Pediatr Neonatol 2008;49(5):197–200

ELSEVIER

CASE REPORT

Spontaneous Multiseptated Cystic Pneumomediastinum in a Term Newborn

Cheng-Ting Lee[1], Po-Nien Tsao[1], Steven Shinn-Forng Peng[2], Suh-Fang Jeng[3], Hung-Chieh Chou[1], Chien-Yi Chen[1], Wu-Shiun Hsieh[1]*

[1]*Department of Pediatrics, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan*
[2]*Department of Medical Imaging, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan*
[3]*School and Graduate Institute of Physical Therapy, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan*

Received: May 15, 2008
Revised: Jun 16, 2008
Accepted: Sep 19, 2008

KEY WORDS:
congenital cystic lung disease;
pneumomediastinum;
pulmonary air leak

Pneumomediastinum usually occurs after assisted ventilation. Spontaneous pneumomediastinum without preceding assisted ventilation, and presenting as a huge cystic mass in a term newborn infant has rarely been reported. We report a term baby who developed respiratory distress at 2 days of age. A huge cystic tumor of the thymus or a congenital mediastinal cystic tumor was initially suggested by chest radiography and computed tomography. Follow-up chest film revealed a "spinnaker sail sign" which is a typical radiographic presentation of pneumomediastinum.

1. Introduction

Localized air collection in the mediastinum of a newborn can be caused by a congenital mediastinal cystic mass[1,2] or by pneumomediastinum resulting from pulmonary air leak.[3] The mediastinal cyst may have various origins; it can have bronchogenic, foregut, mesothelial-derived, neuroenteric, or thymic cystic origins, or can be due to other rare causes.[1,2] Most cases of pulmonary air leak occur in newborns with underlying lung disease, especially if mechanical ventilation is required. Preterm infants are at increased risk because they frequently suffer from respiratory distress syndrome, although prophylactic surfactant replacement treatment may reduce the incidence of air leak.[4] It is challenging for physicians to deal with mediastinal free air collection in a term neonate with no obvious lung disease. Here, we report a term neonate with no perinatal history of lung disease, in whom spontaneous multiseptated cystic pneumomediastinum occurred 2 days after birth. Identification of the nature of the mediastinal lesion is crucial, because surgical intervention may be indicated if a congenital mediastinal cyst is diagnosed.

2. Case Report

A female baby was born at 37 weeks' gestation to a G2P2 mother via an uncomplicated cesarean section

*Corresponding author. Department of Pediatrics, National Taiwan University Hospital and National Taiwan University College of Medicine, 7 Chung-Shan S. Road, Taipei 100, Taiwan.
E-mail: hsiehws@ntu.edu.tw

at an obstetric clinic. Her body weight was 3000 g. The Apgar scores were 8 at 1 minute and 9 at 5 minutes. Prenatal history was remarkable only for maternal asthma under regular medical control. Poor feeding without abdominal distension was noted after birth. At 2 days old, progressive respiratory distress with subcostal retractions and desaturation developed. Desaturation improved under oxygen supplementation but dyspnea persisted, and the baby was therefore transferred to our neonatal intensive care unit for further management.

Initial complete blood cell counts and serum biochemistry profile including C-reactive protein were within normal limits. Initial arterial gas analysis was normal with no evidence of hypoxemia, hypercapnia or metabolic acidosis. Chest X-ray showed a huge radiolucent mediastinal cystic lesion without lung markings over the anterior mediastinum (Figure 1). No obvious thymic shadow was apparent on initial chest radiography. A transillumination test of the chest was negative for pneumothorax. Empirical intravenous antibiotics with ampicillin and gentamicin were administered for fear of respiratory distress due to pneumonia or sepsis. Nasal continuous positive airway pressure support was also given. Chest computed tomography (CT) was performed at 3 days old, to determine the origin of the cyst. A huge multiseptated cystic tumor was detected in the mediastinum, with the thymus compressed to the cephalic aspect (Figure 2). No definite air communication between the cystic lesion and the airway or esophagus was observed. Congenital upper mediastinal cystic tumor, a cystic lesion within the thymus tissue or pneumomediastinum were possible diagnoses. There was no evidence of subcutaneous emphysema. There was no further deterioration in the patient's condition, and she was therefore monitored by close observation. A follow-up chest radiograph at 4 days old showed the area of air lucency to have decreased while the thymus shadow over the upper chest had become more prominent. Feeding and respiration became increasingly stable, and the nasal continuous positive airway pressure support was therefore changed to a nasal cannula with oxygen support at 4 days old. Oxygen support was discontinued at 6 days old.

