

Pulmonary Hypoplasia: A Rare Cause of Chronic Cough in TB Endemic Area

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Abstract

Pulmonary hypoplasia is a rare disease characterized by a defect of lung development more often unilateral. The diagnosis requires several exams to eliminate other causes of pulmonary retraction. We report two cases at the department of pneumophtisiology of the University Teaching Hospital of Point G. The first case is a young adult who was complaining of a chronic cough. Etiological investigation required several exams including spirometry and Computed tomographic scan (CT scan). After elimination of all suspected causes of pulmonary opacity, the diagnosis of pulmonary hypoplasia was retained. The second case is a 2-year-old girl who was born with congenital cardiopathy whose respiratory complications were increasing during her childhood and respiratory explorations discovered pulmonary agenesis. Pulmonary hypoplasia is rare in our medical practice, but attention must be drawn to a retractile pulmonary opacity in young age after elimination of all infectious causes in TB endemic area.

Keywords

A Chronic Cough, Pulmonary Hypoplasia, Bamako

1. Introduction

Pulmonary hypoplasia (PH) is a rare congenital malformation without apparent cause associated with other developmental abnormalities [1]. It is defined as a defect in the lungs characterized by bronchial segmentation often conserved in a

decrease in pulmonary alveoli. It may be unilateral or bilateral, segmental or lobar [2]. The Pulmonary hypoplasia occurs with an alteration of the molecular mechanisms of the early diaphragm and lung development, aggravated by later fetal dynamic stress [3]. The majority of children with pulmonary hypoplasia will have some degree of pulmonary artery hypertension resulting from increased muscularization of the intra acinar pulmonary arteries [4]. With an incidence estimated to be 1 per 10,000 births, 24% of malformations are discovered at birth or adolescence, resulting in significant morbidity and mortality [5]. Mortality is particularly due to congenital cardiac anomalies, but also esophageal, vertebral and renal [2]. A hypoplastic lung is more likely to be atelectasis, fibrotic, functionless, and prone to infection whatever is the etiology. The typical presentation is neonatal respiratory distress syndrome, whereas the diagnosis in adulthood is usually fortuitous or secondary to respiratory symptoms [6] [7]. Cough is the most common respiratory symptom in hypoplasia [7] [8] [9] [10]. It is qualified to be chronic and suspecting tuberculosis (TB) if it exceeds fifteen (15) days according to the national TB program (PNLT) guidelines in Mali. TB incidence in Mali is 57 per 100,000 populations; TB screening strategy is based on cough duration. A chronic cough is a major criterion for TB test in sputum in Mali and all country where TB is endemic [11]. We report two cases of pulmonary hypoplasia fortuitously discovered at the Department of pneumophthisiology of the University Teaching Hospital of Point G in Bamako, Mali. These cases are the first to be published in Mali; their diagnosis was derived from collaborative teamwork between the clinicians and radiologists.

2. Observation

Observation 1: A 24 years old, male, farmer, and tailor with no alcohol and smoking history. He was received at pneumology consultation for a chronic cough with mucopurulent expectoration sometimes fetid. He was presenting Sadoul stage-2 dyspnea for seven (7) years associated with chest pain, heartburn, mild to moderate persistent rhinitis and recurrent cough worsened since 1 month. Physical examination found a low-grade fever of 38°C, a static performance (PS) of 1, a body mass index (BMI) of 20 kg/m², an absence Digital clubbing, the respiratory frequency was 23 cycles/minutes at rest, and oxygen saturation (SpO₂) was 97% at ambient air. Cardiac auscultation was normal. At Chest inspection noted a diminution of the right chest expansion. Pulmonary auscultation found a decreased vesicular murmur at the right side, cracking rales at bottom of the lung and bilateral rhonchi and wheezes located at the third section of the right lung.

