Case Report

Pulmonary interstitial emphysema and continuous positive airway pressure in a premature infant

Sameer Y. Al-Abdi, MRCPCH, *Nalini Singhal,* FRCP(C).

ABSTRACT

Pulmonary interstitial emphysema (PIE) is a recognized sequel of high pressure mechanical ventilation. Nevertheless, with the improvement in perinatal medical care, PIE started to be seen in spontaneously breathing infants. We present a 29-week-old girl who developed PIE on the first day of life, while she was on nasal continuous positive airway pressure, and was successfully managed with high frequency oscillatory ventilation. There is a paucity of these cases in the literature. We report on the occurrence of this entity, early reorganization, and the possible associated or causation factors.

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Four types of air leak syndrome have been described in neonates. Three of these (pneumothorax, pneumomediastinum, and pneumopericardium) are known to occur spontaneously or with augmented breathing. Spontaneous pulmonary interstitial emphysema (PIE) is rarely reported.[1-7] The incidence of PIE in ventilated very low birth weight (VLBW) infants is 32%. Although PIE is diagnosed frequently, there is no standard treatment. The treatment options are conservative or surgical. The conservative approaches are chest positioning, selective main stem bronchial intubation and occlusion, and changing mode of ventilation (low pressure ventilation or high frequency ventilation). Pleurotomy, lung puncture, pneumonectomy, and lobectomy are the surgical options for the patients with severe PIE who have not responded to maximum conservative intervention.[5-7] Our patient was a premature baby with unusual continuous positive airway pressure (CPAP) complication of early PIE. In this report, we would like to highlight the possibility of pregnancy induced hypertension (PIH)/hemolysis, elevated liver enzymes, and low platelet count syndrome (HELLP) as risk factors for PIE, and prophylactic surfactant treatment ameliorating the risk of developing PIE.

Case Report. A 29-week (by dates and early antenatal ultrasound at 11 and 18 weeks) baby girl, delivered in a tertiary care hospital by emergency cesarean section to a 32-year-old Caucasian primigravida mother with PIH and HELLP. The pregnancy was uneventful until the mother presented to the obstetric service complaining of right upper quadrant abdominal pain, and was found to have high blood pressure, abnormally elevated liver function tests and proteinuria. Labetalol and magnesium sulfate were started to control her blood pressure. She received a complete course of betamethasone. She was monitored by daily ultrasound. Emergency cesarean section was performed under epidural anesthesia for poor fetal tracing and breech presentation. The baby had poor respiratory effort at

From the Department of Pediatrics, University of Calgary, Calgary, Alberta, *Canada*.

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Address correspondence and reprint request to: Dr. Nalini Singhal, Director of NICU and Professor, Department of Pediatrics, University of Calgary, Calgary, Alberta, *Canada*. Tel. +1 (403) 9444638. Fax. +1 (403) 9444892. E-mail: nalini.singhal@calgaryhealthregion.ca

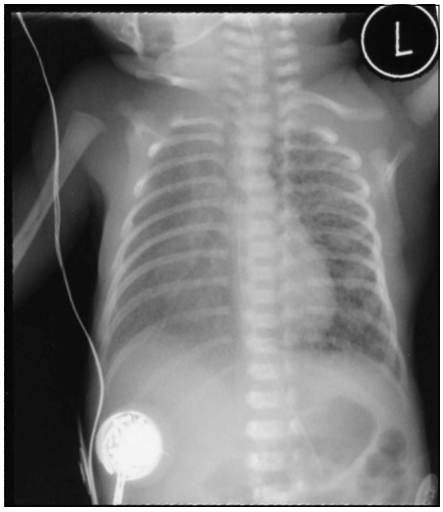

Figure 1 - Chest x-ray showing pulmonary interstitial emphysema in the left lung.

