Congenital Left Upper Lobe Agenesis: Report of a Case

Alkin Yazicioglu*, İbrahim Onur Alici and Erdal Yekeler

Turkiye Yuksek Ihtisas Training and Research Hospital, Department of Thoracic Surgery and Lung Transplantation, Ankara, Turkey

Abstract: Lobar agenesis is an extremely rare condition which is characterized by the absence of the lobar pulmonary artery, pulmonary vein, bronchi and parenchyma. Most frequently the affected lobe is the left upper lobe, and patients usually survive to older ages. In the report below we present a case of the left upper lobe agenesis and bronchiectasis of the superior segment of the ipsilateral lower lobe. The thoracic computed tomography scan and fiberoptic bronchoscopy are important diagnostic tools. The patient remained problem-free after the segmentectomy of the bronchiectatic parenchyma.

Keywords: Agenesis, Bronchiectasis, Lobar, Pulmonary.

INTRODUCTION

Pulmonary agenesis is an uncommon congenital anomaly which is usually diagnosed in pediatric ages, whereas, the exact incidence is not well defined. In their case series, Mardini *et al.* reported an incidence between 0.0034 - 0.0097% for unilateral pulmonary agenesis [1]. In the pulmonary agenesis, the lung parenchyma, pulmonary artery, pulmonary vein and bronchi of the associated lobe are all absent. As a matter of course bilateral pulmonary agenesis is incompatible with life [2]. Lobar pulmonary agenesis is an extremely rare situation, and an affected newborn usually survives to older ages as the pulmonary parenchyma of the unaffected sites are functionally normal.

CASE REPORT

A 43 year-old male was admitted to our hospital with complaints of cough and purulent sputum which had worsened over several months. His past medical history was clear and physical examination was unremarkable except for the presence of coarse crackles on the upper zone of the left hemi-thorax. Also there were wheezing on the same zone. His effort tolerance was perfect. All laboratory investigations were normal and pulmonary function test showed adequate lung volume for an anatomical resection. His pulmonary function test results were FEV1: 2,80 L - 73% of predicted, FVC: 3,60 L - 80% of predicted and their ratio was 0,77.

The chest radiography demonstrated a bronchiectatic field in the left upper zone. Computed tomography (CT) of the thorax revealed the mediastinal shift towards the left hemi-thorax due to the volume loss of the left lung together with a bronchiectatic parenchyma on the area attributed to the left upper lobe (Figure **1a-f**). With the initial diagnosis of bronchiectasis, the patient underwent a fiberoptic bronchoscopy. During bronchoscopy we realized that the left main bronchus continued with left lower lobe bronchus without giving a branch to left upper lobe (Figure **2**).

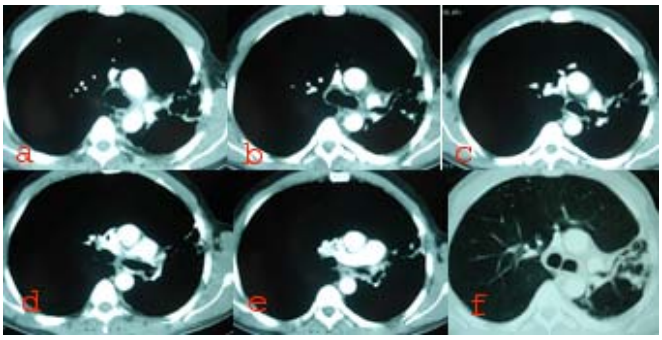

Figure 1a-f: Thoracic CT revealed there was no pulmonary artery branch to the upper lobe and the bronchiectatic changes of the suspected left upper lobe.

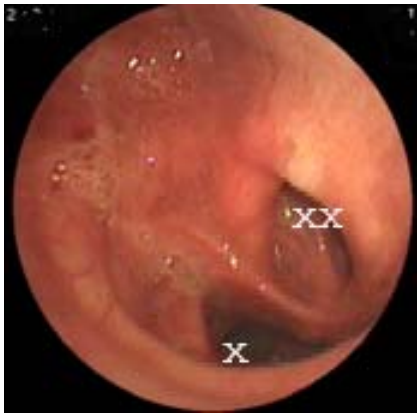

Figure 2: Bronchoscopy revealed there was no bronchial way to the left upper lobe. Two orifices were headed to the lower lobe; superior segment (asterix) and other segments (double-asterix).

