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Case Report

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Case Report: Pulmonary Aplasia in Pregnant Women

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Abstract

Background: Pulmonary aplasia is a rare congenital pathology in which there is unilateral or bilateral absence of lung tissue. It differs from pulmonary agenesis, since in aplasia a closed, short-blind bronchus is present. The symptoms vary from no complaints to various respiratory complaints, however, non-specific clinical features act as a barrier to diagnosis.

Case Report: A 27-year-old woman, primigravide at week 12, presented with dry cough, breathing difficulty, fever 370C, slightly altered health conditions, conscious, oriented. She refers no concomitant diseases, except the frequency of "seasonal flu".

Physical examination/ Radiologic examination: Retraction of the left hemithorax, dullness in percussion and diminished vocal fremitus and breath sounds on the left hemithorax base.

Chest radiography: a homogenous density in the lower part of the left hemithorax with deviation of the trachea and mediastinum to the left.

Echo: left hemidiaphragm without mobility during respiration, left renal agenesis.

ECG: sinusal rythm, P pulmonale, RBBB.

Thoracic CT: Complete atelectasis of the left lung with short-blind left main bronchus and compensatory hyperinflation of the right lung. Unique right lung with total displacement of the mediastinum left as well as hypertrophy, herniation of the right lung tissue left.

It is diagnosed with left pulmonary aplasia, unique right kidney.

At 36 weeks of pregnancy, dyspnea is amplified and the patient undergoes cesarean delivery with spinal anesthesia. Both mother and infant did well.

Conclusion: Unilateral pulmonary aplasia without significant comorbidities may remain undiagnosed until adulthood. Pregnancy and childbirth are successful with multidisciplinary assistance.

Keywords: Pulmonary Aplasia; Pregnancy; Adults.

Introduction

Pulmonary aplasia is a rare congenital pathology in which there is unilateral or bilateral absence of lung tissue. Although similar, it differs from pulmonary agenesis, since in aplasia a closed, short-blind bronchus is present. Aplasia is usually unilateral, as bilateral pulmonary aplasia makes life impossible. It is often associated with other congenital anomalies. Pulmonary hypoplasia is incomplete development of the lungs in an abnormally low number or size of bronchopulmonary segments or alveoli [1-3]. The symptoms appear from no complaints to various respiratory complaints, however, non-specific clinical features act as a barrier to diagnosis [4]. It is detectable before birth, childhood [5], but the literature describes cases discovered in adulthood, even without symptoms [6,7,8], as well as undiagnosed cases discovered during pregnancy [9,10]. Unlike pulmonary hypoplasia which in most cases results from incomplete lung development during prenatal development, pulmonary agenesis and aplasia result from complete developmental arrest of the primitive lung during embryonic life.

Case Report

A 27-year-old woman, is presented in the emergency service of the University Hospital of Lung Diseases "Shefqet Ndroqi" with dry cough, difficulty in breathing, fever 370C for about three months. She is carrying her first pregnancy (12-th week). During this period she had not received medication. In her life, she does not refer to any concomitant diseases, except the frequency of "seasonal flu". According to the patient and confirmation by an obstetrician-gynecologist, the menstrual cycle has been irregular once /3 months. She has been treated with hormone therapy (Clomide) and vitamin therapy and is in the 12th week of pregnancy. She was a lifelong non-smoker, used no illicit drugs, and she is a worker in a hook factory. In the family history does not refer to having had patients with tuberculosis, bronchial asthma or cancer. The patient presents with slightly altered health conditions, conscious, oriented.

On physical examination: It is seen a ascertained retraction of the left hemithorax, dullness in percussion and diminished vocal fremitus and breath sounds on the left hemi thorax base. Heart with rhythmic tones, without pathological sounds and murmurs, frequency 80 bmp, BP 110/79 mmHg. Abdomen is soft, treatable, not dolent. Liver under the costal arch, spleen is not palpable. Blumberg negative. Pathological lymph nodes are not palpated. Examination of the sensory - motory nervous system without pathological data. Free limbs with normal mobility, without oedema.

Pulmonary clinical examination suggested the presence of consolidation, atelectasis or pleural effusion: A chest radiography (Figure 1-3) was performed. Chest radiography showed a homogenous density in the lower part of the left hemi thorax with deviation of the trachea and mediastinum to the left. Blood gas analysis at the entrance: pO2 52.0 mmHg, pCO2 31.1 mmHg, pH 7.415, HCO3- 19.5, BE -4.0, SaO2 87.6%. Normal blood tests results. The patient, in this conditions , is hospitalized with the following diagnosis: left pulmonary atelectasis with secondary infection, respiratory insufficiency type I and treated with amoxiclav 1.2 g 2x1fl iv, O2 therapy 1-2 L/min, multivitamin 3x1 tb.

From thoraco-abdominal echo examination is found: the left hemidiaphragm without mobility during respiration; the free phrenico-costal sinuses; left renal agenesis.

ECG: sinusal rythm, P pulmonale, right bundle branch block.