Chest X-ray showed a dense opacity of the right middle lobe, trachea and mediastinal displacement to the right, inter-costal spaces pinching on the right lung and distention of the left lung (**Figure 1(a)**). The blood cell count was normal, the serology of the acquired immunodeficiency virus (HIV) was negative and tuberculin skin test (TST) was positive (12 mm). Smear Acid Fast Bacilli test was negative with Ziehl Nielsen coloration and absence of *Mycobacterium tuberculosis*

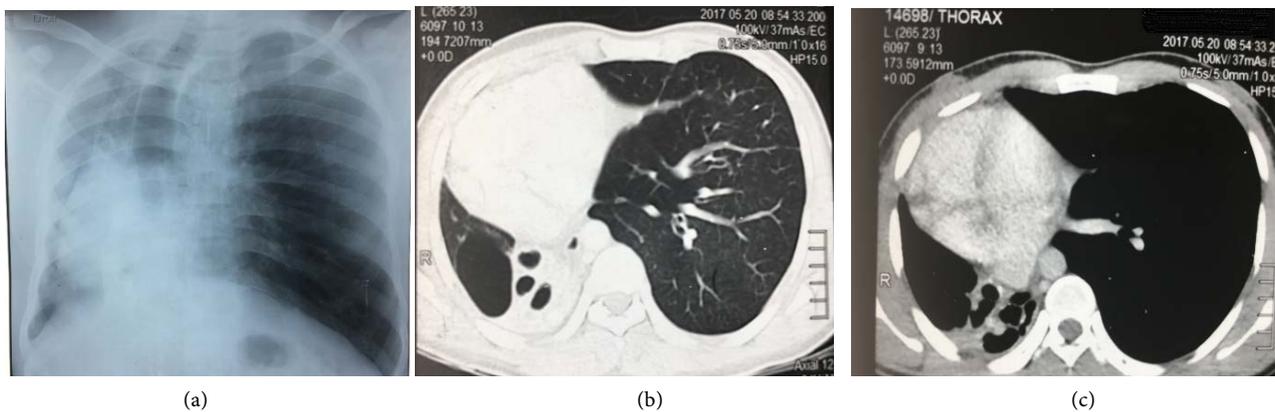


Figure 1. (a) chest X ray → Left thoracic distension and right mediastinal hernia, (b) and (c) computed tomographic scan mediastinal hernia, emphysema bubble, and middle lobe bronchial dilations.

with GeneXpert MTB/RIF[®], the cytology and bacteriological exam of sputum (CBES) did not isolate any germ. Abdominal-pelvic ultrasound, digestive fibroscopy, and sinus radiography were normal. The chest CT scan (**Figure 1(b)** and **Figure 1(c)**) showed a distended and ventilated upper right lobe with bronchiectasis (BD) and emphysema bubbles. A complete collapse of the middle and lower right lobes was observed. Bronchial dilatation interesting the intermediary trunk and the lower lobe associated with the ascension of the diaphragm and liver. The left lung was much distended with posterior and posterior mediastinal lung hernia.

EKG and echocardiography showed a sinus rhythm with right axial deviation, normal cardiac cavities, left ventricle ejection fraction at 65% and minimal leakage tricuspid.

The patient was initially treated as a secondary bacterial infection due to the lack of arguments for tuberculosis. Spirometry showed a mixed ventilation disorder with a forced vital capacity (FVC) at 2.81 (63%), forced expiratory volume to second (FEV₂) at 1.49 (39%), FEV₂/FVC = 53 (64%), the reversibility test was significant with post FEV₂ at 1.71% for 15% of variation. The prick test for inhaled allergens was normal and the six minutes' walk test recorded a distance of 368 m (59%). We retained the diagnosis of right pulmonary hypoplasia with bronchial dilatation and homolateral lung emphysema. He was treated with inhaled based Salmeterol 250 mcg and fluticazone 25 µg twice a day, vaccination against pneumococcal, and chest physiotherapy for bronchial drainage. The 6 months evolution was satisfactory symptoms regression with persistence of a low-grade cough. Regular follow-up every six months has been prescribed to the patient.

Observation 2: A child aged two (2) years and 3 months, female, the first-born of the family was diagnosed with congenital heart malformation type common arterial trunk (CAT) at the age of three months of life. The medical history did not find inbreeding but allergic maternal rhinitis. The initial symptomatology was dominated by recurrent respiratory infections with at least two

