Case Report

An 8-year-old boy with diffuse cystic pulmonary tuberculosis: An intriguing story

Wonashi R Tsanglao, Sulanthung Kikon, Tenukala Aier

From Consultant, Department of Pediatrics, Christian Institute of Health Sciences and Research Hospital, Dimapur, Nagaland, India

ABSTRACT

Diffuse cystic lung disease due to pulmonary tuberculosis (TB) is rare. We describe an 8-year-old boy who presented with acute onset respiratory distress while on a compliant anti-tubercular treatment for a recently diagnosed pulmonary TB. On clinical examination, hypoxemia, clubbing, and features of the right-sided heart failure were observed. High-resolution computed tomography of the chest revealed extensive cystic lung parenchymal changes with ground glassing and consolidation, and echocardiography indicated the presence of pulmonary arterial hypertension. His treatment included supplemental oxygen, sildenafil, prednisolone, and anti-tubercular drugs. At the two-year follow-up, the patient showed complete clinical recovery and resolution of cysts on the chest X-ray.

Key words: Cystic lung disease, Diffuse lung disease, Pulmonary hypertension, Pulmonary tuberculosis

The National Tuberculosis Elimination Program notified 1.8 million cases of tuberculosis (TB) in India in 2020, out of which 5.65% belong to the age group 0–14 years [1]. Children most often present with pulmonary TB, which manifests as airspace consolidation, hilar lymphadenopathy, nodular opacities, tree-in-bud appearance, cavitation, fibrosis, and pleural effusion on imaging [2].

Cystic lung disease (CLD) is an uncommon presentation of pulmonary TB [2], of which little is known about the pathogenesis, management, and long-term outcome. We describe an 8-year-old boy with diffuse cystic pulmonary TB and pulmonary hypertension, who showed complete clinical and radiological recovery at a 2-year follow-up. Through this case report, we highlight an unusual presentation of a common disease and the need for long-term follow-up in such cases.

CASE REPORT

An 8-year-old boy was brought to the emergency department of our institute with complaints of cough and dyspnea for 3 days. The cough was non-paroxysmal, productive, and without diurnal variation, while the dyspnea was severe enough to render him unable to walk around within the house (New York Heart Association class 3) [3]. There was no history of fever, noisy breathing, wheeze, swelling of limbs, or orthopnea. 5 months earlier, he was diagnosed with pulmonary TB at a district hospital, which was based on findings of acid–fast bacilli (AFB) 2+ on sputum microscopy, and bilateral infiltrates with cystic changes on chest X-ray (Fig. 1a). He was started on four drugs of anti-tubercular treatment, and the parents confirmed that he had been compliant with the treatment. Before being diagnosed with TB, he was healthy with a normal birth history and development. He was fully immunized and his family history was unremarkable.

At admission, he was alert, with a heart rate of 124/min, capillary refill time of less than 3 seconds, respiratory rate of 46/min with subcostal recessions, blood pressure of 100/70 mm Hg, and oxygen saturation (SPO_2) of 80% in room air that improved to 99% with supplemental oxygen at 8 Litres/minute with a non-rebreather mask. General examination revealed the presence of grade 3 pan-digital clubbing. His weight was 15 kilograms (<3rd centile for age), height 115 cm (3rd to 10th centile), and the body mass index was 11.34 kg/m^2 (< 3rd percentile) indicating a severely underweight child. On respiratory system examination, the chest movement was symmetrical bilaterally, with no deformity; palpatory and percussion findings were normal. Auscultation revealed the presence of bilateral coarse crackles, with normal vocal resonance. Cardiovascular examination revealed loud P2 and raised jugular venous pressure (5 cm above the sternal angle). Abdomen and central nervous system examinations were normal.