Transthoracic cardiac echo: Very difficult window. Dislocated heart due to pulmonary pathology. The left ventricle is in normal dimensions; no damage to segmental kinetics is observed; systolic function is normal. Thin mitral flaps, with normal movement. Minimal mitral regurgitation. Three sigmoid aortic valve, with normal opening. No transvalvular gradient. Right cameras are difficult to visualize. No tricuspid regurgitation is recorded. No PsAP is recorded. No pericardial effusion.

Thoracic CT was performed to determine the cause of the alleged atelectasis: complete atelectasis of the left lung with short-blind left main bronchus and compensatory hyperinflation of the right lung. Unique right lung with total displacement of the mediastin to the left, as well as hypertrophy and herniation of the right lung tissue left, no pleural fluid, no bone and thoracic wall lesions. Left renal agenesis. Normal aspect of liver, spleen, pancreas and adrenal glands.

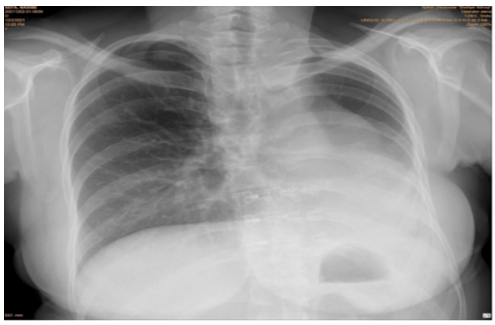

Figure 1: Mediastinum displaced to the left. Left-pulmonary field reduction.

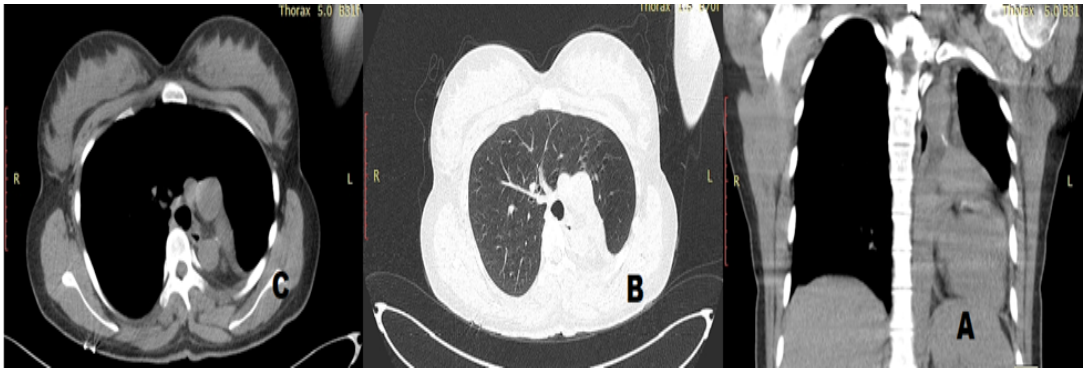

Figure 2: CT scan showing complete atelectasis of the left lung with short-blind left main bronchus and compensatory hyperinflation of the right lung.

It is diagnosed with Left pulmonary aplasia, unique right kidney. On discharge from hospital: pH 7.437, pO2 102.1, pCO2 32.6, HCO3- 21.5, BE -1.9, SaO2 97.9%. The patient is recommended to be followed by the family doctor, pulmonologist for the possibility of secondary infections of the lower respiratory tract and the obstetrician. The patient then had several hospitalizations in the maternity ward between 12-30 and 36 weeks. The performance of vital parameters and biochemical examinations has been normal. At 36 weeks dyspnea is amplified and the patient undergoes cesarean delivery with spinal anesthesia. Baby weighing 2,400 at birth, with no organic abnormalities. After one year of birth, she is re-hospitalized with a history of frequent respiratory infections. She complained from time to time of dry cough, shortness of breath, body weakness. In thoracic CT- scan with IV contrast: Total absence of the sinister pulmonary artery; millimeter rudiment of the primary sinister bronchus (about 5x3 mm). Spirometry is performed:FVC 69.4%, FEV1 73.3%, FEV1/FVC 90.95% (moderate restrictive dysventilatory syndrome).

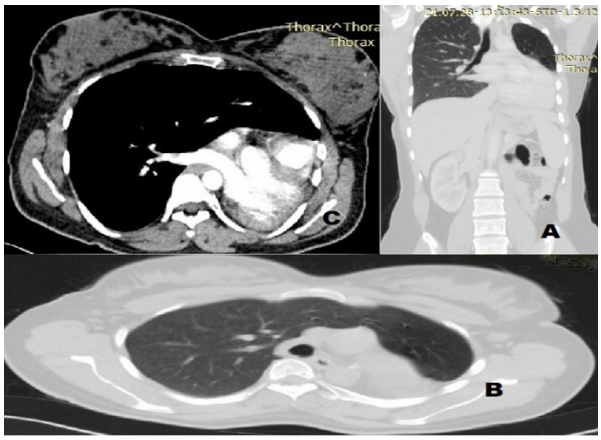

Figure 3: Thoracic T scan with IV contrast showing total absence of the sinister pulmonary artery; millimeter rudiment of the primary sinister bronchus (about 5x3 mm).

