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CASE REPORT

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The Breathing Struggle: A Case Study of Congenital Lung Malformation in a Young Child

Asra Moradkhani[1], Ali Abasi[1], Fatemeh Behbahani Nejad[1], Rasoul Nasiri Kalmarzi[2] and Shilan Mohammadi[3*]

Abstract

Background Congenital lung malformations (CLMs) are among the rare anomalies that can be diagnosed by bronchoscopy and imaging. They can cause various respiratory symptoms and complications, especially in children with congenital heart disease. This is an interesting case report of a child with a rare combination of congenital anomalies affecting the airway.

Case presentation We report a case of a 3.5-year-old boy with multiple congenital anomalies and respiratory problems since birth. He had a history of mild autism, developmental delay, and sensitivity to smell and smoke. He presented with hoarseness, shortness of breath, severe coughing, and severe wheezing, which worsened with the flu. He underwent bronchoscopy and other diagnostic tests, which revealed a posterior laryngeal cleft, a tracheal bronchus, and a very narrow distal trachea. He was treated with nebulizers, antibiotics, and serum therapy and showed improvement. This case illustrates a rare combination of airway malformations that require a multidisciplinary approach.

Conclusions We presented a case of rare pulmonary malformations and chronic respiratory symptoms that improved with conventional pharmacotherapy. Increased awareness and understanding of these anomalies among healthcare providers can lead to earlier diagnosis and improved patient outcomes.

Keywords Congenital lung malformation, Posterior laryngeal cleft, Tracheal bronchus, Case report

Introduction

Congenital tracheobronchial malformations (CTMs) are found in some cases that undergo bronchoscopy, a procedure that allows direct inspection and sampling of the tracheobronchial tree. The use of imaging techniques has enhanced the diagnosis of CTMs [1]. The tracheal bronchus (TB), also known as bronchus suis, was first described in 1785 and represents a rare congenital anomaly characterized by additional tracheal growth during fetal development; it may manifest on either side of the trachea [1–3]. TB originates from the lateral side of the trachea above the carina and provides air to the upper lobe [4]. Posterior laryngeal clefts (PLCs) were

*Correspondence:
Shilan Mohammadi
Shilanm3256@gmail.com
[1]Student of the Research Committee, Kurdistan University of Medical Sciences, Sanandaj, Iran
[2]Cellular & Molecular Research Center, Kurdistan University of Medical Sciences, Sanandaj, Iran
[3]Pediatrics Department, Medical Faculty, Kurdistan University of Medical Sciences, Sanandaj, Iran

first reported by Richter in 1792 [5]. This anomaly results from the failure of fusion of the tracheoesophageal septum or the dorsal aspect of the cricoid ring during the embryonic stage [6].

According to bronchoscopic and bronchographic findings, right TB is more prevalent than left-sided (0.1-2% vs. 0.3-1%) [1, 7]. Also, it is more common in men compared to women [2, 8]. It is often asymptomatic and diagnosed incidentally through a chest CT scan or during bronchoscopy, but it may present with symptoms such as respiratory distress, stridor, tachypnea, cough, wheezing, pneumonia, air trapping, and atelectasis [9–11]. In this report, we present a child complaining of a severe cough and shortness of breath. During the work-up, the child was diagnosed with a posterior laryngeal cleft (grade 1), tracheal bronchus, and very narrow distal trachea.

Patient information

The patient, a three-and-a-half-year-old boy, presented with complaints of hoarseness, shortness of breath, severe coughing, and severe wheezing since birth. Also presented with clubbing, cyanosis, and failure to thrive (FTT). Despite these signs and symptoms, no diagnostic evaluations were conducted by any of the healthcare professionals consulted. The patient received conservative management based solely on a presumptive diagnosis of asthma. His condition had worsened significantly over the past few months. As a result, his parents have referred him to this center for additional diagnostic tests. The patient's companion reported that the flu worsened the symptoms and that the patient had a history of night snoring and wheezing, was unable to speak fully, and spoke in fragments. The parents also mentioned a history of mild autism and that the patient had a drug history of amoxicillin, azithromycin, adrenaline, and multivitamins. He also had an allergy to the smell of perfume and smoke. The patient was the second child in the family from a 32-year-old mother who had a cesarean section due to placenta previa. Due to the low first-minute Apgar score, the patient was given oxygen therapy for two hours. Due to the developmental delay, he started walking alone at 15 months of age and held his head steady at 9 months of age. Based on the information provided by the patient's mother, it appears that he has difficulty learning.