On investigation, arterial blood gas showed a pH of 7.36 with PO2 of 58 mmHg, PCO2 of 40 mmHg, bicarbonate of 20 mmol/L, and lactate of 2.0 mmol/L. He had an elevated total white cell count (14,450 cells per mm^3) with 89% neutrophils and

Access this article online

Received - 31 January 2023
Initial Review - 18 February 2023
Accepted - 07 March 2023

Quick Response code

DOI: 10.32677/ijcr.v9i2.3851

Correspondence to: Dr. Wonashi R Tsanglao, Department of Pediatrics, Christian Institute of Health Sciences and Research, Dimapur - 797 115, Nagaland, India. E-mail: wonash_tsanglao@yahoo.co.in

6% lymphocytes. The erythrocyte sedimentation rate was 40 mm in the 1st h. Chest X-ray showed persistent bilateral radiological changes in the form of infiltrates and cystic changes.

He was empirically started on intravenous amoxicillin-clavulanate (30 mg/kg/dose 8 hourly), and anti-tubercular drugs (Isoniazid 10 mg/kg, rifampicin 10 mg/kg, and ethambutol 20 mg/kg) were continued. Gastric aspirate for AFB and geneXpert were negative. Sputum culture for bacteria and enzyme-linked immune assay test for human immunodeficiency virus were negative. High-resolution computed tomography of the chest showed cystic destruction of lung parenchyma with surrounding collapse consolidations in the right upper lobe and apicoposterior segment of the left upper lobe. There were ground-glass consolidations diffusely distributed in the remaining lungs bilaterally with scattered areas of nodular opacities, cystic changes, and small cavitations (Fig. 2). 2D-echocardiography showed dilated right atrium and right ventricle with moderate tricuspid regurgitation (gradient 39 mmHg) suggestive of pulmonary arterial hypertension (PAH). Prednisolone (1 mg/kg/day), sildenafil (10 mg 8 hourly), and furosemide (1mg/kg/day) were added to his therapy. A noticeable improvement in his clinical status was observed, and by day 14 of the hospitalization, his oxygen requirement dropped to 2L/min on the nasal cannula. On the 20th day, he was discharged at request on home oxygen. At 1-month follow-up, anti-tubercular drugs were stopped on completion of 6 months. Prednisolone was tapered and stopped subsequently.

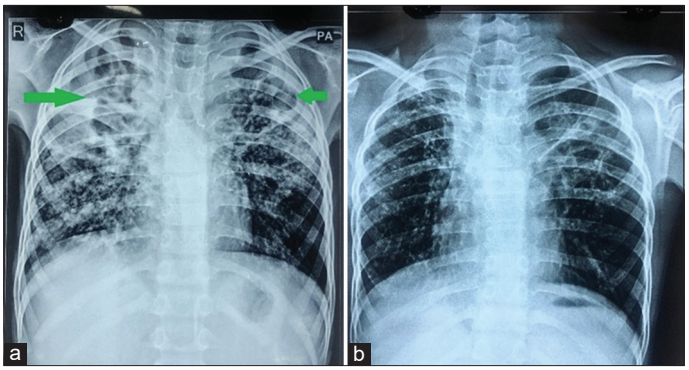

Figure 1: (a) Initial chest X-ray (5 months earlier), showing infiltrates and cystic changes in bilateral upper zones (green arrows) and (b) chest radiograph at 2-year follow-up showing bilateral fibrocalcific opacities and resolution of cystic changes

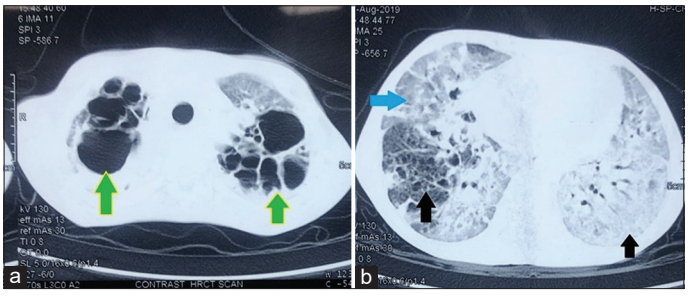

Figure 2: High-resolution computed tomography of the chest at present hospitalization showing (a) multiple cystic changes in the bilateral upper lobes (green arrows) and (b) ground glass opacities (blue arrow), with areas of cystic changes and consolidation (black arrows)

