, and coexisting defects affect clinical manifestations, which can range from asymptomatic or mild breathing difficulty to severe respiratory distress that begins at birth [2,3]. The cause is not

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clearly known, but multiple theories propose an abnormality of airway cartilages and compression by intrinsic vessels. The resulting hyperinflation and subsequent mass effect on adjacent lobes and/or mediastinum are responsible for the respiratory symptoms at presentation. Most patients are symptomatic at an early age, with the majority presenting at less than 6 months of age [1–3]. Usually, a single lobe is involved, with the upper and middle lobes more commonly involved and the lower lobes affected rarely. Bilaterality is exceptionally rare [5].

Imaging plays a crucial role in confirming the condition and separating it from other mimickers such as compensatory overinflation and pneumothorax. Chest radiography is the initial method that will show hyperlucency of the involved lung region and the resulting mass effect. Chest Computed Tomography (CT) is superior in showing the size of the involved lobe(s), compression of adjacent lobes (and, when severe, herniating to the contralateral hemithorax), other coexisting processes such as pneumonia, as well as other congenital cardiovascular conditions. The major imaging features include overinflation of the involved lobe(s), a resulting mass effect, and decreased vascularity in the involved lobes [1,5,6]. Attenuated vascularity is an additional finding that is important to carefully assess as it helps in differentiating congenital lobar overinflation from pneumothorax. For this too, chest computed tomography is mandatory and performs better than radiography, but it is not entirely unambiguous [15].

Case 1

A 10-day-old female neonate presented with breathing difficulties. Birth was at term via caesarian section for an indication of severe oligohydramnios at a local hospital. The baby had grunting and fast breathing since birth and was treated for presumed transient tachypnea of the newborn and early-onset neonatal sepsis with IV ampicillin and gentamycin and later on with the addition of IV ceftriaxone for a total of 9 days. The baby failed to improve and was referred to our center for better neonatology care. Records of radiologic investigations were not found on the referral paper. On presentation, the patient had a pulse rate of 172/min, a respiratory rate of 52/min, a temperature of 36.7°C, and was desaturating (88% on room air). On auscultation, there was bilaterally decreased air entry in both upper lung regions, more on the left side.

Chest radiography showed bilateral hyperlucency with relatively well-maintained bronchovascular markings. There was evidence of left lower lobe collapse, as evidenced by the silhouetted medial left hemidiaphragm. There was also right middle lobe increased translucency with right upper and lower lobe opacity (Fig. 1). Chest and abdominal ultrasounds were performed to evaluate the diaphragm and for further characterization of the lung parenchymal processes, and the right upper and bilateral lower lobe collapses were redemonstrated. With the clinical presentation and multifocal parenchymal densities, the possibility of infection was entertained as a possible cause. The possibility of CLO was not strongly considered at this point since bilateral presentation is

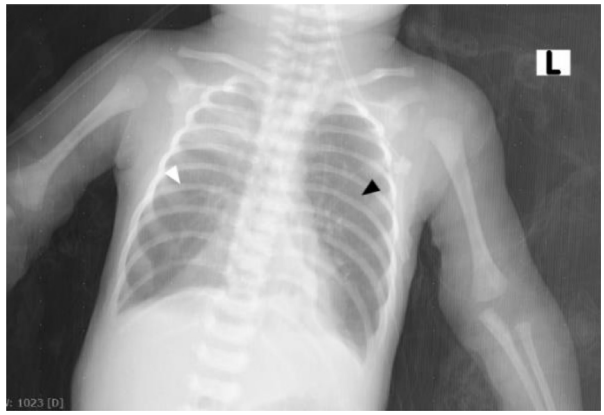

Fig. 1 – Supine chest X-ray shows hyperlucency of the right middle lung (white arrowhead) as well as the majority of the left lung (black arrowhead). Right upper lung as well as bilateral medial lower lobe sharply marginated opacities are also seen.

quite rare. Moreover, from the initial chest X-ray observation, the pulmonary vasculature within the hyperlucent regions appeared normal.