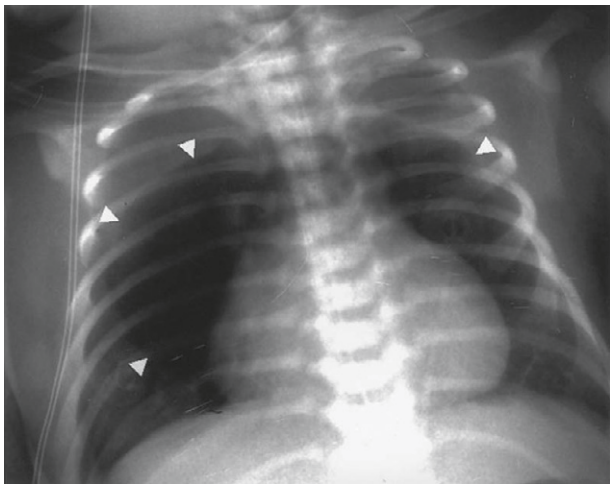

Figure 1 At 2 days of age, anteroposterior chest roentgenogram revealed a wide radiolucent area over the mediastinum (arrowhead).

A chest radiograph taken at 1 week old revealed a "spinnaker sail sign", which is the classic radiographic feature of pneumomediastinum (Figure 3). Pneumomediastinum was accordingly diagnosed and antibiotics were discontinued after completion of the treatment course. The last chest radiograph taken before discharge at 8 days old showed total resolution of the area of air collection. The patient was discharged at 12 days old. There was no evidence of recurrence of pneumomediastinum on a chest radiograph taken 1 week after discharge. Her condition remained stable.

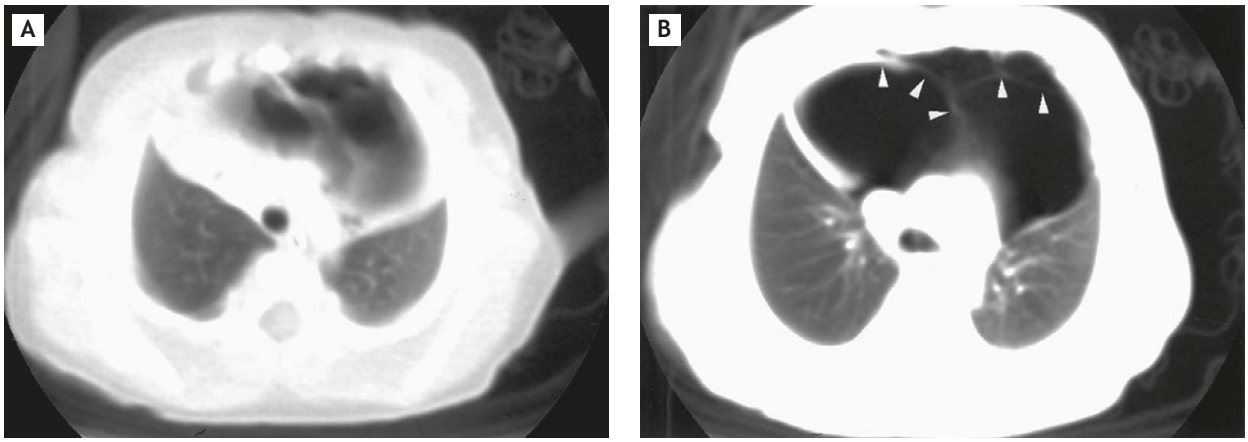

Figure 2 Postcontrast chest computed tomography at 3 days old. The upper portion (A) and the lower portion (B) of the cyst. Note the thin septum within the cyst (arrowhead).