In the physical examination, the patient was alert. Vital signs included a blood pressure of 100/70 mmHg, a pulse rate of 140, a temperature of 37, and a respiratory rate of 30. And had an oxygen saturation of 96%. The throat examination revealed erythema. In the examination of the chest, crackles and expiratory wheezes were heard. He had slight intercostal and subcostal retraction. The auscultation of the heart was normal, and the abdomen and organs were also examined.

The heart function was normal in the echocardiography, and mild TR was found. In the initial examination, the patient underwent a chest X-ray, which revealed an abnormal finding of a separate lung tissue mass. (Fig. 1) A chest CT scan provided a more detailed visualization of the airway anatomy. (Fig. 2) Before this, the patient underwent a flexible bronchoscopy (Fig. 3), which revealed three bronchial openings. Moreover, the bronchoscopy showed a PLC (grade 1) (Fig. 4), TB, and a significant narrowing of the distal trachea. The barium swallow was normal. (Fig. 5) The schematic image of

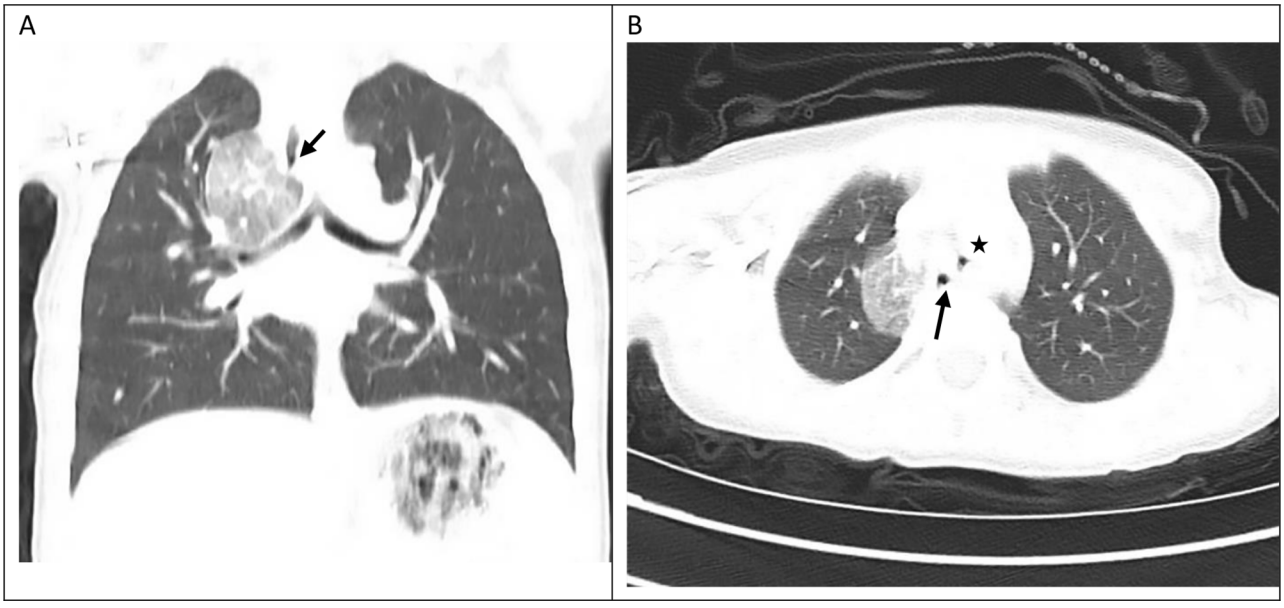

Fig. 1 Chest CT scan findings in a patient with right TB and a distinct parenchyma. **A**. Coronal view showing an arrow pointing to the TB. **B**. Axial view showing an arrow pointing to the TB and a star marking the trachea