Septic doses of IV cefepime and ampicillin were given for 3 days, but the neonate did not improve and continued to desaturate persistently below 90% on 1 L of intranasal oxygen. Three days into her admission, a noncontrast chest CT was performed, which showed left upper lobe (LUL) and right middle lobe (RML) hyperinflation with resulting compressive atelectasis of the left lower lobe, right upper lobe (RUL), and right lower lobes (Fig. 2). Even if the diagnosis of CLO was not apparent from the initial radiography, the CT was more suggestive by clearly showing the marked hyperlucency of the affected lobes and the compressed adjacent lobes.

Subsequently, a right middle and left upper lobectomy were done. Intraoperative findings were consistent with the CT findings, showing a hyperinflated RML and LUL that herniated through right and left posterolateral thoracotomy incisions. Atelectasis of RUL and right lower lobes were also seen. Postoperatively, the baby had a smooth course and was discharged in stable condition. At 2 weeks and 1 month of follow-up visits after discharge, the infant was in good condition and was recovering well.

Case 2

A 2-month-old male infant was referred to us with a complaint of fast breathing since birth. He was born at term by spontaneous vaginal delivery, which was complicated by prolonged duration of labor (lasting 20 hours). The initial APGAR score was not known. The chest radiograph report from the referring hospital stated a right upper lung region opacity. In addition, blood culture was positive for *Escherichia coli*. These subsequently led to a treatment of neonatal sepsis with IV ampicillin, gentamycin, and later on vancomycin and ciprofloxacin for a total of 22 days, but the infant's respiratory distress did

