Giant Pulmonary Bulla Underlying Bronchopulmonary Dysplasia in a Very Low-birth-weight Infant: a Case Report

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Case report

Giant pulmonary bulla underlying bronchopulmonary dysplasia in a very low-birth-weight infant: A case report

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50 **Abstract**

51 **Background:** Acquired cystic lung disease is a serious respiratory complication of
52 bronchopulmonary dysplasia in premature infants. Most cases of acquired cystic lung disease
53 underlying bronchopulmonary dysplasia involve pulmonary interstitial emphysema. Although
54 this is a reversible condition, there are a few instances where surgery might be necessary. An
55 accurate diagnosis is important to decide the therapeutic strategy for acquired cystic lung
56 disease. Here, we report a rare case of a giant pulmonary bulla in an infant treated with
57 bullectomy.

58 **Case presentation:** A male infant born at 23 weeks of gestation with a birth weight of 524 g
59 was initially diagnosed with respiratory distress syndrome. During mechanical ventilatory
60 support, he presented with recurrent pneumothorax and a gradually expanding pulmonary cyst
61 in the right lung. Chest CT at 5 months of age revealed a large cyst located in the subpleural
62 area adjacent to the multiple cystic air spaces. These findings are consistent with the diagnosis
63 of giant pulmonary bulla with pulmonary interstitial emphysema underlying
64 bronchopulmonary dysplasia . At 9 months of age, the giant pulmonary bulla expanded
65 further due to acute bronchitis for which he developed respiratory failure and obstructive
66 shock. This warranted a bullectomy for the giant pulmonary bulla. After the operation, the
67 unresected pulmonary interstitial emphysema lesion did not expand further. He is currently
68 three years old and has no respiratory problems.

69 **Conclusions:** This case demonstrated that chest CT is useful for providing valuable
70 anatomical information necessary in deciding the treatment strategy for acquired cystic lung
71 disease in infants.

72

73 **Keywords:** Acquired cystic lung disease, Bronchopulmonary dysplasia, Giant pulmonary
74 bulla, Infant, Pulmonary interstitial emphysema

75

76 **Background**

77 Acquired cystic lung disease in premature infants is a serious respiratory
78 complication of bronchopulmonary dysplasia (BPD) [1]. This includes various pathologies
79 such as pulmonary interstitial emphysema (PIE) [2-4], pulmonary bulla [5], and pneumatocele
80 [6]. Among acquired cystic lung diseases, most cases are reported as PIE. However, bullous
81 lung diseases are rare in infants. Since the treatment strategies for these pathologic conditions
82 are different, it is important to establish a correct diagnosis. Here, we report a rare case of
83 giant pulmonary bulla in a very low-birth-weight infant and highlight the utility of computed
84 tomography (CT) in deciding the therapeutic strategy.

85

86 **Case presentation**

87 A male infant (525 g) with no congenital pulmonary cysts detected on foetal
88 echography was born at 23 weeks of gestation with low Apgar scores of 1, 3, and 6 at 1, 5,
89 and 10 min, respectively. After birth, the patient was intubated and received synchronised
90 intermittent mandatory ventilation. Initial chest radiographs at birth showed ground-glass
91 appearance and air bronchograms with no cystic lesion consistent with respiratory distress
92 syndrome. The patient was then treated with surfactant. On the 11th day, he developed
93 pneumothorax on the right side, which required the placement of a chest drain. On the 24th
94 day, the lung cyst appeared and gradually expanded in the right upper lobe. Because of the
95 recurrences of pneumothorax, the mechanical ventilator mode was changed to neurally
96 adjusted ventilatory assist or high-frequency oscillatory ventilation to reduce ventilator-

induced lung injury. On the 76th day, he was extubated and continued to receive non-invasive respiratory support as a consequence of BPD. However, the lung cysts gradually expanded.

Chest CT performed on the fifth month of age revealed two types of cystic lesions. The first was a huge solitary cyst with an empty interior and smooth outlines below the pleura. The second was multicystic lesions of various sizes with septations located adjacent to the first. Due to mass effects, there was a left mediastinal shift with left lung atelectasis (Fig. 1). These cysts were diagnosed as giant pulmonary bulla with PIE.

In the interim, his respiratory condition remained stable with oxygen therapy and his body weight gradually increased. He was then discharged from the hospital at 6 months of age. After that, we observed his clinical condition while planning to perform surgical intervention should his respiratory condition deteriorate.

