

Progressive Intraparenchymal Bronchogenic Cyst in a Neonate

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Neonatal parenchymal bronchogenic cysts (BC) are a very rare congenital anomaly. Usually patients with BC are born with severe respiratory distress or cardiovascular insufficiency are asymptomatic till they grow up to older children and adults. We report a case of neonatal BC with a prenatal diagnosis of congenital tracheobronchial cystic anomalies of the right lung. The cyst was 36 mm in diameter at 17 week gestational period but diminished in size to 21 mm at 35 weeks. After birth, chest X-ray demonstrated a growing cyst 50 mm in diameter and gradual displacement of the heart and mediastinum from the right to the left day by day. Right S³ segmentectomy was performed on the 5th day. (Ann Thorac Cardiovasc Surg 2008; 14: 32–34)

Key words: intraparenchymal bronchogenic cyst, neonate, segmentectomy, mediastinal shift

Introduction

There is a wide variety of developmental abnormalities of the respiratory tract and cystic lung disease (CLD) in children, such as congenital pulmonary airway malformations (CPAM), intralobar sequestrations (ILS), bronchial atresia and bronchogenic cysts (BC), which is a relatively rare congenital abnormality. These anomalies are often difficult to diagnose pathologically and coexist because these diseases have similar embryologic origin.

BC are very rare in the neonatal period, their overall prevalence is reported to be about 0.05%,¹ Ten percent of cases of BC are recognized at birth.² Approximately two third are within the mediastinum and one third are intraparenchymal.^{3,4} Up to one third of BC patients remain asymptomatic until they mature. Because many cysts occupy only a small space in a term neonate, it has been noted that BC cause symptoms in older children and

adults. We report here an unique case of neonatal BC with antenatal diagnosis of CLD with the cyst gradually progressively enlarging after birth in spite of the avoidance of positive pressure breathing.

Case Report

A 1-day-old girl was admitted to the neonatal intensive care unit of our hospital with the diagnosis of CPAM. She was born spontaneously after a 39-week-and-3-day gestation, weighting 2,900 g, to a 29-year-old multigravida mother. She received Apgar Scores of 8 and 9 at 1 and 5 min, respectively. When the fetus was at 17 weeks gestational period, a cystic mass of left hemithorax was noted by a sonogram. BC or type-I CPAM was suspected. The cyst was 36 mm in diameter but diminished in size to 21 mm at 35 weeks' gestational period. Arterial blood gas analysis showed hypoxemic and respiratory acidosis with hypercapnia (pH 7.167, pCO₂ 66.4 Torr, HCO₃ 23.1 mmol/L, BE -8.0, SatO₂ 85%) but oxygen saturation increased to be above 95% and pCO₂ went down to about 45 Torr on 30% oxygen in a closed incubator. Manual and mechanical ventilation were avoided because positive pressure breathing could lead to enlargement and rupture of the cyst. Initial immediate chest X-rays demonstrated a

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well defined cystic lesion in the right upper field. Computed tomographic scan (CT) of the chest showed a 35×20 mm air containing cyst as a round low density area in the right S^3 . Chest X-ray demonstrated gradual displacement of the heart and mediastinum from the right to the left day by day (Fig. 1, A and B).

At 5 days of age, the patient underwent thoracotomy assisted with thoracoscopy through right IVth intercostals space. The right S^3 segment was cystic and independently lobulated from S^{1+2} and lingular segment. At first, the tension wall of the cyst was incised to release the contained g air. The pulmonary vein V3 was identified, ligated with double 4-0 Prolene and transected. The anterior trunk of right pulmonary artery was exposed, and the A3 was prepared and transected in the same way. B3 bronchus which was found under the artery was ligated with 4-0 Vicryl and transected. The cyst communicated with B³. Next, S^3 segment separation was performed with an electric cauterly with low output. The postoperative course was uneventful and the patient was discharged on the 12th postoperative day.

A histopathological examination revealed a S^3 segment sized $51 \times 35 \times 15$ mm, with thin-walled, only air filled unilocular cyst without mucoid material contents (Fig. 2). Microscopically, the cyst was lined with ciliated, mucus secreting respiratory columnar epithelium. The relatively central bronchus connected to the cyst was cystic dilated but peripheral bronchi were not dilated. There was no increase in terminal bronchiolar tissue and absence of cartilage and increased elastic tissue surrounding the cyst.

Discussion

BC were first described in 1948 by Maier.⁵⁾ It has been noted that these cysts generally occur in adults or older children. Gerle first proposed the term “broncho-pulmonary foregut malformation” to encompass these pulmonary congenital abnormalities in 1968.⁶⁾ BC are the most common primary cysts of the mediastinum and usually demonstrate no patent communication to the airway and are adherent to major airways or esophagus. Most frequently they are unilocular, round or oval in shape and contain clear fluid, hemorrhagic secretions or air are less common.