He was physically followed up in the outpatient department for the next 4 months and telephonically thereafter in view of the COVID-19 pandemic. Over the next 2 months, oxygen, sildenafil, and furosemide were reportedly discontinued by the parents after noticing tolerance in physical activity and improvement in SPO_2. The patient apparently remained well thereafter. At the two-year physical follow-up, he was asymptomatic and attending school, digital clubbing was absent, and the chest findings were normal. His growth chart also showed an upward trajectory (weight 25 Kg, height 129 cm). Chest X-ray (Fig. 1b) showed a complete clearing of cysts, and echocardiography revealed a structurally normal heart with no evidence of pulmonary hypertension.

DISCUSSION

CLD is characterized by the presence of multiple cysts, defined as thin-walled (<2 mm) round parenchymal lucencies interfaced with normal lung [4]. Differential diagnoses include lymphangioleiomyomatosis, pulmonary Langerhans cell histiocytosis, lymphocytic interstitial pneumonia, amyloidosis, Britt-Hogg-Dube syndrome, centrilobular emphysema, congenital bronchiectasis, cystic adenomatoid malformation of the lung, tracheal papillomatosis, and pulmonary infections (pneumatoceles), including *Pneumocystis jiroveci* pneumonia in immunocompromised hosts [2,4,5].

There have been only a few cases of pulmonary TB manifesting as diffuse CLD reported in both adults and children [2,4-10]. To the best of our knowledge, five cases have been reported in children (Table 1). The pathogenesis of cyst formation in pulmonary TB is unclear. A check-valve obstruction due to granulomatous inflammation and luminal narrowing of the bronchioles has been described by most authors to be the likely mechanism of cyst formation [2,5]. Cysts can also be caused by peribronchiolar fibrosis, or by drainage of necrotic material by a communicating bronchus [2]. In pulmonary TB, the cystic changes are reported to be predominant in the upper lung zones as seen in our case, often surrounded by consolidation; centrilobular; and cavitating nodules [2,5].

There are no established guidelines for treating cystic pulmonary TB. Most of the treatment has been directed at treating the underlying TB. Some authors have reported the use of steroids in a few cases [5,6,9,10]. The clinical course and prognosis of diffuse cystic pulmonary TB are not well known. Most case reports of cystic pulmonary TB have described diffuse air space opacities and/or micronodular opacities as the initial radiologic finding that later evolved into cystic changes [2,6,7]. Complete or partial resolution of cysts following anti-tuberculous treatment, rapid progression to respiratory failure and death, and persistence of cyst after treatment has been reported by previous authors [5,6,7]. Our patient showed complete clinical and radiological recovery at the 2-year follow-up, even though he had extensive lung involvement at presentation. In addition, pulmonary hypertension was an unusual finding in our case. The only pediatric case report we are aware of in children describes an 8-year-old boy with smear-positive pulmonary TB and pulmonary hypertension [11]. Hypoxia, restriction of the pulmonary vascular bed, and decreased functional lung parenchyma are described

