Agenesis of the lung

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We have carefully read the case report "agenesis of the lung" which appeared in the Journal recently (1). We doubt that the data presented are sufficient for establishing the diagnosis of agenesis of the lung versus pulmonary artery agenesis (PAA) with the methods employed, since autopsy was not performed.

At our institution we have diagnosed 4 cases of PAA in the last two years, aged 15–19 yrs, resembling the described case (2). Before admission to our 500 bed hospital, patients were examined at times by various specialists (pediatricians, cardiologists, roentgenologists etc) for repeated respiratory infections since childhood. The initial diagnoses with which the patients were referred were Tuberculosis, McLeod's syndrome, agenesis of the lung, Fallot's tetralogy and lung tumor. All patients had a characteristic chest radiograph (Fig 1) with

1) ipsilateral heart and mediastinum displacement, 2) smaller hemithorax, 3) absent pulmonary artery shadow, 4) ipsilateral hemidiaphragm elevation, 5) ipsilateral absence of vascular pattern and, 6) contralateral lung hyperinflation and herniation beyond the mid line with hyperlucency. Ventilation-perfusion lung scan is the cornerstone of the diagnostic approach, with the perfusion scan showing absence of radioactivity in the affected lung, as was the case described in the Journal [1]. The ventilation scan shows normal or slightly subnormal radioactivity. The perfusion lung scan limits the possible diagnoses to three: PAA, pulmonary artery hypoplasia and pulmonary embolism (3). Other congenital abnormalities (heart abnormalities, right aortic arch, transposition of great vessels etc) are of help in suggesting PAA. Angiography (conventional or digital) confirms the diagnosis. We therefore believe that the described case [1] most probably is PAA.

References


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Since the parents did not give consent for post mortem examination, the autopsy has not been performed in this case, but laboratory studies and clinical course definitively showed that the described case is agenesis of the right lung (1). First of all, anteroposterior view of chest revealed a homogeneously dense right hemithorax, additionally an air shadow of the right main bronchus was not observed.

The overall incidence of associated congenital anomalies was greater than 50%. Patent ductus arteriosus, patent foramen ovale, abnormal origin of great vessels, a vascular ring produced by a right aortic arc and other cardiovascular and non-cardiovascular (genitourinary, skeletal and gastrointestinal) anomalies have been recorded (2). Anomalies of the pulmonary vessels to the affected side were found in all cases in the other report (3). In this report, all had hypoplasia of the pulmonary artery or its branches.

While, chest roentgenograms suggest the diagnosis, careful bronchographic angiocardiographic, and echocardiographic studies, as well as computed tomography in some instances, may be necessary for confirmation.

Half of all reported patients die either at birth or within the first 5 yrs of life. Individuals, with agenesis of the left lung have a longer life expectancy than do those with agenesis of the right lung. Our case of right lung agenesis expired on the forty-fifth day.

As a result, I consider any cardiovascular anomaly to be another associated anomaly in this case.

References


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