episodes of wheezing bronchiolitis and pulmonary arterial hypertension (PAH) was suspected of cardiac ultrasounds. She was prescribed Sildenafil 12.5 mg per day. She was received at pneumology for recurrent rhino-bronchitis. The physical examination showed a slight developmental delay (90 cm height and 9.8 kg weight), increased respiratory rate, and pulse at 178 and SpO₂ of 78% at ambient air. No digital hyppocratism, normal conjunctive coloration, no sign of dehydration and no visible skin lesions, and presence of bilateral corneal hypertrophy with inflammatory mucosa. The pulmonary examination showed right thoracic distension with diminution on the left chest expansion. Lung auscultation perceived abolition vesicular murmur and Ronchi on the left lung, cardiac tip shock was visible in the left side. The cardiac exam found a tachycardia at 130 beats per minute, peripheral pulses were perceived and synchronous, there was no collateral venous circulation or cyanosis. The patient was prescribed nasal hygiene, antibiotics and antihistamine, and azithromycin. Control at Day 7 showed basal oxygen saturation with 88% SpO₂. Biological explorations such as blood cell count, HIV, hepatitis B, and C were normal. The prick test with respiratory allergens (DP, DF Blomia, ALT, ASP, 5G, cat, dog) was negative. Chest X-ray (**Figure 2(a)**) showed a retractile opacity on the half of the left lung associated with compensatory distension on the right lung. The abdominal ultrasound did not indicate abnormality of the abdominopelvic organs. Cardiac ultrasound found the small size of the left ventricle, dilatation right cavities, walls not hypertrophic and overall kinetics conserved. The pulmonary artery trunk was not visible and the two branches arise from the aorta and presence of PAH. Thoracic CT scan (**Figure 2(b)** and **Figure 2(c)**) showed a total collapse of the left lung with short and obstructed bronchus. Pulmonary vessels are present but reduced caliber, total mediastinal deviation to the left with reduction of the left chest; right lung expansion with a trans-mediastinal parenchymal hernia. Chest

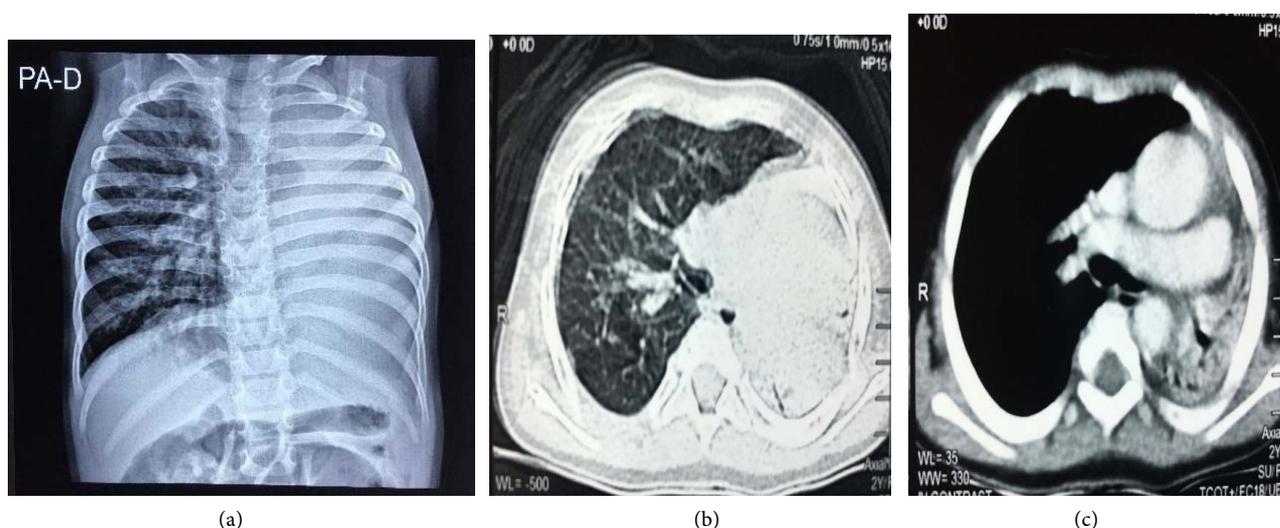


Figure 2. (a) Chest X-ray: Left retractile opacity; Chest scanner (b) right lung distention and mediastinal switch (c) single pulmonary artery and left parenchymal atelectasis.

radiography performed in December 2015 (at 3 months of birth) showed right diffuse alveolar opacities with parenchymal clarities, no retraction, mediastinal enlargement associated with cardiomegaly. A chest CT scan performed in March 2016 showed atelectasis by compression of the left lung, normal vascularization and no detectable lesions; frosted glass appearance of the right parenchyma. The diagnosis of left pulmonary hypoplasia with common arterial trunk and PAH was retained. A treatment based on nasal hygiene (mometasone furoate), pneumococcal vaccine and respiratory physiotherapy for bronchus drainage were prescribed. The 6 months clinical evolution was satisfactorily characterized by a regression of rhinitis and pulmonary symptoms. A regular follow-up every six months has been prescribed.

