Cardiology in the Young

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Surgical management of congenital lobar emphysema associated with CHD

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Brief Report

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Abstract

Congenital lobar emphysema is often associated with CHD in early infancy; however, the surgical strategy for this condition is still controversial. We report three successful cases of emphysematous lung lobectomy before the surgical repair of associated CHD. Aggressive lobectomy preceding cardiac interventions is advised when the management of congenital lobar emphysema is difficult.

In congenital lobar emphysema, the lobes of the lungs become overinflated due to a check valve mechanism; 12–20% of patients with congenital lobar emphysema have CHD.[1] Lobectomy is preferable when conservative treatment is difficult, but it is still controversial whether CHD or congenital lobar emphysema should be treated first.[2] We report three successful cases with aggressive lobectomy preceding cardiac surgery.

Case 1

A girl was born at full-term with retractive breathing. Owing to a heart murmur, echocardiography at 1 month showed a 4-mm ventricular septal defect and a 6-mm atrial septal defect. Radiography showed hyperlucent areas in bilateral lung fields. Cardiac catheterisation showed central venous pressure of 15 mmHg, mean pulmonary artery pressure of 50 mmHg, right ventricular end-diastolic pressure of 15 mmHg, left ventricular end-diastolic pressure of 15 mmHg, pulmonary-systemic blood flow ratio of 2.1, and pulmonary vascular resistance of 6.9 Wood Unit m2. The patient had severe biventricular diastolic dysfunction and pulmonary hypertension. CT was performed, and the congenital lobar emphysema of the right middle and left upper lobes was diagnosed (Fig 1). The right middle lobe and left upper lobe bronchi were not clear on bronchoscopy. As adverse effects of congenital lobar emphysema on circulation were suspected, lobectomy of the right middle lobe, the largest lesion, was performed at 9 months. After lobectomy, respiration improved and cardiac catheterisation showed central venous pressure of 8 mmHg, mean pulmonary artery pressure of 28 mmHg, right ventricular

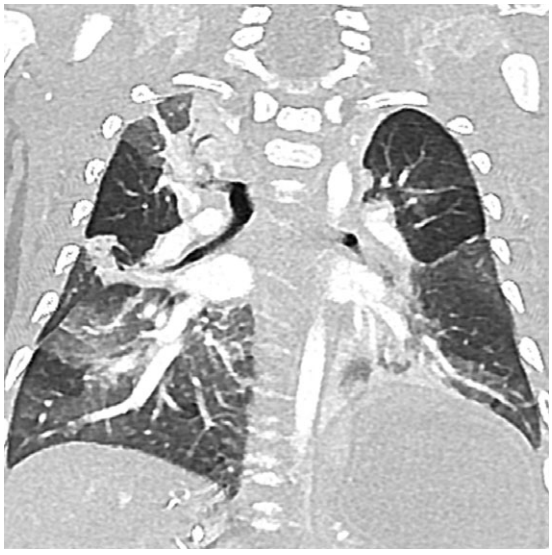

Figure 1. Chest computed tomography shows overinflation of the right middle lobe and left upper lobe. The right upper lobe is compressed by right middle lobe emphysema.

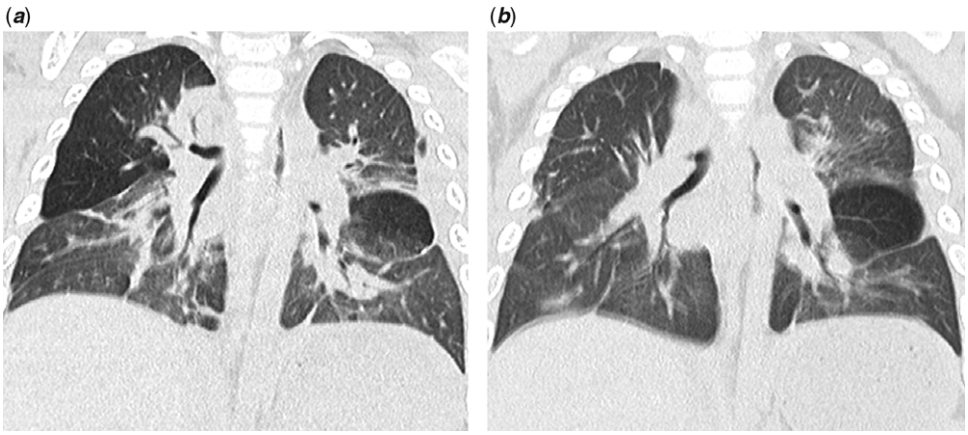

Figure 2. (a) Chest computed tomography shows hyperinflation of the right upper lobe and left lower lobe. (b) Postoperative chest computed tomography shows a residual lesion in the left lower lobe.

