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Unilateral Pulmonary Hypoplasia in an Adult Nigerian: A Case Report and Literature Review

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Abstract
Background: *Pulmonary hypoplasia presenting for the first time in adulthood is rare and can pose a diagnostic challenge without a high index of suspicion and proper investigations.*
Materials and Method: *We present a 42 year old man with an initial diagnosis of lung fibrosis from pulmonary tuberculosis but who on further evaluation was found to have left pulmonary hypoplasia. The history and clinical examination findings and are presented and management discussed. The patient was treated with empirical antibiotics, bronchodilators and referred for surgical evaluation.*
Pulmonary hypoplasia should be suspected in adults with homogenous opacification of the hemithorax.
Keywords: *computed tomography, hypoplasia, pulmonary, unilateral.*

Introduction

The development anomalies of the lung between the 4th and 24th gestational weeks may cause functional damage usually discovered in newborns and infants, and it can be rarely present in adulthood. Practically, the earlier the anomaly is present, the branching of the tracheal-bronchial tree is reduced.[1] Pulmonary hypoplasia is a developmental abnormality of the lung characterized by a decrease in the number of alveoli, cells, and airways with resultant decrease in size and weight of the lungs.[2] Pathologically, the hypoplastic lung has reduced lung weight, fewer generations of airways, and hypoplasia of the corresponding pulmonary arteries.[1] Unilateral Primary pulmonary hypoplasia is rare in adulthood. It is usually present in the neonatal period or in early childhood and is characterized by a decreased number of bronchial segments and decreased/absent alveolar air space.[3] The majority of patients present with severe respiratory distress or repeated pulmonary infections and wheezing, whereas some may be completely asymptomatic.[3] We present the case of 42 year old man who was referred to the pulmonologist unit of Benue state university teaching hospital Maude with an initial diagnosis of pulmonary tuberculosis with lung fibrosis.

Case Report

Mr A.S, A 49 YEAR OLD man was referred to the pulmonologist unit a with diagnosis of lung collapse

due to tuberculosis. He had a two months history of cough with scanty sputum, left sided pleuritic chest pain, dyspnoea and low grade fever. He had background 3year history of exertional dyspnoea with worsened in the preceding one month without orthopnea, paroxysmal nocturnal dyspnoea or leg swelling. There was no weight loss, night sweats and no smoking history or occupational exposure to fumes and dust. There was no previous treatment for tuberculosis. His previous health status was good. Examination revealed a middle aged man who was not in any distress, afebrile, not pale, acyanosed, no finger or toe clubbing and pedal oedema. In the Respiratory system, the respiratory rate was 24cycles/minute, with left trachea deviation, reduced expansion and dull percussion note on the same side. The breath sounds were absent on the left side. Features of emphysema were noted on the right hemithorax. Other systemic examinations were essentially normal.

PA chest radiograph-showed mediasternal shift to the left, homogenous opacity o f the left hemithorax obscuring the cardiac and diaphragmatic margins.

Due to the absence of a history consistent with tuberculosis, further investigation of the patient was carried out. Contrast enhanced Chest CT showed the left lung was reduced in size and hyper dense with no aeration. The left main bronchus and pulmonary artery were reduced in size. The hemi diaphragm on the left was elevated and left scoliosis involving T3-T7 was noted. There was compensatory right sided emphysema with herniation to the left hemithorax. Spirometry revealed a restrictive pattern of lung disease. Echocardiography showed cardiac windows displaced to the 6th left intercostal space in the anterior axillary line with left ventricular hypertrophy. No congenital anomalies were found. The haemogram was normal. Based on the history, examination and investigation findings, a diagnosis of Primary hypoplasia of the left lung with compensatory R lung emphysema was made. He was given empirical antibiotics, counselled and advised on follow up visits.