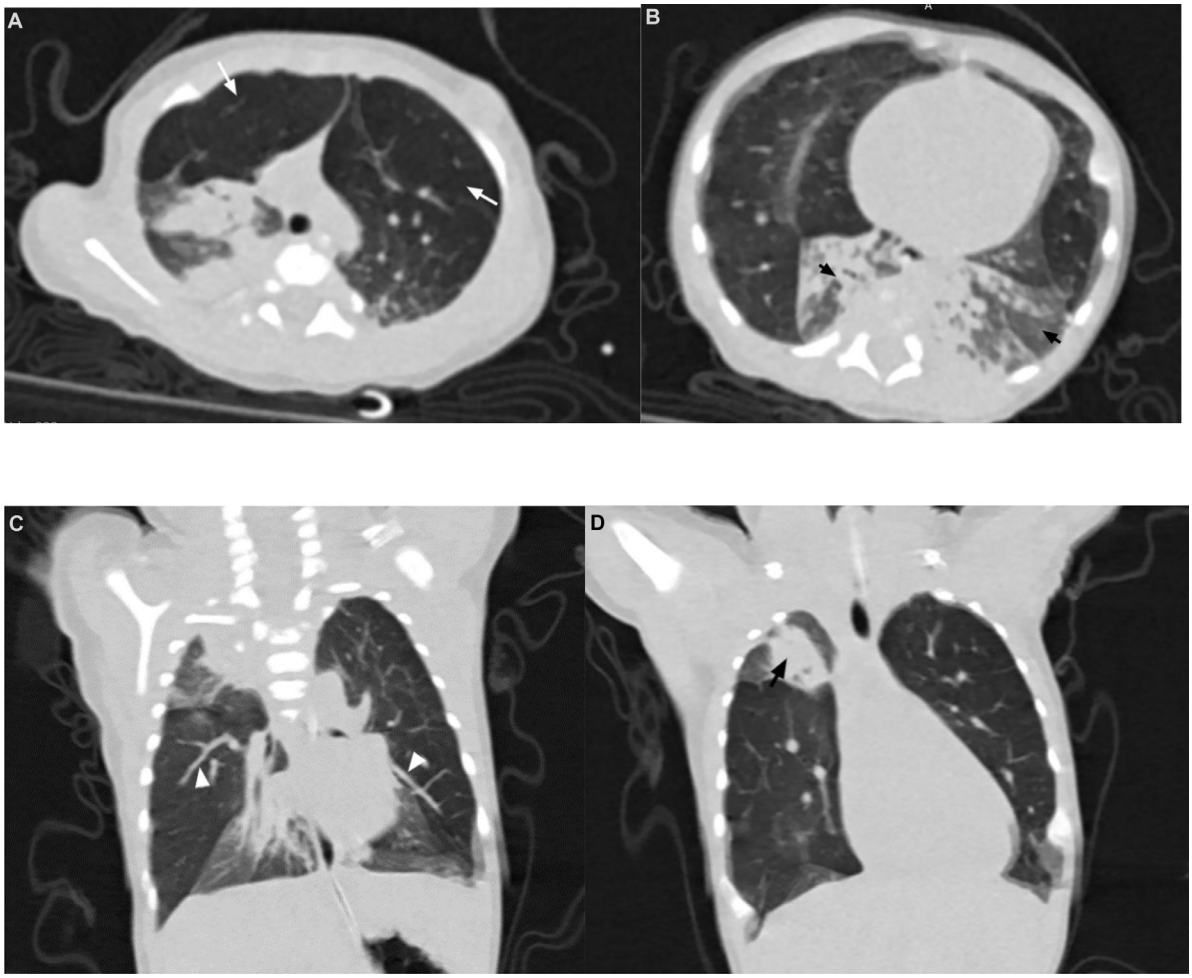

Fig. 2 – Three days later, axial cranial (A) and caudal (B) sections of lung window chest CT show air trapping in the right middle and left upper lobes (white arrows in A) with bibasal lung collapses (black arrows in B). Coronal views (C and D) show RML and LUL disease distribution to better advantage. White arrowheads (in C) point to subjectively prominent appearing vasculature. Black arrow (in D) shows the right upper lung collapse with a focus of consolidation.

not improve, and he was subsequently transferred to our center. On arrival, examination showed the child in acute respiratory distress with a pulse rate of 152/min, a respiratory rate of 72/min, a temperature of 37.2°C, and an oxygen saturation of 85% on room air. Chest examination showed severe inter- and subcostal retractions and decreased air entry in the upper two-thirds of the right chest.

For a detailed evaluation of the chest, a noncontrast chest CT scan was done and showed a hyperinflated right middle lobe that herniated across the midline and a resulting mediastinal shift to the left. Even if the lobe was hyperinflated, the pulmonary vessels were not attenuated. The right and left upper lobes showed evidence of collapse and consolidation (Fig. 3). As a result, an acquired compensatory overinflation due to the RUL pneumonic consolidation was considered. The patient continued with a revised course of antibiotics consisting of IV cefepime for 11 days, but to no avail. A repeat chest X-ray was done 14 days after the CT scan for lack of improvement and showed a persistently hyperlucent right mid-lung after marked improvement of the RUL consolidation (Fig. 3E). Thus, a right middle lobe CLO was diagnosed as it best represented the abnormality. The initial hesitation in considering CLO was due to the fairly normal calibre of the pulmonary vessels in the affected lobe. A right middle lobectomy was done through a right posterolateral thoracotomy incision, which showed an overinflated RML. The child was subsequently discharged in stable conditions. The patient was stable at 2 weeks and 1 month of follow-up visits.