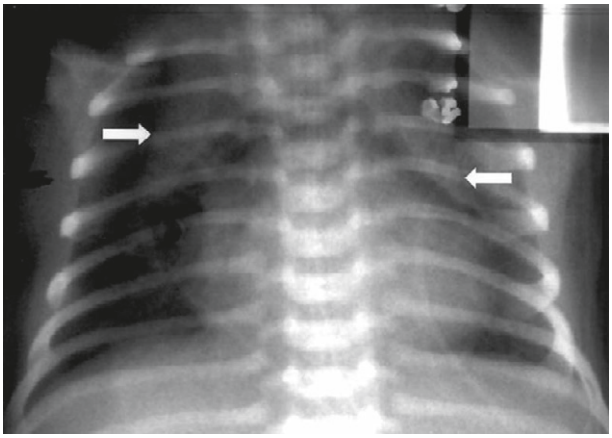

Figure 3 At 1 week old, a typical "spinnaker sail sign" (arrow) showed on supine anteroposterior chest roentgenogram.

3. Discussion

Pneumomediastinum, or mediastinal emphysema, is defined as the presence of gas in the mediastinum. The reported incidence of pneumomediastinum is 4–25 per 10,000 live births.[3,5,6] The incidence may be underestimated because many infants with pneumomediastinum have minimal symptoms.[3] The pathogenesis of pneumomediastinum was first demonstrated experimentally by Macklin:[7] increased intraalveolar pressure leads to alveolar rupture into the perivascular space. Air then dissects along the vascular channels resulting in pneumomediastinum as it leaks into the mediastinum. Most reported cases of neonatal pneumomediastinum have therefore been related to underlying lung disease or following active resuscitation at birth.[4,5,8,9] However, one study reported that most patients may develop neonatal pneumomediastinum in the absence of any known predisposing factors,[3] as in our patient. One possible explanation for the spontaneous development of pneumomediastinum may be the forceful inspiratory effort at birth coupled with some degree of ventilation inhomogeneity, resulting in alveolar overdistention and rupture.[10]

A complete roentgenographic examination with anteroposterior and lateral views is necessary to establish the diagnosis of pneumomediastinum. The role of the transillumination test is to detect sudden, life-threatening pneumothorax, which requires immediate therapy.[11] The commonest diagnostic radiographic sign of pneumomediastinum is the "spinnaker sail sign".[12] On an anteroposterior view, a crescentic configuration of thymic lobes resembles a spinnaker as it is lifted by the buoyant air and separated from other structures of the mediastinum. The initial chest X-ray in our patient showed only a huge multiseptated mediastinal cystic lesion without lung markings and no typical roentgenographic sign of pneumomediastinum. The differential diagnoses based on the imaging findings in our patient included congenital mediastinal cyst, subpulmonary bleb, subpulmonary pneumothorax, large hiatal hernia and pneumopericardium. CT is an important noninvasive modality for the evaluation of mediastinal changes. Siegel et al showed that CT provided diagnostic information additional to that derived from chest radiographs in 82% of pediatric patients with mediastinal abnormalities revealed by traditional roentgenogram, and contributed to substantial changes in therapy in 65% of the patients.[13] The CT image in our patient excluded the possibility of mediastinal anomaly and avoided unnecessary surgical intervention.

Marchand described an investing fascia of the great vessels and trachea, which was continuous with the fibrous pericardium and with the investing fascia of the pulmonary vessels at the lung hilus.[14] The investing perivisceral fascia "constitutes a highway connecting the lung to the mediastinum". He demonstrated this continuity by the injection of colored radiopaque fluid in cadavers. Quattromani et al[15] later observed thymic fascia during autopsies on patients, and suggested that this fascia is continuous with the perivisceral fascia as described by Marchand.[14] The fascia forms the thymic capsule and interlobular septa of the thymus. If the air dissects within this fascia and gains access to the potential space behind the thymus, a specific radiographic appearance of multiseptated pneumomediastinum will be seen.