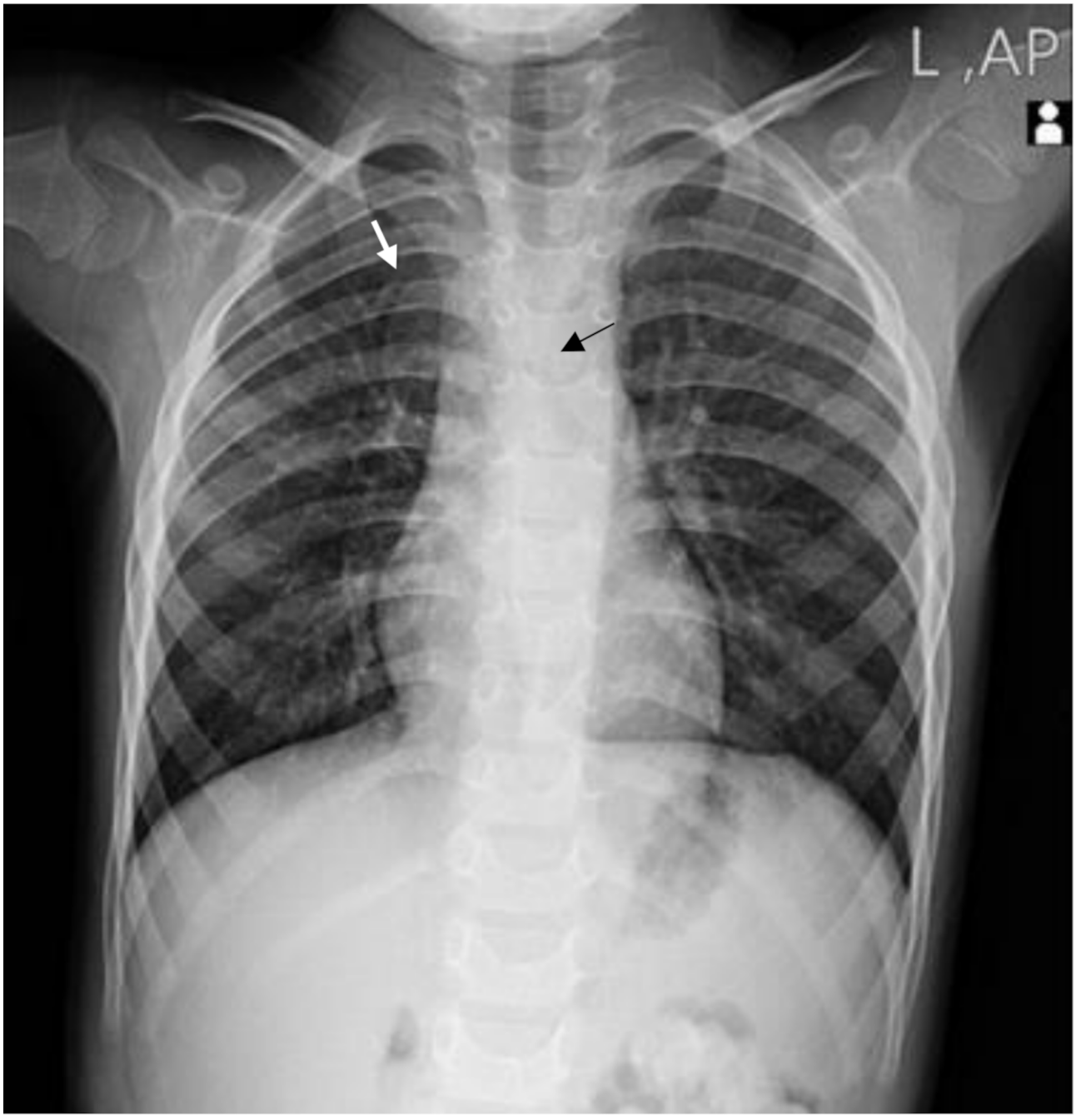

Fig. 2 Chest X-ray findings in a patient with an accessory lung lobe. The white arrow points to an abnormal separate tissue of the lung parenchyma in the right lower lobe. The black arrow shows the approximate image of the narrowing of the distal trachea

the lung and abnormalities related to the studied case is given. (Fig. 6) In this center, the patient was treated with an Epinephrine nebulizer, Pulmicort nebulizer, and Atrovent spray, as well as Ceftriaxone. During hospitalization, the patient experienced a slight decline in overall health status following the administration of the aforementioned treatments. Due to the limited resources and the surgical indication, the patient could not receive further treatment at this center; he was referred to the surgery department for the following treatment, though we have no details on whether any interventional procedure was done. He was discharged with instructions on the warning signs and medication regimen and advised to follow up at the hospital.