At 9 months of age, he was admitted due to acute bronchitis caused by human metapneumovirus. A chest radiograph showed further expansion of the giant pulmonary bulla triggered by the respiratory infection (Fig. 2). His clinical condition could not be controlled by medical treatment such that he presented with respiratory failure and obstructive shock. While PIE is expected to spontaneously regress, the giant pulmonary bulla was thought to be the main cause of respiratory failure. This warranted cystectomy without resection of the PIE. The giant pulmonary bulla was incised, and a few air leakage sites were sutured directly. The cyst sac was empty without remnants of the lung tissues. The visceral pleura was then reinforced using a residual cyst wall. Histopathologic examination of the excised bulla wall revealed that it was derived from the fibrous tissue and was located within the subpleural lung parenchyma. The giant emphysematous cyst was histologically diagnosed as a bulla, excluding congenital cystic lung disease (Fig. 3).

After the operation, the patient had no respiratory problems and did not need any respiratory support. He is currently three years old and is doing well.

Discussion and conclusions

Bulla is defined radiologically as an emphysematous space with a diameter of 1 cm or more and is morphologically located in the subpleural lung parenchyma [7]. Reid et al. proposed a classification for bulla. Type I bulla is characterised pathologically as a narrow-necked empty sac with a clear linear outline in the absence of vascular or airway remnants [7]. The outer surface of the bulla is composed of visceral pleura, while its inner surface is composed of fibrous tissue, derived from the pleura and the destroyed pulmonary tissue [8]. In our case, the expanded cyst was a monocyst with a clear rim of the wall on CT. The cystic wall histologically contained fibrous tissue and was located within the subpleural lung parenchyma. The cyst was diagnosed as a Reid type I bulla based on both radiologic and histopathologic findings. The symptomatic expanded cyst met the following criteria of a giant pulmonary bulla: (1) upper lobe involvement, (2) occupying at least one-third of the hemithorax, and (3) compressing the surrounding lung parenchyma [9]. On the contrary, multiple cystic spaces were adjacent to the giant pulmonary bulla. These showed extra-alveolar air accumulation and a line-and-dot pattern, which is a specific sign of PIE on CT.

The mechanism of PIE is likely from air leakage into the perivascular and peribronchial spaces due to the high airway pressure of mechanical ventilation [10]. This PIE may spread centrifugally along the bronchovascular sheath or lymphatic channels. Air leakage develops into expanded airspaces of the terminal bronchiole and alveolar septal destruction, which leads to subpleural emphysema [10]. Therefore, the pathogenesis of bulla and the adjacent PIE, in this case, was considered to be closely related.

In the management of acquired cystic lung disease, PIE may be reversible. Conservative treatment by selective intubation, selective bronchial obstruction, or decubitus positioning is accepted as the initial management [3-5]. However, the giant pulmonary bulla

can be considered for surgery [2, 8, 11]. Based on the good outcome of our case, cystectomy was considered to be safe and effective as it can leave sufficient residual lung in small infants.

In our case, the chest radiograph could not sufficiently evaluate the structure of the acquired cystic lung disease. However, CT could be utilised to further evaluate the lesions at a higher resolution, which may provide important anatomic information necessary in choosing a therapeutic strategy.

In conclusion, most case reports of acquired cystic lung disease underlying BPD are PIE, and bulla is rarely involved in acquired cystic lung disease. To decide the therapeutic strategy, CT is useful when chest radiograph is equivocal on the evaluation of acquired cystic lung disease.

List of abbreviations

BPD, bronchopulmonary dysplasia; PIE, pulmonary interstitial emphysema; CT, computed tomography

Declarations

Ethics approval and consent to participate: Informed consent was obtained from patient.

Consent for publication: We obtained the consent for publication from the parent.

Availability of data and materials: Not applicable

Competing interests: The authors declare no conflict of interest.

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Authors' contributions: TS, YH, KK and NO interpreted the patient data regarding the lung disease. TO performed the histological examination of the bulla, and TS, KK were major contributor in writing the manuscript. All authors read and approved the final manuscript.

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Figure legends

Figure 1

Chest computed tomography taken at 5 months of age demonstrated a large 30×25 mm monocystic space with a clear rim (asterisk) and multiple cystic air spaces with a line-and-dot pattern (white arrowheads) in the upper right lobe (a, b).

Figure 2

A chest radiograph revealed a giant cyst with a diameter of 80×60 mm, causing left mediastinal left shift and left lung atelectasis.

Figure 3

Histopathologic analysis of the excised bulla wall (black arrows), visceral pleura (black arrowheads), and lung parenchyma. The wall of bulla was located below the visceral pleura adjacent to the lung parenchyma and was formed by fibrous tissue (Elastic Van Gieson staining, $\times 200$).

Figures

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