Table 1: Cases of cystic pulmonary tuberculosis reported in children

Author	Study design	Findings	Histology	Treatment	Outcome
Ray *et al*. [5]	Case report	13 - year-old female with fever for 1 month and breathlessness for 10 days. CT chest: Multiple thin walled cysts seen in both the lungs (left>right; upper lobe>lower lobe); diffuse bilateral ground glass opacities and centrilobular nodules	Granulomatous inflammation AFB positive	4-drug anti-tubercular treatment (ATT) and Steroid.	End of treatment CT chest (6 months): Reduction in the number and size of cysts with decrease in diffuse lung opacities and nodular lesions.
Periwal *et al*. [6]	Case report	14-year-old female with fever for 3 months, breathlessness and weight loss for 1 month. CT chest: Bilateral pneumothorax, multiple cystic changes , and ground glass haziness	Extensive necrosis and cystic changes. Bronchocentric granulomatosis. AFB positive	4-drug ATT and steroid Mechanical ventilation.	Died due to respiratory failure
Jana *et al*. [8]	Picture of the month	10-month-old girl, known case of Rifampicin resistant pulmonary TB on ATT for 2 months, presented with breathlessness for 5 days. Chest X-ray: Extensive bilateral consolidation that progressed to cystic changes.	Not done	Second line ATT	Repeated episodes of respiratory distress after 3 months.
Tsanglao *et al*. [9]	Case report	8-month-old boy, recently diagnosed rifampicin sensitive miliary TB, on ATT (6 weeks) presented with fast breathing for 2 days. HIV negative. CT Chest: multiple cysts in bilateral upper lobes, left lower lobe; bilateral extensive centrilobular nodules, consolidation, ground glass opacities.	Lymphocytic interstitial pneumonia. AFB negative	4-drug ATT and Steroid (prednisolone) for 16 weeks.	Asymptomatic at 15-month follow up. Chest X-ray : resolution of cysts.
Mohari *et al*. [10]	Case report	2-month-old male with fever and tachypnea for 6 weeks. CT chest: progression from diffuse nodular infiltrates to multifocal cystic changes.	Not done AFB positive	4-drug ATT Steroid Mechanical ventilation (High-frequency oscillatory ventilation)	Asymptomatic at 6-month follow up

as the possible reasons for the development of pulmonary hypertension and cor pulmonale [11].

CONCLUSION

Although rare, pulmonary TB should be considered as a differential diagnosis in diffuse CLD. There is a need to identify and follow-up on more cases of this rare form of a relatively common disease to understand its clinical course, management, and long-term outcome. It is imperative to look for the presence of pulmonary hypertension in pulmonary TB with diffuse lung parenchymal changes.

REFERENCES

1. Central TB Division, Ministry of Health and Family Welfare. India TB Report 2021. New Delhi: JK Offset Pvt. Ltd; 2021. p. 352.
2. Perim J, Pimenta ES, Marchiori E. Cystic tuberculosis: A very unusual aspect of a common disease. Pulmonology 2020;26:400-3.
3. The Criteria Committee of the New York Heart Association. Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels. 9th ed. Boston: Little, Brown and Co; 1994. p. 253-6.
4. Park S, Lee EJ. Diagnosis and treatment of cystic lung disease. Korean J Intern Med 2017;32:229-38.
5. Ray A, Suri JC, Sen MK, Khanna A. Cystic lung disease in tuberculosis: An unusual presentation. Lung India 2013;30:351-3.
6. Periwal P, Khanna A, Gothi R, Talwar D. Bronchocentric granulomatosis with extensive cystic lung disease in tuberculosis: An unusual presentation. Lung India 2016;33:320-2.
7. Lee HJ, Goo JM, Im JG. Rapid and irreversible cystic change of pulmonary tuberculosis in an immunocompetent adult. J Thorac Imaging 2003;18:254-6.
8. Jana M, Bansal S, Bhalla AS. Lodha R, Pritviraj SK. Cystic lung disease in tuberculosis. Indian J Pediatr 2021;88:734-35.
9. Tsanglao WR, Miachieo N, Ao MK, Aier T, Kikon S. Lymphocytic interstitial pneumonia in an infant with diffuse cystic pulmonary tuberculosis. Indian Pediatr Case Rep 2022;2:21-4.
10. Mohari N, Raj D, Lodha R, Kabra SK. Prolonged high-frequency oscillatory ventilation in tubercular multifocal cystic lung disease. Respir Care 2012;57:2111-4.
11. Sharda N. Cor pulmonale as a rare presentation of pulmonary tuberculosis in a child. Pediatr Infect Dis J 2013;32:424.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Tsanglao WR, Kikon S, Aier T. An 8-year-old boy with diffuse cystic pulmonary tuberculosis: An intriguing story. Indian J Case Reports. 2023; 9(2):57-59.