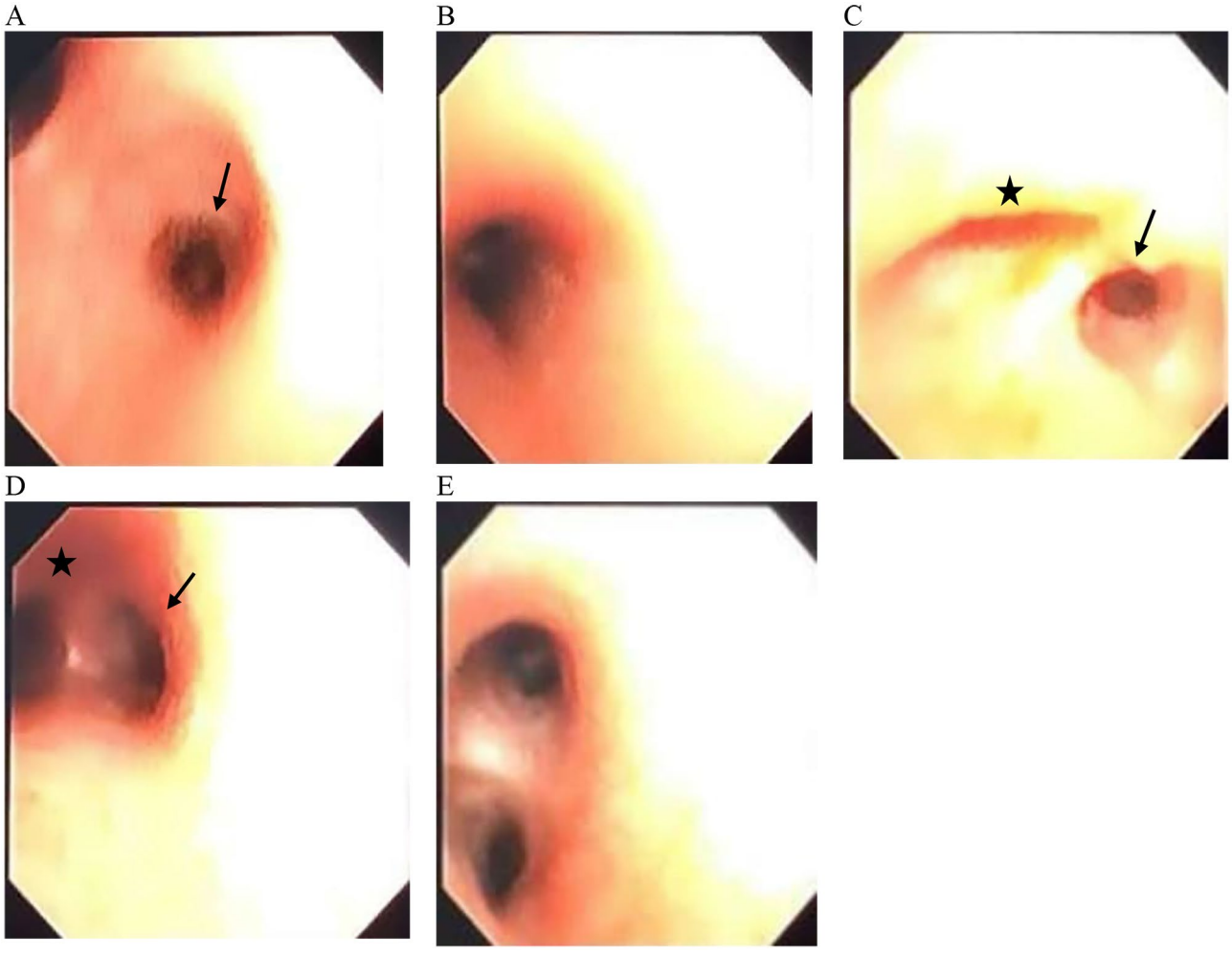

Fig. 3 Bronchoscopic findings in a patient with TB. **A**. An arrow indicates the opening of the TB in the right main bronchus. **B**. A close-up view of the TB. **C** and **D**. An arrow points to the TB opening and a star marks the trachea. **E**. The carina, where the trachea bifurcates into the left and right main bronchi

