

Spontaneous Diffuse Pulmonary Interstitial Emphysema (PIE) in an unventilated infant

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Citation

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Abstract

Pulmonary Interstitial Emphysema (PIE) is a well documented complication of mechanical ventilation, but it also seen in unventilated premature babies. When an apparently healthy, near-term 2 week old infant suddenly developed respiratory distress, PIE was excluded clinically, despite the characteristic honeycomb appearance of the lungs on chest X-ray. However, rare cases of term unventilated babies with PIE are reported in the literature and this was later found to be the underlying diagnosis.

This case is reported to highlight the challenges and complexities of routine medical practice, as well as the need for clinicians to keep in mind rarer differential diagnoses like PIE in a healthy term baby.

INTRODUCTION

PIE is the abnormal collection of gases inside the connective tissue of the peribronchovascular sheaths, interlobular septa, and visceral pleura, secondary to alveolar and terminal bronchiolar rupture. It is a severe condition usually seen as a complication of mechanical ventilation in preterm neonates¹, occurring in 91% of infants with birth weights below 999grams². However, it can occur in premature babies without ventilation or any evidence of respiratory distress syndrome (RDS).

The short term effects of PIE are: ventilation-perfusion mismatch in the affected lung tissue; pressure effects on the normal adjacent lung and mediastinum; and an increased occurrence of pneumothoraces and air emboli. In the longer term, the presence of PIE in very-low-birth-weight infants is associated with a high risk of mortality and significant long-term morbidity, including an increased risk of chronic lung disease. PIE may present as diffuse bilateral involvement or a unilateral lesion.

CASE SUMMARY

Baby A was born at 36+2 weeks gestation by elective caesarean section for twins. He was born in good condition, apgar score 8 at 1 minute, 9 at 5 minutes. He did not require any form of resuscitation after birth. Due to his low birth weight of 1.578kg, he was admitted to the special care baby unit for monitoring of feeding and growth. On day 14, he

suddenly deteriorated. He was found cyanosed, tachypnoeic and in significant respiratory distress. There was no evidence of wheeze, stridor, crepitations or cardiac failure. There was no associated choking or coughing episode and no preceding upper respiratory tract symptoms. A full septic screen was performed, which was later found to be negative. The X-ray chest done at the time of the acute collapse showed the characteristic honeycomb appearance suggestive of PIE with mediastinal shift (Figure 1). However, PIE was discounted as the baby was near term at birth, was now two weeks old, and had not required any resuscitation after birth.

Figure 1



He was started on nasal prong oxygen and covered with broad spectrum antibiotics. After 2 days he was transferred to a tertiary unit in view of an increasing oxygen requirement for further management. His X-ray chest was repeated but remained unchanged.

Following the transfer to the tertiary paediatric respiratory centre, he continued to deteriorate. He was intubated and ventilated, and a CT scan showed large cystic mass in the left hemi-thorax with mediastinal shift to the right. A diagnosis of congenital cystic adenomatoid malformation (cCAM) was made based on the history, degree of hyperinflation and extent of the mediastinal shift. The mediastinal shift became so severe that he underwent a left pneumonectomy. This resulted in a marked improvement and he was quickly weaned off the ventilator into air.

Histological analysis of the resected lung demonstrated changes consistent with PIE, not cCAM. A second CT scan post pneumonectomy showed less severe cystic lesions in the right lung, similar those seen in the now resected left lung. Thus the final diagnosis was confirmed to be bilateral, spontaneous pulmonary interstitial emphysema.

The baby is now clinically well with no further respiratory distress. Twin 2 remained well throughout.

DISCUSSION

Whilst PIE is usually associated with either mechanical ventilation³ or prematurity^{4,5}, rare cases have been reported of term unventilated babies with PIE^{6,7}. PIE has been linked to over distention secondary to high peak inspiratory

pressures, however findings from animal studies suggest that it may be more associated with structural and maturational factors rather than with over distention alone. This is thought to explain why PIE can present as primary problem without any history of respiratory distress syndrome (RDS) or mechanical ventilation⁸.

In this case, the baby did not have any respiratory distress and did not receive any kind of mechanical ventilation. Thus mechanical factors can be excluded as a cause of the PIE. The baby was however a twin, slightly premature, and perhaps most significantly, of low birth-weight. There are no reports in the literature of any association between twinning and PIE. Prematurity is a well recognized risk factor, however this is more usually associated with extreme prematurity¹², rather than 36 weeks. Very low birth weight <1500g is also an independent risk factor⁹, although the significance of a low birth weight greater than 1500g is unknown. We postulate that the unknown factors that caused the low birth-weight also caused a delayed maturation in the lungs with concomitant structural problems that resulted in bilateral PIE. This however remains theoretical and the true cause of the PIE is unknown.

In routine clinical practice, PIE would not be considered a likely candidate to explain sudden respiratory distress in a apparently healthy, near-term infant at 2 weeks age. Even given the characteristic honeycomb picture on X-ray, PIE was discounted at both the local and tertiary units as unlikely on clinical grounds. Whilst a lung biopsy could have identified PIE as the underlying diagnosis rather than a cCAM, whether this would have changed the outcome is unclear.

Treatment options for PIE are limited. Minimal pressure ventilation or high frequency oscillation ventilation are the mainstays, although positioning and ECMO may also have a role. Other treatments, such as selective bronchial intubation or surgical pleurectomy have been tried in the past with little success^{9,10}. The key question is whether or not sufficient resolution of the hyperinflation and mediastinal shift would have occurred without surgery to enable the baby to come off the ventilator. The patient certainly improved rapidly post surgery and is currently doing very well. Pneumonectomy at such a young age is exceeding rare and there is little available information concerning morbidity and mortality. In a study of 45 patients aged 2 days to 12 months who underwent either lobectomies or pneumonectomies, the only death occurred due to underlying cardiac disease,

although significant post-operative complications did occur in 7 (15%) of patients. It is worth noting that being symptomatic pre-surgery and younger age did increase the likelihood of post-operative complications¹¹. In the longer term, the capacity of the remaining lung to grow during the first few years of life is associated with relative preservation of postoperative pulmonary function in children with pneumonectomies and lobectomies¹², albeit with a fixed reduction. However the relative flexibility of their tissue structures carries significant risks for development of scoliosis, thoracic cage abnormalities and postpneumonectomy syndrome (airway compromise due to extreme mediastinal shift). The use of space filling devices has had positive outcomes in such cases in older children¹³, and it is likely that this patient will require further surgical interventions to minimize complications.

Overall, when all these factors are considered, along with the fact that the remaining lung also showed signs of PIE, the longterm outcomes for this patient are impossible to predict. This case is reported to highlight the challenges and complexities of routine medical practice, as well as the need for clinicians to keep in mind the rarer differential diagnoses.

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