Case 3

A 4-month-old female infant was referred to us with a 1-month history of a nonwhooping and nonbarking type of progressively worsening cough. She was relatively asymptomatic during her first 3 months of life and had an uneventful perinatal course. On examination, the infant was desaturating (initially 86% on room air, which normalized to 98% with 2 L intranasal oxygen), had a pulse rate of 140/min, a respiratory

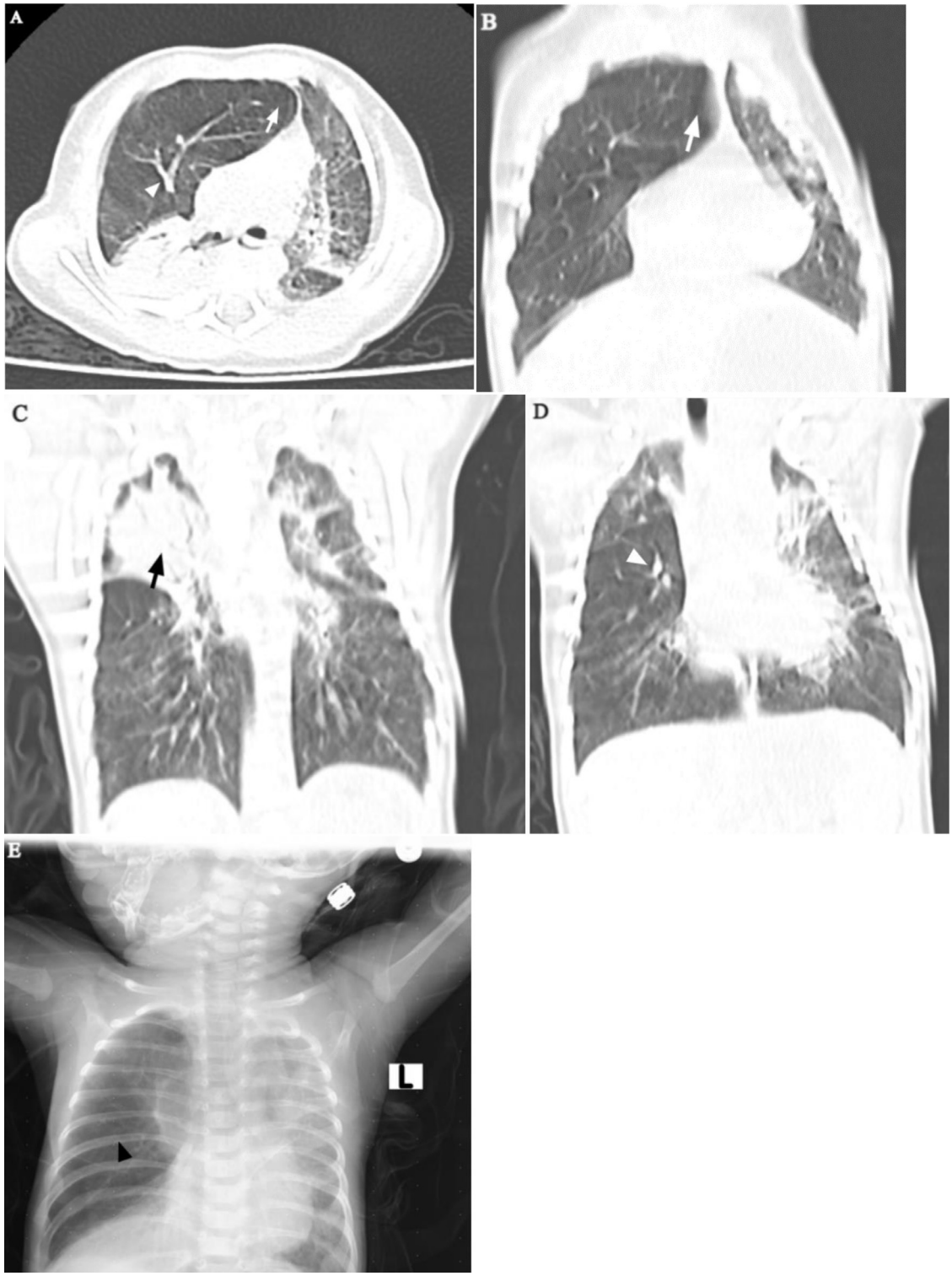

Fig. 3 – Axial (A) and successive coronal views (B-D) of chest CT in lung window show hyperinflated RML with left ward herniation through the anterior mediastinum (white arrows). Caliber of vessels are relatively preserved (white arrowheads in A and D). Consolidation is seen in the posterior segment of the right upper lobe (black arrow in C). Chest radiograph (E) acquired 14 days after the chest CT shows resolution of consolidation but persisting large area of lucency (black arrowhead) in right lung consistent with RML overinflation.