It is sometimes difficult to pathologically diagnose neonatal cystic lesions and inflamed intraparenchymal cysts may be particularly impossible to definitively diagnose. T Parenchymal cysts without pathological definitive and typical diagnosis have recently been treated “in-

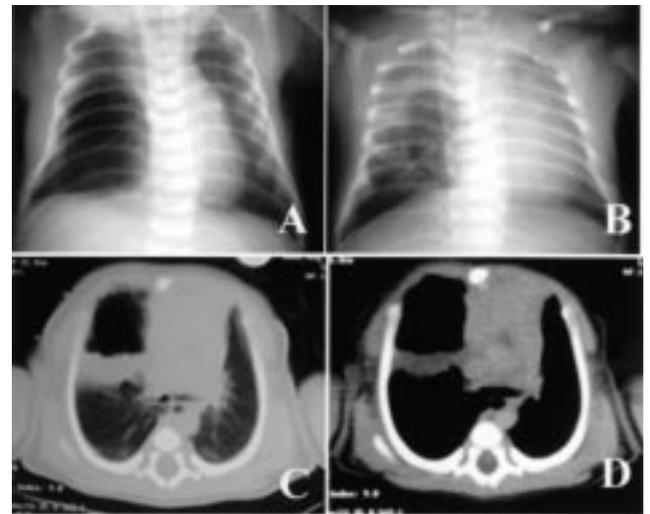


Fig. 1. Chest X-ray demonstrated a hyperexpanded right lung with a gradual displacement of the heart and mediastinum from the right to the left day by day (A: Day 2 after birth; B: Day 4 after birth).

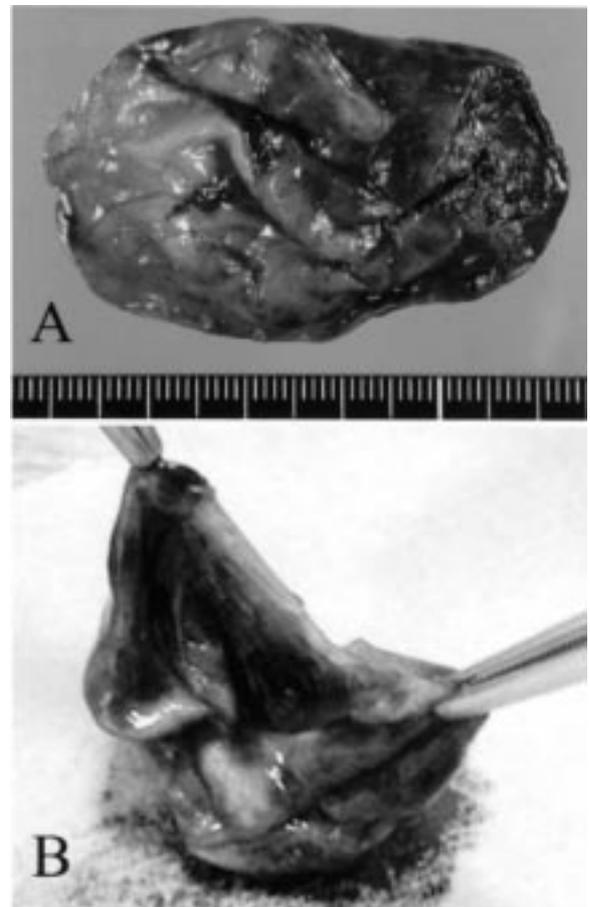


Fig. 2. Gross section of the excised bronchogenic cyst occupying right S^3 segment (A: Resected segment; B: The content was a cyst cavity without mucoid material).

flamed intraparenchymal cysts (IIC).” BC typically has cartilaginous structures and submucosal glands in its wall, and are the most common primary cysts of the mediastinum or are adherent to major airways or the esophagus. The present case was pulmonary parenchymal type and as it did not have cartilaginous structures in its wall, the diagnosis of IIC may be plausible. However as in this case there was a lack of alveolar tissue, the distinction from CPAM is apparent.

BC are often discovered on prenatal sonography such as CPAM and ILS, and have check valves that lead to their progressive enlargement. Enlarged cysts cause a mediastinal shift or pneumothorax due to rupture into pleura.^{7,8)} If they produce a mediastinal shift in utero, aspiration of the cyst may be indicated. The present case did not have mediastinal shift due to cyst expansion in utero, but had gradually expanded shifted after birth. Such enlargement usually occurs in neonates and young infants and many complications were reported such as superior vena cava syndrome, pericardial or atrial compression, bronchial atresia and pulmonary artery stenosis by mediastinal type BC.

Most BC patients without antenatal diagnosis are recognized by their symptoms of severe respiratory distress or cardiovascular insufficiency at birth positive pressure breathing with manual or mechanical ventilator may be carried out because of their low respiratory function. However, this treatment can lead to enlargement and rupture of the cyst and possible fatality.

Surgical therapy is effective as a treatment of BC and should be performed as early as possible for postnatal patients because of the risk of pulmonary compression, infection, or malignant degeneration.⁹⁾ Ideal surgical procedure is a paraenchyma-saving operation and should be selected whenever possible. However, the most important surgical point is complete excision of BC tissues around the parenchyma and should be resected if the di-

viding line is unclear because recurrent cases have been reported following incomplete excision and multiple attempts at percutaneous aspiration,^{10,11)} Lobectomy or segmentectomy is the procedure of choice.

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