*Address correspondence to this author at the Alacaatli mah. 3272. Cad. Sonmez Residence 4/11 Yasamkent/ Cankaya/ Ankara/ Turkey; Tel: +905334177985; Fax: +903123552135; E-mail: yazicioglu.md@gmail.com

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With a diagnosis of bronchiectasis we decided to perform an operation to the bronchiectatic field. Before general anesthesia an epidural catheter was inserted for analgesic control and after anesthesia endotracheo-bronchial tube was deployed. A left posterolateral thoracotomy was performed through the fifth intercostal space and the bronchiectatic field of the left lower lobe superior segment was observed. During the resection, it was also observed that there was no parenchyma which would be attributed to the left upper lobe with a distinct fissure. The left main pulmonary artery readily continued to the left lower lobe without giving branches of the upper lobe; there was no pulmonary vein of the left upper lobe also. The left main bronchus continued directly to the lower lobe branch without giving an upper lobe branch. The bronchiectatic segment was related to the superior segment of the lower lobe. Therefore, the patient was diagnosed as left upper lobe agenesis with a bronchiectasis of the lower lobe superior segment. An anatomical segmentectomy was performed to the lower lobe superior segment. During the operation there was no anesthetic management discomfort. After the operation, the patient had an uneventful recovery and was discharged on the sixth post-operative day. The pathologic diagnosis was consistent with an ordinary bronchiectasis.

DISCUSSION

Agenesis of the lung is an uncommon developmental defect in which there is complete absence of one or both lungs. The essential components of a functional lung:- parenchyma, pulmonary artery and vein, and bronchus-, are all absent. This developmental anomaly was first described by de Pozzis in 1673 and the first proposed classification of underdevelopment of the lung was introduced by Schneider in 1912 (Class I - Agenesis, total absence of bronchus and lung; Class II - Aplasia, rudimentary bronchus without lung tissue; Class III - Hypoplasia, bronchial hypoplasia and variable but reduced amount of lung tissue) [3]. In 1977, Spencer divided pulmonary agenesis into (1) bilateral complete agenesis, (2) unilateral agenesis with (a) complete absence of bronchi, (b) rudimentary bronchus present but no pulmonary tissue, (c) poorly developed main bronchus with poorly organized parenchyma, and (3) lobar agenesis [4,5]. Agenesis of one or two lobes of a single lung is an extremely rare condition and the exact incidence of this anomaly is not known. According to Berrocal *et al.* the most affected lobe is the left upper lobe [6].

The etiology of pulmonary agenesis is not well defined, although, genetic, teratogenic and mechanical factors have been suspected as possible causes [6]. The agenesis of the pulmonary tract might have been an embryologic developmental pathology. The tracheo-bronchial groove appears in the fourth week of gestation and lung buds are supplied by a pair of bronchial arteries [5]. An unknown reason for interruption of the arterial development is postulated to result in necrosis of an established primitive lung bud and smaller distal airways [5]. Unavoidably, pulmonary or lobar agenesis occurs.

Associated congenital abnormalities, such as cardiovascular diseases (more frequently patent ductus arteriosus and patent foramen ovale), spinal, genitourinary and musculoskeletal pathologies has been reported [1,5]. Mardini *et al.* presented four cases with pulmonary agenesis; all of them had congenital heart defects, and three of them had musculoskeletal pathologies [1]. The coexistence of these malformations supports the embryologic underdevelopment during the fourth week of gestation.

Thoracic CT is helpful to determine lobar agenesis with an associated mediastinal shift to ipsilateral hemi-thorax, a normal contra-lateral lung with a compensatory hyperinflation and a herniation across the midline [6]. In lobar agenesis, the volume of the affected hemi-thorax is reduced; as a result, a mediastinal shift towards the affected site and the herniation of the contra-lateral lung could be observed [5]. The contra-lateral lung is in normal structure and function, but it has been subjected to compensatory hypertrophy [3]. In our case, there was a mediastinal leftward shift and a compensatory hyperinflation with a herniation of the contra-lateral normal lung.

Fiberoptic bronchoscopy plays a major role to determine the lobar anatomy of the bronchial system. In lobar agenesis cases, there is no bronchial branch to the affected lobe; our case had no left upper lobe bronchus. The left main stem bronchus directly continued with the lower lobe and divided into two branches: one for superior segment of the lower lobe and the remaining was travelling caudally to give basal segments of the same lobe. Pulmonary angiography is helpful in demonstrating the absence of pulmonary and bronchial arteries of the absent lung or lobe [6].

As the patients may easily survive without an undeveloped lobe, the true prevalence of the condition

is yet undefined and the possibility of this uncommon developmental pathology should be kept in mind. Thoracic CT scans, fiberoptic bronchoscopy and pulmonary angiography are important diagnostic tools; and the combination of these diagnostic methods leads the clinician to the suspicion of pulmonary or lobar agenesis.

CONFLICT OF INTEREST

Authors declare that there is no conflict of interest.

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