Discussion

In 1673, De Pozze described the first ever case of pulmonary agenesis in a female cadaver, and Muhamed reported the first case of unilateral lung agenesis from India in 1923 in an autopsy. Pulmonary agenesis is a rare congenital anomaly with an incidence between 0.0034% and 0.0097% [11]. Most of these patients with associated anomalies present in childhood and are diagnosed earlier. However, those with only lung aplasia, can remain asymptomatic for several years or can have recurrent lower respiratory tract infections and diagnosis can be delayed due to confusion with other common entities [11].Although the diagnosis is often made during childhood, asymptomatic patients can be diagnosed later due to the absence of comorbid anomalies [8]. It can be isolated or associated with congenital malformations, most often with heart abnormalities. It is a rare congenital anomaly. The main etiology of the disease is unknown while genetic, iatrogenic and viral factors as well as vitamin A deficiency during early pregnancy can result in failure to develop primitive lung germ.

In 1912, Schneider originally classified lung agenesis in three groups which was later modified and made more practical by Boyden in 1955. Depending upon the degree of defect, lung agenesis patients can be classified into three groups. Group 1 (Agenesis): Complete absence of bronchus, lung and pulmonary vasculature; Group 2 (Aplasia): Rudimentary bronchus is present but lung parenchyma and vessels are absent; Group 3 (Hypoplasia): Hypoplastic lung, bronchus or vasculature is present. In our case, patient had Type 2 - left lung aplasia with only left renal agenesis and hence, had a better prognosis [8,11]. Right lung agenesis is more frequently associated with congenital anomalies and has poor prognosis as compared to left lung agenesis [11]. Left sided lung agenesis is present in 70% cases and is associated with a more favourable outcome [12].

Our patient was examined in the third month of pregnancy due to respiratory symptoms. Several maternal physiological changes occur during pregnancy due to the growing of the uterus and hormonal changes. This changes are more relevant in unilateral aplasia. There are changes in the shape of the thorax and the diaphragm rises into the thoracic cavity. The increased level of oestrogen causes the nasopharynx mucosa to become hyperemic and oedematous, leading to nasal congestion. The ventilation increases. The effects of progesterone and the central nervous system chemoreceptors cause hyperventilation, which leads to an increase in PaO2 and a decrease in partial pressure of carbon dioxide in the maternal blood, allowing for a gradient conducive to carbon dioxide transfer from foetal to maternal circulation [13]. Symptoms of pulmonary agenesis are unspecific and their apperance changes between individuals [14]. These factors increase the difficulty of the diagnosis. Therefore, there is considerable delay in diagnostic time, even if it is possible to discover pulmonary agenesis at or pre-birth [14]. Our patient at the end of the first trimester of pregnancy had exacerbation of respiratory symptoms and a decrease in PaO2. These due to increased oxygen requirement during pregnancy, as demanded by the growing fetus, placenta and maternal organs in women with compromised systems.

The chest radiograph shows a homogenous opacity, loss of hemi thorax volume with displacement of the mediastinal structures and elevation of the hemi diaphragm. Compensatory contralateral lung hyperinflation is common with normal lung herniation in the contralateral hemi thorax. CT may demonstrate a rudimentary main bronchus. Pulmonary angio-CT is the confirmatory test for ipsilateral absence of pulmonary vasculature. Echocardiography is mandatory for ruling out cardiac anomalies and pulmonary hypertension. Pulmonary hypertension is a common complication of lung agenesis due to loss of pulmonary vasculature.

Left sided lung agenesis is present in 70% cases and is associated with a more favourable outcome [12]. Most patients with associated anomalies present in childhood and are diagnosed earlier; however patients with only lung agenesis, as our patient with only concomitant renal agenesis, can remain asymptomatic for several years or can have recurrent lower respiratory tract infections and diagnosis can be delayed due to confusion with other common entities.

In a normal pregnancy, oxygen consumption and metabolic rate are increased because of the added demands of the products of conception. Both are even higher during labour and delivery. Given that the rise in tidal volume is limited in patients with a restrictive physiology, worsening of symptoms is expected as pregnancy progresses and also during labour. For the same reason, pregnant individuals with restrictive disorders, such as our patient, tolerate exercise poorly, since exercise requires increments in tidal volume, in addition to those needed in pregnancy. Delivery plans should be discussed with the obstetric anesthesiologists and usually include early epidural anesthesia to minimise the rise in minute ventilation and oxygen consumption as well as avoiding the Trendelenburg position and high levels of anesthesia to avoid an aggravation of the restrictive disorder. Depending on the degree of restriction at baseline, further aggravation may lead to respiratory compromise with difficulties with ventilation and oxygenation and respiratory failure [9].

Oxygen consumption during a uterine contraction may triple. Maternal oxygen reserve is decreased due to this increased consumption and decreased functional residual capacity [13].

The patient had a successful delivery by caesarean section with epidural anesthesia and a healthy child. Successful treatment is multidisciplinary.

Conflict of Interest: There is no Conflict of Interest.

Ethical Consideration: None.

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