end-diastolic pressure of 6 mmHg, left ventricular end-diastolic pressure of 11 mmHg, pulmonary-systemic blood flow ratio of 2.8, and pulmonary vascular resistance of 1.4 Wood Unit m2. As the patient showed an indication for intracardiac repair, surgery was performed the following month, and she was discharged without difficulty in perioperative management. Although there was a residual lesion in the left upper lobe, she had no respiratory disturbance even after 1 year.

Case 2

A boy was born at 36 weeks' gestation weighing 2520 g. He was diagnosed with 21 trisomy, double outlet right ventricle, and complete atrioventricular septal defect. Pulmonary artery banding was performed at 3 months. Retractive breathing and hypoxemia on crying appeared at approximately 5 months of age. CT showed hyperinflation of the right upper and left lower lobes (Fig 2a). Bronchoscopy showed no tracheomalacia or tracheal stenosis. Despite hospitalisations for airway infections and aggressive conservative treatment, the patient developed respiratory failure at 1 year and 6 months, requiring partial resection of the left lung under emergency extracorporeal membrane oxygenation, which was weaned the next day and the patient extubated on the third post-operative day. However, additional resection of the right upper lobe was performed because of severe hyperinflation of the right upper lobe on CT. Although there was a residual lesion (Fig 2b), his respiratory condition was stable and intracardiac repair was performed at the age of 2 years. The post-operative course was good.

Case 3

A girl with transposition of great arteries with ventricular septal defect was born at 31 weeks' gestation weighing 1390 g. She was treated with patent ductus arteriosus clipping in the neonatal period and pulmonary artery banding at 3 months. Her respiratory status was unstable since then, and CT showed hyperinflation of the left upper lobe. Non-invasive positive pressure ventilation management was continued, but the lesion remained unchanged even after growth, and left upper lobectomy was performed at 11 months. She was weaned from non-invasive positive pressure ventilation management 3 weeks post-operatively. Arterial switch operation was performed, and she progressed well without difficulty in perioperative management.

Discussion

Concomitant congenital lobar emphysema and CHD require circulatory and respiratory management. As symptoms of congenital lobar emphysema are similar to those of heart failure, congenital lobar emphysema may be diagnosed after CHD treatment.[3] Respiratory or haemodynamic abnormalities that cannot be explained by CHD alone require the evaluation of the lung parenchyma, and CT is useful. In all cases, CT and bronchoscopy were performed; however, the cause of bronchial compression was not determined. Gentle non-invasive positive pressure ventilation management was an effective conservative treatment, but positive pressure airway may further aggravate pulmonary hyperinflation.[4,5] Lobectomy should be considered for cases of prolonged treatment.

We suggest lobectomy prior to intracardiac repair if congenital lobar emphysema does not improve and conservative treatment is unsuccessful despite pulmonary flow control by palliative surgery. Conversely, a study reported three infants with congenital lobar emphysema complicated by ventricular septal defect whose respiratory impairment improved after lobectomy and cardiac surgery was not required.[6] Dogan et al reported a case in which lobectomy was necessary because the patient could not be weaned from cardiopulmonary bypass after intracardiac repair.[1] Prior lobectomy provides safe respiratory management perioperatively of cardiac surgery and promotes the development of healthy lungs. If haemodynamic status improves after lobectomy, it may be possible to manage the patient without surgical intervention for heart disease.

There are reports of bilateral congenital lobar emphysema. Not all lesions require resection. A case study reported that three of four patients had good outcomes with unilateral lobectomy.[7] Our patients also had residual lesion but did not require additional surgery after more than a year. It should be noted that there is a risk that congenital lobar emphysema may affect the circulation as well as respiration, as in case 1. Even congenital lobar emphysema without CHD can lead to haemodynamic instability under conditions of tension physiology. We must be careful about circulation in pneumothorax and intrapulmonary occupational lesions such as congenital pulmonary airway malformation.

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Conclusions

All patients in this report achieved good outcomes by prior lobectomy. If conservative treatment is difficult, it is advisable to perform lobectomy before treating cardiac diseases.

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Conflicts of interest. None.

Ethical standards. This case report does not involve human and/or animal experimentation.

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