3. Discussion

Congenital pulmonary malformations are rare and complex. Most of them are discovered prenatally in developed countries or during the first month of life, but late discovery is also frequent depending on the severity of the symptoms [2]. The majority of malformations originate between 4 and 16 weeks of pregnancy, during the development of the aerial tree [12]. They are classified in 3 types according to Schneider: type 1 is pulmonary agenesis (complete absence of bronchus and parenchyma), type 2 is pulmonary aplasia (rudimentary blind bronchus with no parenchyma), and type 3 is pulmonary hypoplasia (incomplete bronchial development and varying degrees of parenchyma) [4]. Our patients respond to type 3, which would be secondary to the mechanical compression of the homolateral lung by intra-abdominal organs that remained in the thoracic position, the absence of regulation of the intra-pulmonary fluid and the oligoamnios are involved in the genesis of pulmonary hypoplasia [3] [12] [13].

It more often unilateral and can affect regardless of ages. Their frequency varies between 1/10,000 and 1/200,000 birth seems to be underestimated [14]. The clinical diagnosis remains difficult because there are no clinical symptoms or signs characteristic of pulmonary agenesis [6]. The circumstances of discovery of pulmonary agenesis reported in the literature are multiple, sometimes accidental during a medical checkup [15], investigation of a chronic cough or chest pain in a teenager or an adult [7] [8], investigation for respiratory distress or stunted growth during the first days of life, confirming the congenital aspect of the disease [16]. A discrete male predominance is reported authors [17] or equal frequency regardless of gender [7] as found in our series. The literature review found an inconsistent predominance of the left side in 70% which seems to be less associated with cardiac abnormalities [9] [14]. In our series, we reported both attainment left and right side. The cardiac anomaly was associated with left pulmonary hypoplasia in our second observation. Physical signs such as chest asymmetry, diminution of chest expansion, chronicity of symptoms and images chest X-ray in our context raise the discussion of lung destruction post-infection especially tuberculosis. The ventilatory compensation at the expense of the vascularization compensation would explain the occurrence of PAH. It was asso-

ciated with agenesis in 19% of cases and in 88% if there is a cardiac malformation [16] [18]. That could explain the precocity in one of our cases that has a TAC. The occurrence of primary pulmonary hypoplasia is rare and seems to be of low symptomatic character before puberty, as found in our series. The presence of comorbidities has a negative impact on prognosis [17] [19].

Suspicion is made on chest radiography and the confirmation requires several complementary exams to eliminate infectious etiology such as tuberculosis sequelae, trachea-bronchial obstacles. Thoracic T Scan is a key element for certainty diagnosis. It helps to visualize bronchial, parenchyma and vascular abnormalities [1] [7] [8] [17]. The pulmonary functional assessment, in particular, the spirometry was performed in one of our cases and revealed a mixed ventilatory disorder. A similar result has been reported by Abdelfatah [8]. Lung scintigraphy can estimate the functional contribution of the lung; it was not feasible in our context because of the lack of technical platform [15].

The treatment is essentially symptomatic in the absence of surgical indication as described in the literature [1]. The emphasis was placed on medical treatment associating physiotherapy for bronchial drainage, pneumococcal vaccination, nasal hygiene, and bronchodilators. The treatment has improved the quality of life of our patients by reducing the frequency of exacerbations and patients' tolerance to the effort.

4. Conclusion

Pulmonary hypoplasia remains a rare diagnosis in our medical practices. It is under-diagnosed or/and the delay would be explained to its scarcity and non-specificity of the symptoms. It must be investigated in any chronic respiratory infections associated with unilateral chest opacity or diminution of lung volume at chest X-ray. The realization of the scanner is necessary to differentiate tuberculosis sequelae in low-income countries endemic.

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Conflicts of Interest

The authors have no conflict of interest.

Ethical Aspects

Voluntary Consent from case#1 and parental consent in case#2 were obtained for the Case reports being published.

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