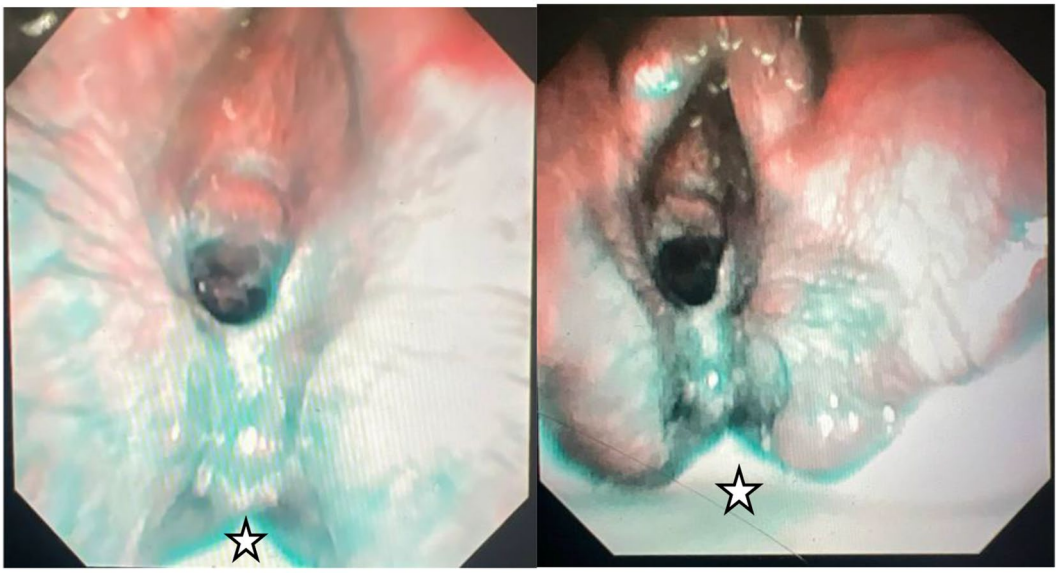

Fig. 4 Bronchoscopic findings in a patient with TB. a star marks the posterior laryngeal cleft (grade 1)