The imaging findings in our patient support the possibility that neonatal pneumomediastinum, unlike that in older children and adults, tends to loculate locally and has multiple internal septa, because neonates have a strong thymus and fascia to contain the air in the mediastinum and so prevent further dissection of air into the subcutaneous tissue of the neck.[16] This fascia and thymus atrophy with age, and subcutaneous emphysema is thus more frequent in older patients with pneumomediastinum.

In conclusion, pneumomediastinum in newborns with no obvious predisposing lung disease may be initially hard to diagnose. A large area of air collection over the anterior mediastinum may simulate a congenital mediastinal cyst. "Bubbly" radiolucency in the chest should be treated conservatively, unless severe respiratory decompensation occurs.

References

1. Ochsner JL, Ochsner SF. Congenital cysts of the mediastinum: 20-year experience with 42 cases. *Ann Surg* 1966;163: 909–20.

2. Takeda S, Miyoshi S, Minami M, Ohta M, Masaoka A, Matsuda H. Clinical spectrum of mediastinal cysts. *Chest* 2003;124: 125–32.
3. Morrow G 3rd, Hope JW, Boggs TR Jr. Pneumomediastinum, a silent lesion in the newborn. *J Pediatr* 1967;70:554–60.
4. Hansen TN, Corbet A, Gest AL, et al. Principles of respiratory monitoring and therapy. In: Taeusch HW, Ballard RA, Gleason CA, eds. *Avery's Diseases of the Newborn*. Philadelphia: WB Saunders, 2005:648–69.
5. Lubchenco LO. Recognition of spontaneous pneumothorax in premature infants. *Pediatrics* 1959;24:996–1004.
6. Chasler CN. Pneumothorax and pneumomediastinum in the newborn. *Am J Roentgenol Radium Ther Nucl Med* 1964;91: 550–9.
7. Macklin C. Transport of air along sheaths of pulmonic blood vessels from alveoli to mediastinum. *Arch Int Med* 1939;64: 913–26.
8. Steele RW, Metz JR, Bass JW, DuBois JJ. Pneumothorax and pneumomediastinum in the newborn. *Radiology* 1971;98: 629–32.
9. O'Gorman LD, Cottingham RA, Sargent EN, et al. Mediastinal emphysema in the newborn: a review and description of the new extrapleural gas sign. *Dis Chest* 1968;53: 301–8.
10. Chernick V, Avery ME. Spontaneous alveolar rupture at birth. *Pediatrics* 1963;32:816–24.
11. Kuhns LR, Bednarek FJ, Wyman ML, et al. Diagnosis of pneumothorax or pneumomediastinum in the neonate by transillumination. *Pediatrics* 1975;56:355–60.
12. Moseley JE. Loculated pneumomediastinum in the newborn: a thymic "spinnaker sail" sign. *Radiology* 1960;75:788–90.
13. Siegel MJ, Sagel SS, Reed K. The value of computed tomography in the diagnosis and management of pediatric mediastinal abnormalities. *Radiology* 1982;142:149–55.
14. Marchand P. The anatomy and applied anatomy of mediastinal fascia. *Thorax* 1951;6:359–68.
15. Quattromani FL, Foley LC, Bowen A, et al. Fascial relationship of the thymus: radiologic-pathologic correlation in neonatal pneumomediastinum. *Am J Roentgenol* 1981;137: 1209–11.
16. Low AS, Tan-Kendrick AP, Loh M, et al. Spontaneous multiseptated pneumomediastinum in a newborn baby: the spinnaker sail is rigged–CT features with pathologic correlation. *Pediatr Radiol* 2003;33:712–5.