Lobar Agenesis of the Left Upper Lung

— A Case Report —

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Lung agenesis is a rare developmental anomaly. It can range from total bronchial and parenchymal agenesis to mild pulmonary parenchymal hypoplasia of one or both lungs.

A case of lobar agenesis of the left upper lung in a 15-year-old boy is presented. The patient had mild exertional dyspnea. Pulmonary angiography revealed the absence of the left upper pulmonary artery and vein. Bronchography showed no branching of bronchus to the left upper lobe. Intravenous pyelography revealed incomplete duplication of the right urinary tract.

Key Words: Lung agenesis, Congenital malformation

INTRODUCTION

Underdevelopment of the lung is a rare developmental anomaly in which there is complete absence (agenesis) or hypoplasia of one or both lungs (Borja et al., 1970; Swischuk et al., 1979). The majority of reported cases had associated congenital anomalies of cardiovascular, musculoskeletal, or gastrointestinal systems. Anomalies of genitourinary or central nervous system were also reported (Mendelsohn et al., 1977; Knowles et al., 1988 Say et al., 1979).

We describe a case of lobar agenesis of the left upper lung with incomplete duplication of the right urinary tract.

CASE REPORT

A 15-year-old male who had a heart murmur detected at Asan Hospital was referred for investigation of the cardiac problem. There was a history of shortness of breath on exertion.

Physical examination showed a normal boy with body weight 53kg (50th percentile) and height 165cm (50th percentile). There was no deformity of the chest wall. The peripheral pulses were normal with a regu-

la rate of 80 beats/min, and the blood pressure was 110/70mmHg. The first heart sound was normal, the second heart sound was widely split and fixed. A grade 2-3/6 systolic murmur was audible over the upper left sternal border and left infraclavicular region. There was no ejection click. The lung was clear and the liver was not enlarged.

The electrocardiogram demonstrated a sinus rhythm of 80 beats/min without abnormal findings. A two-dimensional echocardiogram showed no evidence of structural abnormalities. The chest radiograph showed a marked shift of the mediastinal structures to the left side with a decrease in size of the left hemithorax, and evidence of an increase in volume of the right lung (Fig. 1). The patient underwent cardiac catheterization, and the measured oxygen saturations were as follows: superior vena cava 78%, inferior vena cava 81%, mid-right atrium 80%, right ventricular apex 81%, left ventricle 96%, and ascending aorta 95%. The pressure measurements were right ventricle 34/0-4 mmHg, main pulmonary artery 31/10 mmHg, left ventricle 105/0-9 mmHg, and ascending aorta 115/70 mmHg. Pulmonary angiography revealed absence of the left upper pulmonary artery (Fig. 2) and vein. The aortogram showed right-sided aortic arch. The bronchography of the left side showed tracheal shift to the left hemithorax and no upper lobe bronchus (Fig. 3). A chest CT revealed herniation of some portion of the right upper and middle lung into the left side through the anterior retrosternal space.

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Fig. 1. Chest radiograph shows a marked shift of the mediastinal structures to the left side with a decrease in size of the left hemithorax, along with evidence of an increase in volume of the right lung.

Fig. 3. Brochogram shows the absence of the left upper lobe bronchus and the tracheal shift to the left hemithorax.

Fig. 2. Pulmonary arteriogram illustrates the absence of the left upper pulmonary artery.

A pulmonary function test revealed mild to moderate obstructive type of ventilation. The measured findings of pulmonary function were as follows: FVC 111%, FEV1 77%, FEV1/FVC 61%, FEF25-75 37%, FET25-75 193%, and MVV 68%. Intravenous pyelography revealed duplication of pelvis and upper ureter of the right urinary tract.

Surgical treatment was not attempted. He was discharged to be followed up at the out-patient clinic.

DISCUSSION

Underdevelopment of the lungs can range from total bronchial and parenchymal agenesis to mild pulmonary parenchymal hypoplasia (Borja et al., 1970; Mendelsohn et al., 1977). The majority of reported cases had associated congenital defects involving cardiovascular, musculoskeletal, gastrointestinal, or genitourinary systems (Knowles et al., 1988; Say et al., 1979; Costas et al., 1977).

The incidence of this anomaly is not known. A paper reported a prevalence of 0.0034 percent among hospital admissions (Borja et al., 1970). There appears to be no sex predilection in this condition (Steinberg et al., 1966).

Pulmonary underdevelopment (agenesis) has been classified morphologically by the extent to which bronchopulmonary tissue is absent. Earlier classification were modified by Spencer (1977) who divided pulmonary agenesis as follows: (1) bilateral complete agenesis; (2) unilateral agenesis with (a) complete absence of bronchi, alveolar tissue, and blood supply, or (b) rudimentary bronchus present but not invested by pulmonary tissue, or (c) poorly developed main bronchus with fleshy mass and poorly organized parenchyma; and (3) lobar agenesis or lesser forms of congenital anomaly. A more useful clinical classification based on angiographic data (Mardini et al., 1985) would di-

vide the patients first into two groups: one in which there is the total absence of pulmonary parenchyma and the ipsilateral pulmonary artery in the absence of any systemic arterial supply, and the other in which there is hypoplastic pulmonary tissue. This group can be subdivided into two subgroups: one in which the affected lung receives its blood supply from the pulmonary artery, and the other in which the affected lung is sequestered and receives a large amount of systemic arterial blood supply. Pneumonectomy or lobectomy is required in the first subgroup only when recurrent respiratory infection results from abnormal bronchopulmonary tissues. Surgery may be indicated in the second group when the systemic blood flow causes congestive heart failure of recurrent respiratory infections.

Cardiac catheterization and angiography are essential procedures for diagnosis and elucidation of anatomic abnormalities and identification of associated cardiac defects (Mardini et al., 1985). Bronchography is also required to evaluate the defect. A chest CT or MR imaging may be of considerable value in the assessment of congenital pulmonary anomalies including this defect (Naidich et al., 1988). Bronchoscopy may be indicated in a patient with repeated infections in a hypoplastic lung who may be a candidate for surgery (Mardini et al., 1985).

Patients with pulmonary agenesis may be asymptomatic, and the diagnosis may not be made until adulthood (Shenoy et al., 1979). However, there are usually some pulmonary manifestations including exertional dyspnea as in our case. Cyanosis and respiratory difficulties may be observed in early infancy. Pulmonary hypertension and congestive heart failure can result from changes in the pulmonary arteriolar system in the contralateral lung (Dimich et al., 1967). Tracheal compression has been reported to be caused by a normal but deviated aorta associated with right-sided pulmonary agenesis (McCormick et al., 1979).

Right-sided pulmonary agenesis has been thought to have a graver prognosis than that of the left, with death occurring earlier and with greater frequency (Steinberg et al., 1966; Smith et al., 1958). The worse prognosis for the right-sided defect could be due to a higher incidence of associated cardiac abnormalities or to the greater mediastinal shift produced by right-sided agenesis, leading to more significant dis-

tortion of the bronchial and vascular structures (Thurlbeck et al., 1975). Left-sided agenesis has been reported to occur more frequently than right-sided (Valle, 1955).

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