Case Report

A Pediatric Case of Pulmonary Hypertension Associated with Congenital Bronchial Atresia

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Congenital bronchial atresia is a rare malformation that causes recurrent lung infections and pneumothorax. Pulmonary hypertension has not been reported as a complication in pediatric population, to the best of our knowledge. Our present patient is a 2-year-old girl with left pulmonary emphysema and pulmonary hypertension. She received pulmonary vasodilators and underwent left upper lobectomy. Pulmonary vasodilators were only partially effective; lobectomy was ineffective for pulmonary hypertension. Six years later after lobectomy, the patient was still receiving pulmonary vasodilators, and her pulmonary hypertension was gradually improving.

Keywords: congenital bronchial atresia, pulmonary hypertension, lobectomy, pulmonary vasodilators

Background

Congenital bronchial atresia is a condition in which the bronchi are closed during the fetal period; mucus secreted by the bronchi is accumulated centrally, forming a mucus plug. Emphysema results from air trapping due to the so-called check valve mechanism during collateral ventilation (such as Kohn's pores).[1)] It is a relatively rare abnormality with an estimated prevalence of 1.2 per 100,000 people and can be associated with recurrent pulmonary infections and pneumothorax.[2)] Congenital bronchial atresia complicated with pulmonary hypertension is extremely rare and has been reported only in adults.[3)] Apparently no case of pulmonary hypertension in childhood has ever been reported, based on our search results. Informed consent for this case report was obtained from the parents.

Case Presentation

A 2-year-old girl was referred to our hospital after a close examination for failure to thrive revealed emphysema on the left lung and pulmonary hypertension. Her height was 84.3 cm and body weight 8.7 kg, being equivalent to −2 standard deviations. Her SpO_2 was 98% on room air at rest. Blood tests showed no abnormal values, including α_1-antitrypsin deficiency that causes an early onset of emphysema and chronic obstructive lung disease. A radiograph showed emphysematous lesions in the left lung field and rightward deviation of the heart silhouette. Echocardiography revealed the following findings of pulmonary hypertension; the ventricular septum was shifted toward the left ventricle ("D shape" in a short axis view) (Fig. 1) and a pressure gradient derived from tricuspid regurgitation was 69 mmHg. A non-enhanced chest computed tomography (CT) scanning (Fig. 2) showed hyperinflation of the left upper lobe, cystic lesions in the A3 branch of the pulmonary artery, and rightward deviation of the mediastinum. These, together with the bronchography and fiber bronchoscopy findings, led to a diagnosis of left B3 bronchial atresia. Cardiac catheterization revealed a mean pulmonary artery pressure (mPAP) of 46 mmHg, pulmonary vascular resistance (PVR) of 8.8 Wood units m^2, pulmonary artery wedge pressure of 9 mmHg, and cardiac index (CI) (the Fick method) of 4.2 L/min/m^2. We did not administer the oxygen challenge test because her

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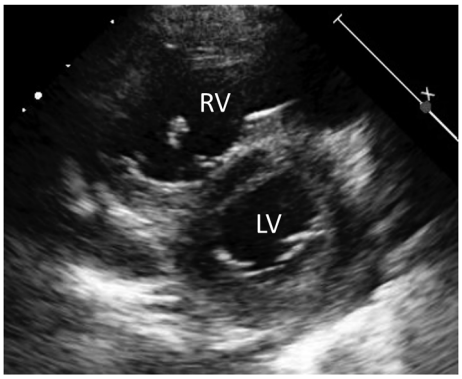

Fig. 1 The ventricular septum is deviated toward the left ventricle, as shown by transthoracic echocardiography. This indicates that a right ventricular systolic pressure is comparable to that of the left ventricle, and thus pulmonary hypertension.
LV, left ventricle; RV, right ventricle

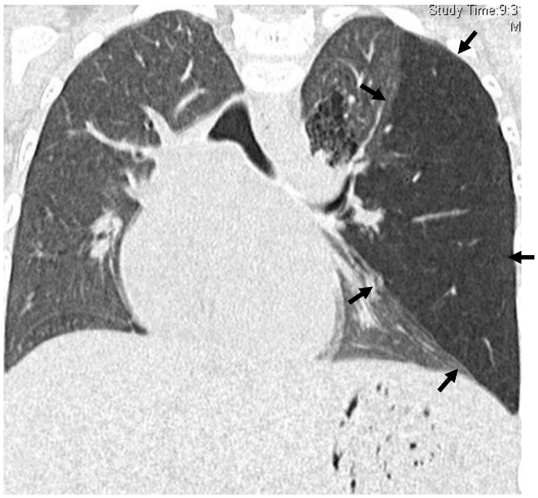

Fig. 2 A coronal section on non-enhanced computed tomography shows hyperinflation of the left upper lobe (arrow).

