CASE REPORT

Spontaneous Multiseptated Cystic Pneumomediastinum in a Newborn by an Uncomplicated Delivery
Yu-Tsun Su¹, Ming-Lun Yeh², Yuan-Yi Huang¹, Yung-Cheng Lin¹, Yu-Cheng Tsai¹, Chen-Kuang Niu³, Chien-Yi Wu⁴
¹Division of Pediatric Pulmonology, Department of Pediatrics, E-Da Hospital/I-Shou University, Kaohsiung City, Taiwan
²Division of Pediatric Surgery, Department of Surgery, E-Da Hospital/I-Shou University, Kaohsiung City, Taiwan
³Division of Pediatric Pulmonology, Department of Pediatrics, Chang-Gung Memorial Hospital, Kaohsiung City, Taiwan
⁴Division of Neonatology, Department of Pediatrics, E-Da Hospital/I-Shou University, Kaohsiung City, Taiwan

Abstract

Pneumomediastinum usually occurs in newborns as a result of an eventful delivery or associated respiratory diseases. Spinnaker sail sign is the typical finding on radiography. We report a term newborn delivered by an uneventful vaginal delivery who presented with a multiseptated cystic lesion on computed tomography. Due to respiratory distress and an undetermined pathology of a mediastinal mass, surgical intervention was initially scheduled. Finally, the patient was diagnosed with spontaneous multiseptated cystic pneumomediastinum by the disease's limited course and typical spinnaker-sail sign in follow-up chest radiography. Neonatal pneumomediastinum may present as a “bubbly” radiolucency in chest radiography initially, and may display multiseptated cystic lesions in computed tomography in addition to typical spinnaker-sail sign in chest X-ray. (J Pediatr Resp Dis 2014;10:13-16)

Key words: pneumomediastinum, spinnaker-sail sign, multiseptated cystic lesion

INTRODUCTION

Air collection in the mediastinum in a newborn results from pneumomediastinum (PM) and congenital mediastinal cystic lesions.¹ PM is frequently associated with some type of insult or trauma at birth, including prematurity, difficult delivery, pneumonia, meconium aspiration syndrome, or mechanical ventilation.²,³ It presents as radiolucency features in chest X-ray and the treatment strategy depends on the nature of the lesions. In addition to mediastinal diseases, intrapulmonary lesions, including congenital cystic adenomatoid malformation (CCAM), and congenital lobar emphysema (CLE) have radiolucency or emphysematous image in radiographic finding.² Herein, we report on the finding from a term baby was born by uneventful vaginal delivery, but presented with respiratory distress after birth. Bubbly radiolucency in the initial chest radiography, and multiseptated cystic lesions in high resolution computed tomography (HRCT) were observed. Surgical intervention was initially scheduled but then was suspended. The patient was finally diagnosed with spontaneous neonatal multiseptated cystic pneumomediastinum because of the disease’s limited course and spinnaker sail sign in follow-up chest radiography.⁴-⁷

CASE REPORT

This full-term neonate was born by uneventful vaginal delivery at gestational age of 37 weeks and 4 days, to a G2P2 mother. His birth body weight was 2910 gm, and the child’s Apgar scores were 8 at 1 minute and 9 at 5 minutes after birth. However, respiratory distress, subcostal retraction, and nasal flaring developed after
birth. The infant was transferred to neonatal intensive care unit and the chest radiography showed ground-glass density over both upper lungs and superior mediastinum, and relatively radiolucency over the upper lungs (bubbly radiolucency) (Fig. 1). Flexible bronchoscopy was performed, which revealed a blind-end tracheal bronchus, without other trachea-bronchial airway anomalies. The HRCT revealed several giant bullae or air-cysts formation with internal septation over bilateral anterior lung zones (crossing the midline) (Fig. 2 and Fig. 3). The differential diagnoses included severe focal /lobar emphysema (CLE) with giant bullae formation, or CCAM type 1, or spontaneous PM (less likely). Blind-end outpouching over the right posteriolateral wall of distal trachea, blind-end tracheal bronchus (tracheal diverticulum) was suspected, and a prominent soft tissue lesion was discovered over the superior mediastinum. Because of a deterioration of the patient’s respiratory condition, we changed the ventilator mode from nasal continuous positive airway pressure to nasal intermittent mandatory ventilation. The surgical intervention was scheduled 5 days thereafter. Unexpectedly, the respiratory condition improved and the follow-up chest radiograph revealed regressive change of the bubbly radiolucency, and was compatible with spinnaker sail (Fig. 4) on day 9. Spontaneous multiseptated cystic PM was the final diagnosis and spontaneous resolution by nature. The infant was discharged on day 12 with a diagnosis of neonatal PM.

