CONGENITAL MALFORMATION OF LUNG: LOCAL BRONCHOSCOPIC EXPERIENCE

Ghulam Shabbier, Najmul Hassan Shah, Akhtar Sherin and Ahmad Fawad

Department of Medicine,
Khyber Teaching Hospital, Peshawar.

SUMMARY

A retrospective analysis of all bronchoscopic procedures from 1998 till 2001 was performed at Khyber Teaching Hospital, Peshawar was carried out to determine the frequency of congenital malformations of lung. Both the lesions were agenesis of left lung. It is concluded that these lesions although rare may be considered in the appropriate clinical circumstances.

INTRODUCTION

Congenital malformations of lung are rare findings on bronchoscopy. Anatomic variations in the airways and congenital agencies of lung are the two conditions usually encountered on bronchoscopy. Usually these malformation are overlooked, while using other conventional investigative modalities, of these agenesis of lung has serious implications for patients, as they are usually wrongly investigated and treated for repeated chest infections.[1] To determine the frequency of congenital malformations of lung on fiberoptic bronchoscopy.

MATERIAL AND METHODS

The reports of bronchoscopies performed at bronchoscopic unit of Khyber Teaching Hospital Peshawar between 1999 to 2001 were retrospectively analyzed for frequency of congenital malformations of lung. We were looking for malformations like tracheal agenesis, tracheal stenosis, tracheobronchomegaly, anatomical variations of bronchi and agenesis of lung.

RESULTS

Total bronchoscopies performed during the period 1999 to 2001, were 176. Out of these, 98 patients were male and 78 were female. Aged ranged between 15 years to 66 years with a mean of 35 years and median of 42.5 years. The only congenital malformation encountered was agenesis of left lung in two patients. Both patients were female. One of the patients was 25 years old from Kohat and other was 21 years old

from Mardan. In 25 years old lady bronchoscopy reported as complete obliteration of left bronchus about 1 to 2 cm below the carina, mucosa was normal looking and the bronchus had formed a cul de sac. While in 21 years old lady bronchoscopy revealed left main bronchus suddenly ending 2 cm below the carina in a cul de sac with a normal looking mucosa.

DISCUSSION

Congenital anomalies of the lung are rare. These include anatomical variations in the trachea and bronchi, of which the bronchial isomersion syndromes are more common.[2] Congenital malformations of trachea include agenesis (aplasia, absence), tracheal stenosis due to inherent defect of trachea and tracheobronchomegaly (bilateral right or left) have been identified.[3]

Agenesis of lung, a rare congenital anomaly has been reported with various degrees of developmental deficiency of the lung tissue that is bilateral pulmonary agenesis,[4] unilateral lung agenesis,[1] subtotal agenesis of two lobes[5] and a single lobe agenesis.[6] Bilateral lung agenesis is very rare. Unilateral lung agenesis is more common and over 200 cases have been reported.[2] Left lung is more commonly absent than the right.[7,8] Some confusion exists in use of terms agenesis, aplasia and hypoplasia but most authors have followed the following classification.[2,8,9,10]

1. Agenesis: complete absence of an entire lung and its airways with no vascular supply to the effected side.
2. Aplasia: small tracheal outpouching or rudimentary bronchus but no lung parenchyma or blood vessels surrounding it.
3. Hypoplasia: lobar agenesis and lung that is smaller than normal but in which the major bronchial subdivisions are preserved, although the lung tissue is poorly developed.

Agenesis of lung is usually diagnosed in infancy but may occasionally remain undiagnosed until adult life.[8] Half of all the reported patients die either at birth or within the first 5 years of life.[11]

The etiology of these malformations is unknown. About 60% of patients with agenesis of lung are said to have other congenital anomalies like vertebral defects, congenital heart diseases, tracheoesophageal fistula, genito urinary, gastro intestinal and limb malformations.[12,13,14,15,16,17,18] The association with other congenital anomalies appears to be greater with agenesis of right lung than left lung.[9] The extent and severity of associated anomalies are reflected in the prognosis.

Agenesis of lung itself doesn't give rise to symptoms unless complicated by bronchopulmonary diseases and other associated anomalies. Provided it is not associated with other major anomalies it may allow long survival and there are several reports of unilateral lung agenesis demonstrated at autopsy in adults with no history of respiratory complaints who died of other pathology.[2] However many patients die before the second decade of life.[9] Patients usually present with repeated chest infections. Patients with lung agenesis are more prone to infections as mediastinal shift and malrotation of carina hinder drainage of functioning lung.

Typical signs in agenesis of lung are elevation of the corresponding hemidiaphragm, compensatory hypertrophy of the contra lateral lung, mediastinal, tracheal, and cardiac shift to the defective side and a dense homogenous opacity on the involved side. Tomography, bronchography, bronchoscopy and angiography may be required to establish the degree of under development or to differentiate agenesis from other conditions.

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