blood pressure declined throughout the examination. Other diseases resulting in pulmonary hypertension were ruled out. We diagnosed pulmonary hypertension associated with pulmonary disease in group 3 of the Nice classification,[4] although there were no reports regarding pulmonary hypertension due to congenital bronchial atresia in childhood. We administered pulmonary vasodilators to reduce pulmonary artery pressure. The patient was started on macitentan (0.25 mg/kg/day) and tadalafil (1.0 mg/kg/day) sequentially. One year after introduction of these agents, mPAP was 35 mmHg, PVR 5.7 Wood units m^2, and CI (the Fick method) 4.4 L/min/m^2, showing a trend toward improvement, but pulmonary hypertension remained. The oxygen load test indicated its result as reactive. We judged that compression onto the normal lung tissues by emphysematous lesions was one of the causes of pulmonary hypertension. After consulting with our multidisciplinary team, a left upper lobectomy was performed. The perioperative course progressed well without any complications. Pathological findings showed a mucus plaque in the lumen of the B3 bronchus. The finding was consistent with bronchial obstruction. There were no histologic findings suggestive of pulmonary hypertension. Cardiac deviation tended to improve after lobectomy. One year after the procedure, however, echocardiography showed no obvious decrease in the estimated pulmonary artery pressure (tricuspid regurgitation pressure gradient 53 mmHg) comparing with the preoperative figure. The patient continues to be treated with the combination of the two drugs. Thereafter, pulmonary hypertension improved. At 6 years after lobectomy, a peak pressure gradient estimated by tricuspid regurgitation was 35 mmHg on echocardiography, whereas mPAP was 21 mmHg, PVR 3.7 Wood units m^2, and CI (the Fick method) 3.6 L/min/m^2 on catheter investigation.

Discussion

We reported a pediatric case of congenital bronchial atresia complicated by pulmonary hypertension.

It has been mentioned that congenital bronchial atresia is pathogenetically caused by impaired blood flow to the bronchial arteries during fetal life, and that the bronchi close after 16 weeks of gestation when bronchogenesis is completed.[5] In our patient, pathological investigation showed a mucus plaque in the bronchus, which was considered to be the result of bronchial closure. In a previous report, pulmonary hypertension was diagnosed in a 66-year-old woman with dyspnea, and there were no lung lesions other than congenital bronchiectasis.[3] The authors considered its mechanism was prolongedly increased pulmonary blood flow in the healthy lungs in their report. This hypothesis, however, most unlikely apply to our case, because the duration of increased pulmonary blood flow was relatively short to promote pulmonary hypertension in the healthy lungs. Moreover, it was also unlikely that interruption of pulmonary blood flow to the left upper lobe alone would

result in an increase in pulmonary vascular resistance in other healthy areas. We therefore assumed that the emphysematous lesion was compressing the healthy areas, and judged that lobectomy of bronchial atresia lesion should work. As a result, lobectomy alone did not improve her pulmonary hypertension.

According to our research, there are no definitive guidelines for management of congenital bronchial atresia in pediatric patients. Relevant to our case, Zarfati et al. proposed that conservative management appeared safe, and surgery should be reserved for patients with symptomatic or complicated circumstances.[6] We determined that pulmonary arterial hypertension (Nice classification, group 1) was one of the mechanisms of pulmonary hypertension in our patient. We applied use of pulmonary vasodilators partly because the oxygen load test indicated a reactive feature. In reality, pulmonary vasodilators were partially effective in this case. Still, surgical resection was indicated because we contemplated that compression onto the normal regions of the lungs by emphysema was one of the significant causes of pulmonary hypertension. After several years of follow-up, this patient's pulmonary arterial pressure showed a trend of a decrease by continuing intake of two pulmonary vasodilators. The mechanism of pulmonary hypertension remains unclear in this patient. Systemic growth could have been a factor to improve pulmonary hypertension. Although pulmonary vasodilators was seemingly effective, the primary mechanism of pulmonary hypertension might have been hypoplasia of the pulmonary vasculature or the lungs themself in this particular child.

Conclusion

In this report, we described a case of an infant with congenital bronchial atresia who presented with pulmonary hypertension in early childhood. Lobectomy was ineffective, and pulmonary vasodilators were only partially effective. Pulmonary hypertension gradually improved as the patient grew. Careful follow-up is necessary.

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Conflicts of Interest

None.

Author Contribution

Tomomi Fujimura drafted the original manuscript. Masataka Higuchi and Hiroshi Ono established the diagnosis, managed the patient, and contributed to drafting and critical revision of the manuscript. All authors read and approved the final manuscript.

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