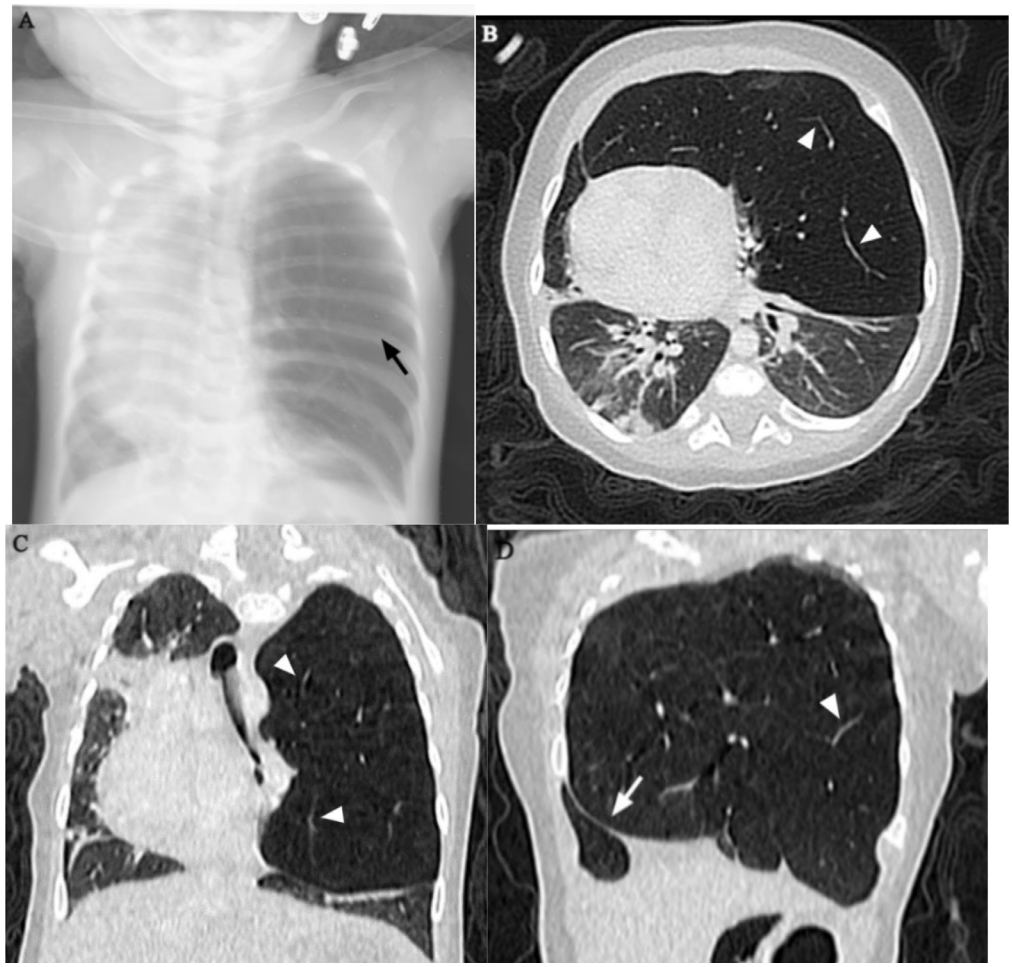

Fig. 4 – Initial supine chest radiograph (A) shows markedly hyperlucent and hyper inflated left upper and mid lung regions with compressive atelectasis inferiorly and marked right ward mediastinal shift (arrow). Axial (B), and consecutive posterior (C) and anterior (D) coronal sections of chest CT in lung window show severe hyperinflation of the LUL with decreased vasculature (white arrowheads in B-D). On the coronal images, marked right ward tracheal and mediastinal shift is also noted with associated mass effect on the right lung. The arrow (in D) points to severely shifted anterior junctional line.

rate of 54/min, and a temperature of 35.6°C. On auscultation, there was diffusely decreased air entry over the left chest.