Figure 1. CXR showed a patchy density over the RUL, bilateral increased infiltration, and radiolucency over the superior mediastinum (bubbly lesions).

Figure 2. The pre-contrast CT revealed upper portion of multiseptated cyst over the anterior region of both lungs.

Figure 3. The post-contrast CT revealed lower multiseptated cysts over the anterior region of both lungs.

Figure 4. Follow-up CXR revealed spinnaker-sail signs.
DISCUSSION

To our knowledge, there are only few case reports about spontaneous PM in a term neonate born by a smooth delivery.\textsuperscript{4,5,8-10} The initial CXR revealed bubbly radiolucency and the CT reported multiseptated cystic lesions. Spontaneous neonatal PM was finally diagnosed using the typical spinnaker sail sign on the follow-up CXR.

PM is defined by air collection in the mediastinum. It results from increased intraalveolar pressure, which leads to a pressure gradient between the alveolar and perivascular space. This excessive pressure gradient causes alveolar rupture of air, along the perivascular space, and into the mediastinum, which causes PM.\textsuperscript{1,4} If the air extends into pleural cavity, pneumothorax occurs. PM in neonates is divided into a spontaneous form or one associated with underlying lung disease, including difficult delivery, pneumonia, meconium aspiration syndrome, or mechanical ventilation, prematurity with respiratory distress syndrome.\textsuperscript{2,4} The incidence in symptomatic infants is reported as 25 per 10,000 infants with live births. The incidence is probably underestimated since spontaneous PM infants are usually asymptomatic or show minimal respiratory symptoms.\textsuperscript{4} Spontaneous PM is very rare in an uneventful delivery. The most likely mechanism is the forceful inspiration effort at birth in the ventilation nonhomogeneous lung, resulting from an excessive pressure gradient and subsequent spontaneous PM.\textsuperscript{11}

The definitive diagnosis of neonatal PM depends on the typical “spinnaker sail” sign on chest anteroposterior X-ray, which was first described by Moseley.\textsuperscript{6} The air collection may extend to one side or both sides in anterior part of the thoracic cavity, and elevate the thymus to create the crescentic shadow like a “spinnaker sail” or “angel wing” sign.\textsuperscript{8-10} In some cases, “bubbly” radiolucency lesions may be the initial radiographic features in neonatal PM.\textsuperscript{5} When the air extends into the interlobular area within the thymus capsule, the multiple internal septa result to the appearance of multiple septated cystic lesions in the mediastinum, and multiloculated multiseptated cystic appearance in CT findings.\textsuperscript{4,5,7}

During the neonatal period, the initial presentations of respiratory distress and radiograph in PM are similar to other mediastinal mass or lung diseases.\textsuperscript{2,3} The differential diagnose of radiolucency cystic like lesions included neonatal PM, congenital mediastinal cyst, and intrapulmonary lesions, including CCAM, and lobar emphysema.\textsuperscript{2,4} Further detailed information about the real position and components of the radiolucency lesions can be available by the CT examination.\textsuperscript{12} The neonatal PM and the other diseases are similar in CT findings, however, the clinical course may be different. PM usually resolves with conservative therapy. Mediastinal cystic lesions including bronchogenic cysts, thymic cysts, etc., usually persist and need surgical intervention. In addition to mediastinal diseases, intrapulmonary lesions, including congenital cystic adenomatoid malformation (CCAM), and congenital lobar emphysema (CLE) have radiolucency or an emphysematous image on radiographic findings and require surgical intervention in cases of severe disease.\textsuperscript{2,3,5,12} Therefore, multiseptated cystic lesions can be found in many pathologic conditions, and we must take the clinic condition into consideration when making the diagnosis and the treatment strategy. Spontaneous multiseptated cystic pneumomediastinum should be considered in differential diagnosis of a term neonate born by an uncomplicated delivery.

In the present case, the chest X-ray revealed bubbly radiolucency lesions. Due to progressive respiratory distress, HRCT was performed, which showed multiple septated cystic lesions in anterior thoracic cavity. The differential diagnoses associated with radiolucency cystic like lesions included neonatal PM, congenital mediastinal cyst, and intrapulmonary lesions, including CCAM, and lobar emphysema. Surgical intervention was performed to explore the pathology and relieve the respiratory distress. The final diagnosis of spontaneous PM was made depending on the limited course of the disease, and the typical spinnaker sail sign in follow-up chest radiography. Multiple septated cystic PM must be considered in infants with bubbly radiolucency on radiography.

In conclusion, spontaneous neonatal pneumomediastinum in a term baby with uncomplicated delivery is scarce. However, this condition needs to be considered in infants with “bubbly” radiolucency in chest radiography, multiseptated cystic lesions on CT, in addition to classical spinnaker-sail sign in chest X-ray.
REFERENCES


