Case Report

Left lung agenesis discovered by a spontaneous pneumothorax in a 20-year-old girl

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ABSTRACT

Lung agenesis is a rare condition which prognosis widely depends on associated malformations. Clinical presentation is so variable and diagnosis is often made in childhood. Here, we present a case of a 20-year-old girl who was admitted because of a spontaneous pneumothorax. Explorations concluded at a left lung agenesis, a hyperinflated right lung crossing the midline with a corresponding pneumothorax. There was no malformation else. This congenital condition and treatment for this rare presentation are discussed in detail.

KEY WORDS: Congenital, lung, pneumothorax, surgical treatment

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INTRODUCTION

Unilateral agenesis of the lung is an extremely rare condition.[1] More than half of patients have associated malformations.[2] Rarely detected in the intrauterine life, lung agenesis is often discovered in childhood after repeated pulmonary infections, frequent hemoptysis, cardiorespiratory failure, or as part of polymalformative syndrome.[3-5] Here we report a case of a 20-year-old girl presenting a left lung agenesis discovered by a spontaneous pneumothorax.

CASE REPORT

A 20-year-old girl was admitted to our hospital because of a brutal chest pain. Physical examination revealed increased respiratory rate and absence of breath sounds on left of chest. A chest radiograph revealed a bilateral pneumothorax and a left basithoracic opacity [Figure 1]. The chest CT-scan revealed the absence of left main bronchus and left lung parenchyma, and the existence of a right lung hyperinflated crossing the midline with a corresponding pneumothorax [Figure 2]. An echocardiography confirmed the absence of left pulmonary artery without any malformation else. An abdominal CT-scan showed no abnormalities. A surgical pleurodesis was indicated for our patient because of a spontaneous pneumothorax occurring on a single lung. In per operative, by right thoracotomy, there was no left pleural cavity, a cardiac torsion and a herniated right lung presenting many blebs [Figure 3]. Pleurodesis consisted on a mechanical abrasion of the parietal pleura. The postoperative course was uneventful and our patient was discharged on postoperative day 4.

DISCUSSION

Lung agenesis is an extremely rare malformation whose prevalence is estimated at 34 per million live births.[3] The true incidence of this anomaly is unknown because 50% of cases are stillborn and more than 20% die at birth or during their first few months.[6] Right agenesis is rarer but more severe than the left, probably because of an excessive

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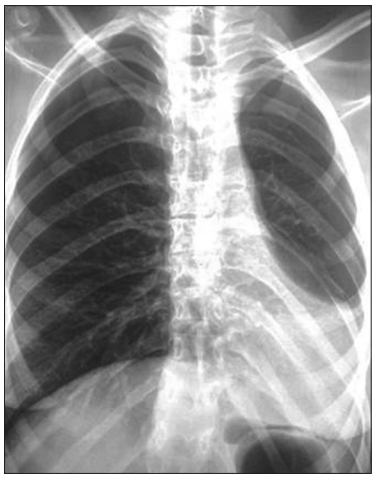

Figure 1: Chest X-ray showing a bilateral pneumothorax, a left basithoracic opacity, and an abnormal cardiac silhouette

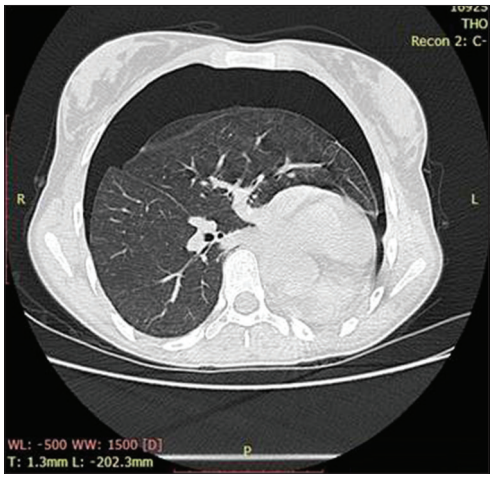

Figure 2: Chest CT-scan showing the absence of the left lung, a right lung crossing the midline with a corresponding pneumothorax, and a posterior push of the heart