Radiologic findings were consistent with left upper lobe CLO, and a chest radiograph showed overinflation of the upper two-thirds of the left lung with atelectasis of the left lower lung and contralateral mediastinal shift. The findings were confirmed by a chest CT done the next day (Fig. 4). A left posterolateral thoracotomy was performed with intraoperative findings of an overinflated LUL. The patient was eventually discharged in stable condition. The infant was healthy at her 2-week follow-up, and then care was transferred to another hospital that is close to the patient's hometown.

Discussion

We reviewed 3 surgically confirmed cases of CLO in 2 females and 1 male patient. One had bilateral CLO involving the LUL and RML, and the others had single lobe involvement of the RML and LUL. CLO occurs between 1 in 20,000 to 30,000 live births [4]. It usually affects a single lobe, with the LUL (43%) and RML (32%) being the 2 commonly involved lobes. RUL (21%), and lower lobe (2%) are less common [2,5]. The LUL and RML are the only 2 lobes affected in all of our cases. Bilaterality is exceptionally rare, with 26 cases reported so far, according to our knowledge. Garge et al. [6] reported 3 cases from the author's institution and reviewed 22 previous cases from 14 reports since 1963. Zinaye and Zeray [7] from Ethiopia also reported a single case of bilateral CLO in 2022. The combination of LUL and RML involvement appears to be more common (17 cases out of 26 compiled by Garge et al. [6] had this combination). The treatment and recovery, as expected, will be more challenging as bilateral thoracotomies carry a higher chance of complications.

CLO, as shown in our cases, causes overinflation of 1 or more lobes, with subsequent mass effects on the adjacent lobes. The cause of air trapping in 50% of the cases is un-

known. The proposed mechanism includes bronchial cartilage dysplasia, extrinsic compression by aberrant vessels, and intrinsic abnormalities like polyalveolar lobes [8,9]. Due to the effect such an overinflation can have on adjacent and contralateral lung tissue and the mediastinum, it can lead to hypoxia that may require emergency intervention. Most patients present within their first 6 months of life with shortness of breath and wheezing [1–3]. Consistent with this epidemiologic profile, 2 of our cases presented immediately after birth and 1 at 4 months of age. Especially among newborns, the condition often mimics an infectious process, and as such, patients might be initially treated with a course of antibiotics before CLO is suspected from further radiologic examinations performed for poor treatment response [5,10–13]. This sequence of events took place in 2 of our cases. CLO can be potentially confused with pneumothorax, as both conditions appear hyperlucent on radiography. Erroneous chest drainage insertions have indeed been reported. Prabhu et al. [15] highlighted this important challenge in their review of inadvertent chest tube insertion among children with congenital cystic lucent lung lesions. Five such occurrences were found in CLO patients who presented with variable degrees of dyspnea. All cases were diagnosed with tension pneumothorax from initial radiographs, which showed hyperinflated lung fields and mediastinal shift. The temptation to insert chest tubes is understandable, as the imaging findings are compatible with the presumed diagnosis. Despite the attempt, respiratory distress worsened in all, and surgical emphysema and a new moderate pneumothorax developed in 3. Consequently, nonimprovement led to a chest CT examination, which properly diagnosed CLO. The writers admit faint vascular markings (within the hyperlucent areas) were seen on the initial radiographs and stress the importance of carefully assessing the level of vascularity on initial radiographs to differentiate pneumothorax from overinflated lung. Similar sequence of events (initial misdiagnosis of tension pneumothorax based on initial radiographs leading to unnecessary interventions with subsequent clinical worsening and eventual corrected diagnosis based on CT) were also reported by other authors [14,16].