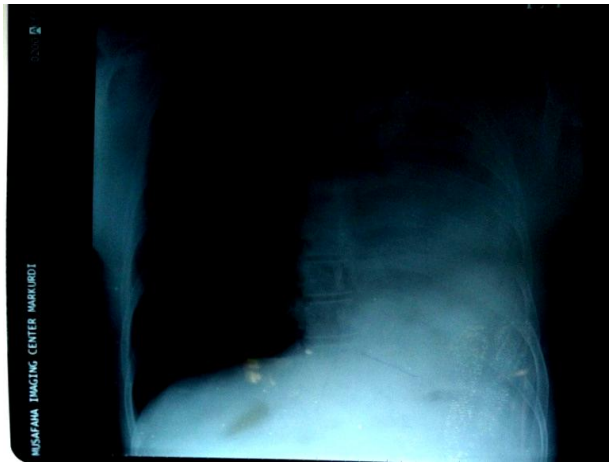

Figure 1: Chest radiograph showing complete opacification of the left hemithorax.

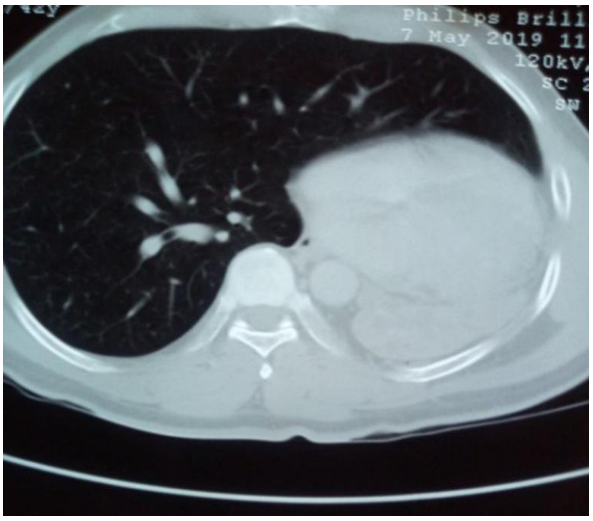

Figure 2: Axial CT chest demonstrating hypoplastic left lung with shift of mediastinal structures to the ipsilateral side.

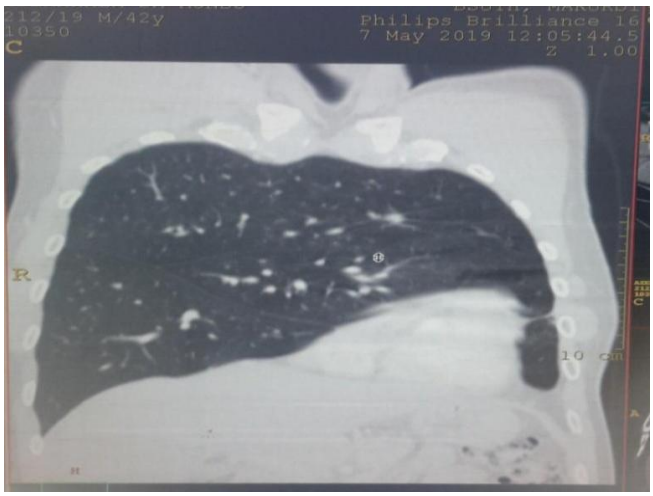

Figure 3: Coronary CT: Compensatory hypertrophy of the right lung with herniation into the left chest and mediastinal shift.

Figure 4: Coronal chest CT in left pulmonary hypoplasia. Note the hypoplastic left main bronchus.

Discussion

Unilateral pulmonary hypoplasia is a rare congenital anomaly. Although its exact prevalence is not well known, it is estimated to be seen in about 1 - 2 out of every 12,000 births.[4] The development of the lungs and developmental malformations too takes place in the 3rd and 4th week of intrauterine life. Primary pulmonary hypoplasia may be caused by deficient thyroid transcription factor 1, GATA factors, hepatocyte nuclear factor (HNF310), epidermal growth factor and its receptor; mitogen-activated protein kinase.[5]More commonly, it is secondary arising as a result of small fetal thoracic volume, prolonged oligohydramnios, early rupture of membranes at 15-28 weeks gestation, longer latent period before delivery, decreased fetal breathing, decreased pulmonary perfusion, congenital heart diseases, and trisomies.[5] Initially classified by Schneider and Schawatbe and later modified by Boyden depending upon the stage of development of the primitive bud, agenesis of the lung is further classified into 3 types.