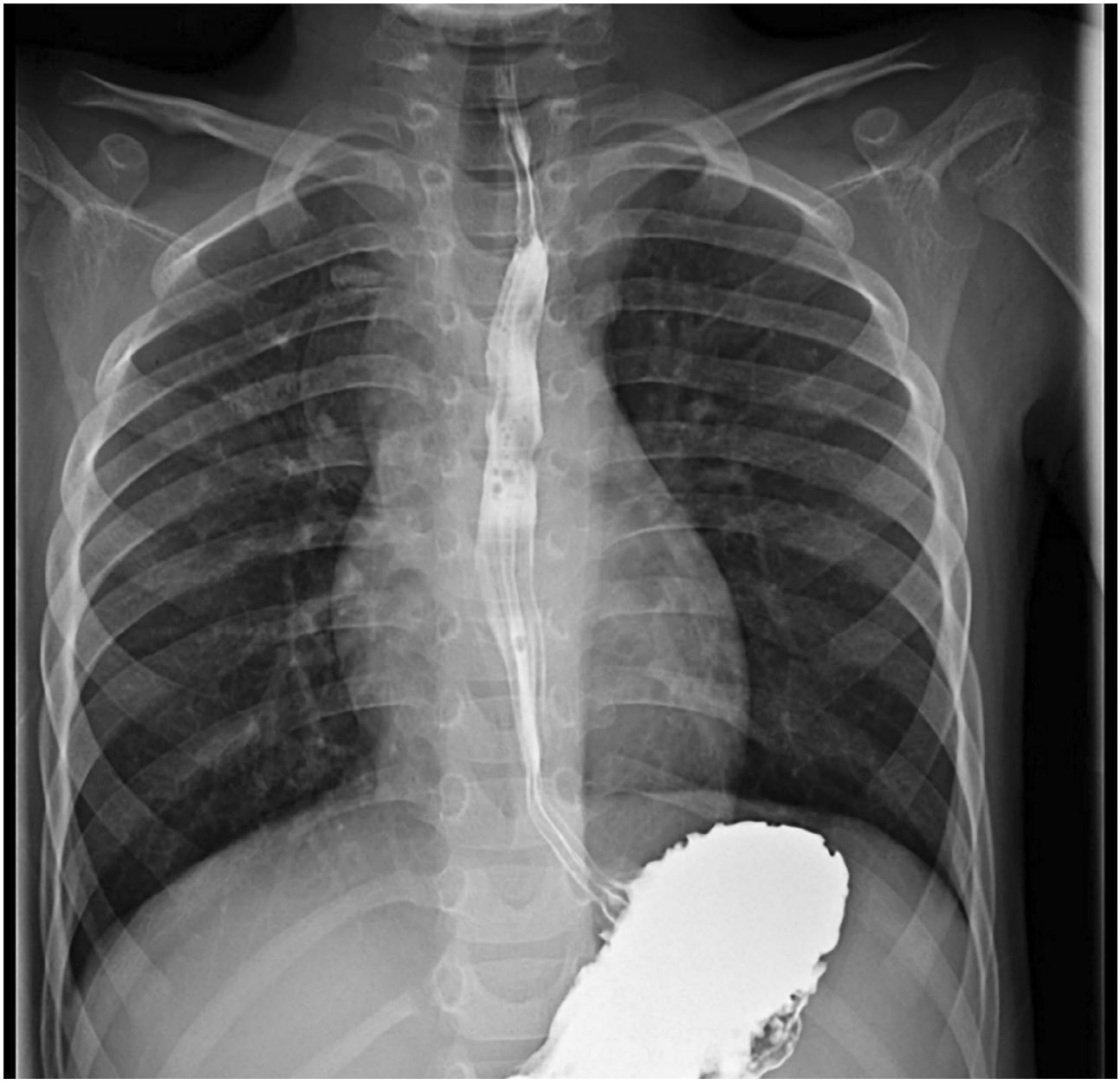

Fig. 5 Barium swallow demonstrates the integrity of the esophagus. No defect or leak is observed

Discussion

Congenital lung malformations (CLMs) are a group of rare and diverse disorders characterized by airway obstruction or narrowing, impacting pulmonary function. Typically, these anomalies are initially identified by signs indicative of increased respiratory effort, such as respiratory distress and retraction [12, 13]. The spectrum of morbidity and mortality associated with CLMs ranges from severe complications like fetal hydrops to milder forms presenting as asymptomatic lesions [14]. The gamut of CLMs includes various vascular, thoracic, and pulmonary anomalies, extending from the upper airway to the lung parenchyma, and encompasses conditions such as lobar pulmonary sequestration (PS), bronchial atresia, cysts, and emphysema [15]. Pediatric bronchoscopy has been instrumental in diagnosing a variety of tracheobronchial disorders, offering direct visualization and sampling. This technique is particularly useful for assessing patients with symptoms like stridor, persistent atelectasis, recurrent segmental pneumonia, or those who may remain asymptomatic for extended durations [16].

In the case presented, flexible bronchoscopy uncovered several anomalies, including a PLC, TB, and significant stenosis in the lower half of the trachea. TB is characterized by a range of bronchial anomalies within the main bronchus, extending into the upper lobe region