Imaging provides crucial diagnostic input by showing the affected lobe(s), adjacent mass effects, concurrent infection, associated congenital heart disease, as well as ruling out obstructive pathology. The cardinal postnatal imaging features on radiography and CT are segmental or lobar overinflation which will persist or progress on subsequent imaging, hyperlucency with compression or displacement of adjacent lobes, and when severe, the mediastinum shift as seen in our third case [1–5].

A chest radiograph is the initial imaging modality, as it is safe and conveniently available in most neonatal care settings. It is of paramount importance to pick the mediastinal shift early and suggest the diagnosis. The cardinal and gold standard study, though, is chest CT, as it is mandatory for a detailed evaluation of the whole chest, the lobar involvement, as well as concurrent complications, clearly and early. As discussed above, all our cases were confirmed by chest CT. One additional imaging finding that is more easily assessed with chest CT is related to the appearance of the vasculature within the affected lobes. Attenuated vasculature has been proposed as a helpful clue to differentiate CLO from its mimickers [12,17,18]. But we did not find any objective imaging criteria to measure decreased vasculature in CLO, and thus we used subjective assessment. In 2 of our cases, the CT scan showed vascularity that was not markedly decreased. In our isolated RML case, the superimposed pneumonia and the seemingly prominent vessels made us less considerate of CLO. In this case, the lucent region was judged to be caused by compensatory hyperinflation related to RUL pneumonia. In our bilateral case too, the extreme rarity of bilateral CLO in addition to the fairly normal calibre of the pulmonary vessels on the initial radiography made us doubt the diagnosis of CLO. But on the chest CT, even though the vessels still appeared preserved, a detailed evaluation of the other cardinal signs of CLO—overexpanded and hyperlucent lobes with adjacent mass effect—persuaded us to the diagnosis of CLO. Admittedly, the worsening respiratory distress of the child also drove us to consider CLO as a strong possibility. The underrecognization of bilateral CLO is consistent with previous experiences from different authors. In the bilateral CLO case reported by Zinaye and Zeray [7], the initial consideration was either a right lung hypoplasia with compensatory hyperinflation of the left lung or a left lung hyperinflation due to a vascular ring. Eventually, a chest CT settled the diagnosis. In the review by Garge et al. [6], recurrent admissions for pulmonary infections and asthma were considered in some of the reviewed cases before chest CT shed light on the possibility of bilateral CLO.

The level of vascular attenuation can be underestimated on the initial radiograph, and it can usually be better recognized on follow-up imaging that shows the persisting abnormality. Kennedy et al. [12] showed that the number of radiographs that correctly displayed decreased vessel calibre on the affected lobe increased from 7 to 10 between the initial and final chest X-rays. Given this, a wholesome consideration of all the imaging features and strong clinical suspicion are needed for an early and correct diagnosis.

Thoracotomy of the involved lobes, either thoracoscopically or by open surgery, based on local expertise, is the definite treatment modality, and it is vital to prevent ensuing complications. The procedure needs to be done as early as possible in severely symptomatic cases. In selected stable and asymptomatic cases, close observation can be chosen in the hopes of avoiding surgery. Standard frequency and duration for follow-ups are not specified, but generally symptoms lead management[1,10,19]. All our cases underwent an open thoracotomy within the same admission period and had a smooth postoperative course.

In conclusion, CLO can have varied clinical presentations with its diagnostic challenges related to the possibility of infection and pneumothorax. Even if single-lobe involvement is the most common presentation, multiple-lobe involvement is still possible. A chest CT scan is an indispensable imaging modality for the overall assessment of CLO and, in particular, for the detailed description of the classic signs. Although these are usually clearly depicted, a dilemma can arise regarding the level of diminished vascularity in the overinflated lobes on either chest radiography or chest CT. For this reason, a wholesome tactic based on clinical and radiologic findings is necessary.

Patient consent

Written informed consent was obtained from the parents of the 3 presented children for anonymized patient information to be published in this article.