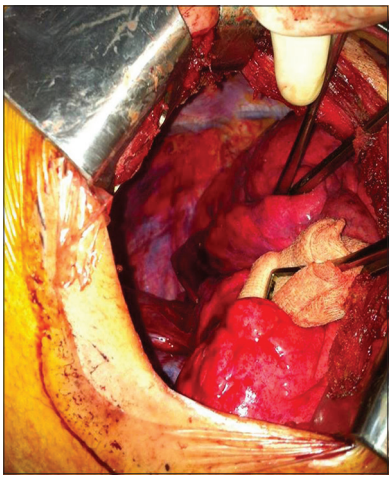

Figure 3: Per-operative view by right thoracotomy. There is no left pleural cavity. The two internal thoracic arteries are unusually seen by this view

carina shift and an improper drainage of the functioning lung.[4,5]

Lung agenesis is associated in 50 -75% with other malformations, especially cardiovascular, urogenital, gastrointestinal and osteoarticular ones.[1,2,6] Mortality is higher and earlier in these cases, generally within the first 5 years of life.[2,5]

Schneider[7] classified this malformation into three categories that has been subsequently modified by Boyden.[8]

Type I (Agenesis): Complete absence of lung, bronchus, and blood vessels to the affected side.

Type II (Aplasia): Existence of a rudimentary bronchus with complete absence of lung tissue.

Type III (Hypoplasia): Presence of variable amounts of lung parenchyma, bronchial tree, and supporting vasculature.

Our patient has been classified as type I.

Normal development of the lung begins in 26th day of intrauterine life from the foregut, and continues postnatally.[2] A disturbance during the 4th week of gestation could cause an unequal division between the two lung buds resulting in a unilateral agenesis or hypoplasia.[5]

Many factors were suggested to be responsible for this defect, such as consanguinity, deficiency of A vitamin, intrauterine infections, and environmental agents and drugs,[3,5] but the exact etiology is still unclear.

Clinical presentation is remarkably variable. Lung agenesis is often discovered in childhood after repeated pulmonary infections, or as part of polymalformative syndrome. A pneumothorax as a discovery circumstance has, to our knowledge, never been reported.

The absence of lung parenchyma is not usually evident at the x-ray chest because of a controlateral upper lobe hypertrophy crossing the midline. The resulting image of basithoracic opacity may mimic lobar collapse, pleural effusion, or diaphragmatic hernia.

The chest CT scan or MRI is the clef of diagnosis. Bronchoscopy may be useful to demonstrate rudimentary bronchus. Other explorations are indicated to detect associated malformations. In our case, echocardiography and abdominal CT-scan were negative.

Treatment of spontaneous pneumothorax may be medical or surgical. Surgery is indicated in case of persistence despite drainage or incomplete lung re-expansion, first recurrence, or bilateral pneumothorax.[9,10] First episode of spontaneous pneumothorax is also an indication for surgery when that affects a single lung.[11]

Surgical treatment is not easy because of working on the functional lung that cannot be conventionally

excluded. Several options may be used: Low tidal volume ventilation,[12] selective lobar isolation,[13] a cardiopulmonary bypass[14] or extracorporeal membrane oxygenation.[15] In our case, we have opted for a conventional thoracotomy with low tidal volume ventilation, with a good outcome.

Prognosis depends essentially on associated malformations. Tomomi[16] reported a series of eight patients having unilateral pulmonary agenesis or aplasia with cardiac or tracheal associated malformations. In this series, intrahospital mortality after surgical repair exceeded 30%. Isolated unilateral lung agenesis may, however, be compatible with a normal life.[5] Successful pregnancy[17] and survival to adult age (up to 77 years)[18] are reported.

CONCLUSIONS

Lung agenesis is a rare but a serious condition which should be early diagnosed and followed up. Prognosis, apart from malformations, is that of a single functional lung. Pneumothorax occurrence is dangerous and should be surgically treated.

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Conflicts of interest
There are no conflicts of interest.

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