birth necessitating intermittent positive pressure ventilation with flow inflating face mask bagging for less than a minute. Apgars were 5 and 9 at one and 5 minutes, and the cord pH was 7.26 with base deficit of -7 mmol/L). Anthropometric measurements were appropriate for her gestational age: birth weight 1100 grams, head circumference 26 cm and length 37.5 cm. Detailed physical examination revealed a normal premature baby with mild respiratory distress, chest recession, flaring of alae nasi and the saturation by the pulse oximetry were reading 87-94% on nasal prong at rate of 1.5 LPM (liter per minute) of 35% blended oxygen. At 2 hours of age, she was apneic and was started on variable flow-active nasal CPAP (generator) at 5 cm of water. Capillary blood gases were: pH 7.31, partial pressure of carbon dioxide (PCO_2) 49 mm Hg, partial pressure of oxygen (PO_2) 58 mm Hg, bicarbonate (HCO_3) 24 mmol/L and base deficit -3 mmol/L. Magnesium was 2.30 mmol/L (normal 0.65-1.05 mmol/L). Chest radiography was carried out at 3 hours of age and was consistent with mild hyaline membrane disease. The 2nd chest x-ray was done at 22 hours of age for increasing oxygen requirement, and this showed PIE in the left lung (**Figure 1**). On the second day of life, the oxygen requirement and work of breathing increased so the baby was intubated and the first dose of bovine lipid extract surfactant (BLES) was given. Synchronized intermittent ventilation (SIMV) was used in the first 24 hours after intubation. On the third day of life, high frequency oscillatory ventilation (HFOV) was started due to worsening PIE and the second dose of BLES was instilled. For the next 2 days, there was no improvement of PIE on HFOV. The mean airway pressure was decreased to 5.5 cm of water, with adequate blood gases. Both lungs were partially collapsed for 2 days. On the 7th day of life, the lungs revived and PIE resolved. During HFOV the pH was 7.29-7.37, PCO_2 34-58 mm Hg, and PO_2 52-67 mm Hg. The baby was kept on HFOV for 4 days, and on SIMV mode for 1.5 days, and then extubated to nasal canula, and a few days later was discharged to level 2 unit in stable condition and normal follow up chest x-ray. During the HFOV period, the baby was on morphine infusion and received pancuronium for 2 days. Antibiotics were given until sepsis was ruled out by negative cultures, she had normal cranial ultrasound at one week of age.

Discussion. Pulmonary interstitial emphysema is a condition in which the trapped air seeps out through alveolar or terminal airways, and is dispersed into the pulmonary interstitium. The diagnosis of PIE is usually made by the characteristic finding of radiolucent areas on a plain chest radiograph. It can also be diagnosed by computerized tomography (CT) which is characterized by hyperradiolucent cystic structure with line and dot pattern, however, this is rarely carried out. Pulmonary interstitial emphysema has been classified into acute or persistent; a pathological classification depending on the presence or absence of giant cells. It is also described as localized, where there are large cysts up to 3 cm in diameter in one or 2 lobes of the lung, or generalized where there are diffuse small cysts less that 0.3 cm in diameter in all the lobes of the lung.

Pulmonary interstitial emphysema generally develops following positive pressure ventilation in a premature baby, the more premature the baby higher the risk of developing PIE. A literature review revealed that 5 premature babies developed PIE in the absence of positive pressure ventilation.[1-5] These 5 babies had a birth weight of 800-2640 gms, their gestational age was 28-35 week, 4/5 were female, 3 were delivered by emergency cesarian section, carried out for HELLP syndrome. Left lung was effected in 4/5 babies. Diagnosis of PIE was made at 2-18 days following birth. Our patient was female, her mother had PIH and HELLP, and she was delivered by emergency cesarean section. The diagnosis of PIE was made at 22 hours of age. Predisposing factors to the development of PIE in the absence of positive pressure ventilation are prematurity, female gender, and delivery complicated by PIH or HELLP syndrome, or both. The left lung is more likely to be effected. The effect of PIH/HELLP syndrome on the developing lung is unclear. Zhou et al,[8] in a study looked at the pathology of the lung and found that moderate to severe PIH in 49 babies was associated with decreased lung weight. These babies could be inferred as having mildly hypoplastic lungs, thereby predisposing them to PIE. It has been demonstrated

that giving prophylactic surfactant early reduces the incidence of all air leak syndromes.[9] However, our baby had the surfactant quite late and this might be another contributing factor to development of PIE.

In conclusion, preterm infants born to mothers with PIH or HELLP are at increased risk of developing PIE even if they are not ventilated. Therefore, a high index of suspicion strategy should be used to diagnose PIE in nonventilated premature newborns with increasing respiratory distress. Besides, it could be extrapolated from these few cases, that this type of PIE varies from the classical scenario with high inspiratory pressure ventilation. Association of PIE and HELLP needs further study. Furthermore, PIE/HELLP might be another justification for early surfactant treatment in premature babies.

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