REFERENCES

[1] Farling P. Anesthesia and uncommon diseases. Br J Anaesth 2006;97(6):904.
[2] US Elsevier Health [Accessed 29.7.23]. Caffey's pediatric diagnostic imaging, Available from: https://www.us.elsevierhealth.com/caffeys-pediatric-diagnostic-imaging-2-volume-set-9780323497480.html.
[3] Tural-Kara T, Özdemir H, Çiftçi E, İnce E. Congenital lobar emphysema. Saudi Med J 2016;37(7):796–8.
[4] Demir OF, Hangul M, Kose M. Congenital lobar emphysema: diagnosis and treatment options. Int J Chron Obstruct Pulmon Dis 2019;14:921–8.
[5] Ozçelik U, Göçmen A, Kiper N, Doğru D, Dilber E, Yalçin EG. Congenital lobar emphysema: evaluation and long-term follow-up of thirty cases at a single center. Pediatr Pulmonol 2003;35(5):384–91.
[6] Garge S, Mahalik S, Jain P. Bilateral congenital lobar emphysema: a rare entity and a therapeutic challenge. Ann Pediatr Surg 2022;18(1):91.
[7] Zinaye A, Zeray A. Bilateral congenital lobar overinflation (CLO) - a rare presentation of an uncommon condition. Radiol Case Rep 2022;17(10):3481–4.
[8] Warner JO, Rubin S, Heard BE. Congenital lobar emphysema: a case with bronchial atresia and abnormal bronchial cartilages. Br J Dis Chest 1982;76:177–84.
[9] Tapper D, Schuster S, McBride J, Eraklis A, Wohl ME, Williams A, et al. Polyalveolar lobe: anatomic and physiologic parameters and their relationship to congenital lobar emphysema. J Pediatr Surg 1980;15(6):931–7.
[10] Bawazir OA. Congenital lobar emphysema: thoracotomy versus minimally invasive surgery. Ann Thorac Med 2020;15(1):21.
[11] Tempe D, Virmani S, Javetkar S, Banerjee A, Puri S, Datt V. Congenital lobar emphysema: pitfalls and management. Ann Card Anaesth 2010;13(1):53.
[12] Kennedy CD, Habibi P, Matthew DJ, Gordon I. Lobar emphysema: long-term imaging follow-up. Radiology 1991;180(1):189–93.
[13] Cataneo DC, Rodrigues OR, Hasimoto EN, Schmidt AF, Cataneo AJM. Congenital lobar emphysema: 30-year case series in two university hospitals. J Bras Pneumol 2013;39(4):418–26.
[14] Congenital lobar emphysema simulating a pneumothorax | Anales de Pediatría [Accessed 13.8.23]. Available from: https://www.analesdepediatria.org/en-congenital-lobar-emphysema-simulating-pneumothorax-articulo-S2341287923000947.
[15] Prabhu SM, Choudhury SR, Solanki RS, Shetty GS, Agarwala S. Inadvertent chest tube insertion in congenital cystic adenomatoid malformation and congenital lobar emphysema-highlighting an important problem. Indian J Radiol Imaging 2013;23(1):8–14.
[16] Reed A, Lucas SF, Nowacka A, Eze C. Iatrogenic pneumothorax in a 4-week-old girl: new diagnosis of congenital lobar emphysema. BMJ Case Rep 2020;13(2):e233302.
[17] Daltro P, Fricke BL, Kuroki I, Domingues R, Donnelly LF. CT of congenital lung lesions in pediatric patients. Am J Roentgenol 2004;183(5):1497–506.
[18] Thakral CL, Maji DC, Sajwani MJ. Congenital lobar emphysema: experience with 21 cases. Pediatr Surg Int 2001;17(2–3):88–91.
[19] Karnak I, Senocak ME, Ciftci AO, Büyükpamukçu N. Congenital lobar emphysema: diagnostic and therapeutic considerations. J Pediatr Surg 1999;34(9):1347–51.