Agenesis: (Type 1) is complete absence of the pulmonary artery and absence ipsilateral of pulmonary parenchyma and bronchus.

Aplasia: (Type 2) is rudimentary bronchus with complete absence of pulmonary parenchyma.

Hypoplasia: (Type 3) is presence of variable amounts of pulmonary parenchyma, bronchi, and supporting vessels.[6] Monaldi categorizes developmental disorders of the lung to four categories. Group I: No bifurcation of trachea; Group II: Only rudimentary main bronchus; Group III: Uncompleted development after bifurcation of the main bronchus; Group IV: Incomplete development of small segment and subsegmental bronchi of the corresponding lobe.[4] Our patient belongs to Monaldi group III, having an undeveloped left main bronchus and absence of pulmonary parenchyma.

Most cases present in neonates, infants, and childhood, but a few cases may remain asymptomatic until adolescence and adulthood and are discovered incidentally.[7] However, it may be infrequently present in adults not producing any symptoms thus rendering its diagnosis problematic.[6] Clinical course (severity of symptoms and their time of occurrence) is decided by the degree of hypoplasia present in the patient.[4,8] Our patient presented late in middle age with mild symptoms. This explained by the well developed, emphysematous.

Diagnosis may be established with help of chest x-ray, CT thorax, fiber optic bronchoscopy and if possible pulmonary angiography and bronchograph.[9] Contrast enhanced CT (CECT) is sufficient for the diagnosis of hypoplasia of the lung.[6] Plain radiographs demonstrate decreased aeration of the affected hemithorax and a small thoracic cage. A common finding is a shift of the mediastinum to the side of the hypoplasia, accentuated during inspiration due to increased compensatory ventilation of the other lung. A retrosternal soft tissue density can also be visible on lateral chest films representing heart and mediastinum that are displaced into the anterior chest.[3] CT scan may be required to establish the degree of under development and to differentiate hypoplasia from other conditions that may closely mimic it radiographically: atelectasis from other causes, severe bronchiectasis with collapse and advanced fibrothorax.

In our environment tuberculosis ranks high on the list of differential diagnosis. In our patient the

absence of any radiological features and negative gene expert negates the diagnosis. Total atelectasia, pleural effusion, pneumonia, dia-phragmatic elevation, diaphragmatic hernia, pulmonary aplasia, and pneumonectomy should be considered in differential diagnosis of these patients.[4] The closest diffential diagnosis is Swyer-James syndrome. In Swyer-James syndrome, the affected lung parenchyma appears hyperlucent, a result of air trapping owing to bronchiolar obstruction and blood vessels within the affected lung parenchyma are decreased in number and calibre.[4] It is excluded from the radiological findings of the CT scan of the patient as it is the left lung which is affected here but the normal right lung is hyperlucent (probably owing to compensatory emphysema) and blood vessels are also normal in the right lung as seen on contrast-enhanced CT scan.[4,10] In our patient,te findings were also a normal right lung (with compensatory emphysema) and blood vessels which excludes Swyer-James syndrome.

Adult patients with hypoplasia are treated normally with antibiotics for infections, bronchodilators and apophlegmatisant.[10] Our patient was treated with empirical antibiotics, a steroids and beta 2 agonist combination. He was also counselled and referred for surgical evaluation.

Recommendation

In clinical practice, a chest x-ray revealing complete or incomplete opacification of a hemithorax with volume loss should raise a strong the suspicion of the possibility of unilateral pulmonary hypoplasia.

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