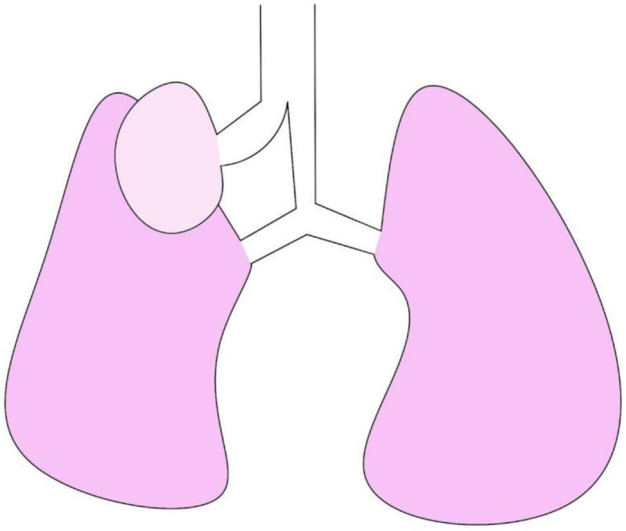

Fig. 6 The schematic representation depicted above illustrates the tracheal bronchus, an isolated pulmonary mass, and the distal tracheal stenosis

[13], and in this instance, was situated on the right side, aligning with literature that suggests a higher prevalence of right-sided TB [17]. Bilateral TB has been associated with narrowing of the distal trachea, as noted in prior studies [18]. The PLC observed was classified as grade 1, following the criteria set forth by Benjamin B et al. [19], which describes a supraglottic cleft extending above the posterior cricoid cartilage. Notably, our case exhibited a unique pulmonary parenchyma within the right TB, diverging from previous reports, and was remarkable for presenting multiple pulmonary anomalies concurrently, identified during diagnostic evaluations.

The pharmacological treatment administered to the child in this case was meticulously evaluated for its efficacy and appropriateness. Given the absence of extrapulmonary diseases and cardiac anomalies, the conventional pharmacotherapy aimed at alleviating respiratory symptoms was deemed accurate and effective. The treatment's success underscores the importance of individualized therapeutic strategies, particularly in cases involving complex congenital anomalies. The precision in managing the child's condition reflects a deep understanding of the interplay between anatomical peculiarities and clinical manifestations, ensuring a favorable prognosis without the need for invasive procedures.

Children with congenital heart disease exhibit a significantly increased susceptibility to TB, with a thirteenfold higher risk [20]. Considering the potential association between TB and cardiac anomalies, the patient was referred for a comprehensive cardiac assessment. The subsequent cardiac examination and echocardiography revealed no abnormalities. Other conditions often correlated with TB include Down's syndrome and various chest and lung abnormalities [21]. Our patient, fortunately, did not present with any extrapulmonary diseases, which favorably influenced the prognosis and negated the necessity for interventions beyond pharmacological management of the condition and respiratory symptoms. However, the absence of follow-up data is a limitation of the current report. We aim to address this in future updates, which will provide insights into the patient's progress and the long-term efficacy of the chosen treatment strategy. This will help to further elucidate the decision-making process in similar clinical scenarios.

Conclusion

In conclusion, our case report highlights the coexistence of multiple rare pulmonary anomalies with chronic respiratory symptoms, illustrating the complexity of such medical presentations. The patient's treatment in the hospital involved conventional pharmacotherapy, which successfully alleviated the respiratory distress. The tracheal bronchus, in particular, demonstrates a wide range of clinical manifestations that are influenced by the specific anatomical malformation and the patient's overall health, especially the presence of immunodeficiency states. These can vary from recurrent respiratory tract infections necessitating interventional procedures to completely asymptomatic cases that do not require surgical intervention. Therefore, clinicians must maintain a high index of suspicion for rare pulmonary anomalies in patients presenting with persistent respiratory symptoms. Such vigilance can facilitate timely diagnosis and significantly enhance patient outcomes by tailoring interventions to the unique aspects of each case.

Abbreviations

CTMs	Congenital Tracheobronchial Malformations
TB	Tracheal Bronchus
PLCs	Posterior Laryngeal Cleft
CLMs	Congenital Lung Malformations
PS	Pulmonary Sequestration

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Not applicable.

Author contributions

A.M. conceived of the presented study, A.M. and A.A. wrote the manuscript in consultation with R.N.K and SH.M. reviewed. A.M. and F.B.N. edited and revised the manuscript. All authors discussed the results and contributed to the final manuscript, have read and approved it, and ensure that this is the case.

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Data availability

The datasets used during the current study available from the corresponding author on reasonable request.

Declarations

Ethical approval
The study was reviewed and approved by the Research Ethics Committees of Kurdistan University of Medical Sciences Medicine. The project was found to be by the ethical principles and the national norms and standards for conducting Medical Research in Iran. Written informed consent to participate in this study was provided by the participant's legal guardian.

Consent for publication
Informed consent was obtained from the patient's parents for the publication of this case report.

Competing interests
The authors declare no competing interests.

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References

1. Ghaye B, Szapiro D, Fanchamps J-M, Dondelinger RF. Congenital bronchial abnormalities revisited. Radiographics. 2001;21(1):105–19.
2. Mohammadi S, Sharifi R, Motavasel E, Shariati MBH. Tracheal bronchus: Case Report. Acta Medica Iranica; 2023.
3. Thilagavathy G, Srinivasan R, Aruna S, Bonny J, Siddhuraj C. A rare case of a tracheal bronchus. J Clin Diagn Res. 2012;6(6):1056–8.
4. Eddine HJ, Arab WA. Tracheal bronchus: Case Report and Literature Review. SN Compr Clin Med. 2021;3(1):399–403.
5. Richter C. Dissertatio médico de infanticide inartis obstetriciae: Thesis for Doctor of Medicine. Leipzig; 1792.
6. Blumberg JB, Stevenson JK, Lemire RJ, Boyden EA. Laryngotracheoesophageal cleft, the embryologic implications: review of the literature. Surgery. 1965;57(4):559–66.
7. Schweigert M, Dubecz A, Ofner D, Stein HJ. Tracheal bronchus associated with recurrent pneumonia. Ulster Med J. 2013;82(2):94.
8. Barat M, Konrad HR. Tracheal bronchus. Am J Otolaryngol. 1987;8(2):118–22.
9. Reis JC, Martins CR, Otta LH, Tardelli MA, Amaral JLGd. Complication related to tracheal bronchus in infant: case report. Revista brasileira de anestesiologia. 2006;56:72–7.
10. Zheng Y-J. Diagnosis of tracheal bronchus in children Yue-Jie Zheng, Ji-Kui Deng, Dao-Zhen Zhang, Yun-Geng Gen. World J Pediatr. 2007;3(4):286–9.
11. Panigada S, Sacco O, Girosi D, Tomà P, Rossi GA. Recurrent severe lower respiratory tract infections in a child with abnormal tracheal morphology. Pediatr Pulmonol. 2009;44(2):192–4.
12. Andrade CF, Ferreira HPC, Fischer GB. Congenital lung malformations. Jornal Brasileiro De Pneumologia. 2011;37:259–71.
13. Sandu K, Monnier P. Congenital tracheal anomalies. Otolaryngol Clin North Am. 2007;40(1):193–217.
14. Palla J, Sockrider MM. Congenital lung malformations. Pediatr Ann. 2019;48(4):e169–74.
15. Bush A, Chitty L, Harcourt J, Hewitt RJ, Nicholson AG. 18 - congenital lung disease. In: Wilmott RW, Deterding R, Li A, Ratjen F, Sly P, Zar HJ, et al. editors. Kendig's disorders of the respiratory tract in children (Ninth Edition). Philadelphia: Elsevier; 2019. pp. 289–e3378.
16. Remy J, Remy-Jardin M, Artaud D, Fribourg M. Multiplanar and three-dimensional reconstruction techniques in CT: impact on chest diseases. Eur Radiol. 1998;8:335–51.
17. Lawrence DA, Branson B, Oliva I, Rubinowitz A. The wonderful world of the Windpipe: a review of Central Airway anatomy and Pathology. Can Assoc Radiol J. 2015;66(1):30–43.
18. Cope R, Campbell JR, Wall M. Bilateral tracheal bronchi. J Pediatr Surg. 1986;21(5):443–4.
19. Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. Annals Otology Rhinology Laryngology. 1989;98(6):417–20.
20. Chassagnon G, Morel B, Carpentier E, Le Ducou H, Sirinelli D. Tracheobronchial branching abnormalities: lobe-based classification scheme. Radiographics. 2016;36(2):358–73.
21. McLaughlin F, Strieder D, Harris G, Vawter G, Eraklis A. Tracheal bronchus: association with respiratory morbidity in childhood. J Pediatr. 1985;106(5):751–5.

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