Pulmonary interstitial emphysema: a case report and review of the literature

Enfisema intersticial pulmonar: relato de caso e revisão da literatura


Abstract
Pulmonary interstitial emphysema is a rare condition that generally affects low-weight preterm infants submitted to mechanical ventilation. The prognosis is variable, depending on early diagnosis and treatment. The radiologist plays a key role in this scenario. The authors report a case of persistent pulmonary interstitial emphysema, describing the main characteristics of such entity.

Keywords: Pulmonary interstitial emphysema; Newborns; Mechanical ventilation.

Resumo
O enfisema intersticial pulmonar é uma condição rara que acomete, em geral, recém-nascidos pré-termos de baixo peso submetidos a ventilação mecânica. O prognóstico é variável e dependente do diagnóstico e tratamento precoces, ressaltando-se a importância do radiologista neste cenário. Os autores relatam um caso de enfisema intersticial pulmonar persistente, abordando as principais características desta entidade.

Unitermos: Enfisema intersticial pulmonar; Neonatos; Ventilação mecânica.

INTRODUCTION

Still poorly known by non-specialist radiologists, pulmonary interstitial emphysema (PIE) represents an uncommon acute complication in preterm, low-weight newborns who develop respiratory distress syndrome requiring mechanical ventilation (although such characteristics are not essential to define the condition)(1,2). The genesis of such a condition is attributed to the effects of the high pressure levels provided by the mechanical ventilation on a poorly complacent lung, determining alveolar rupture and air leakage into the pulmonary interstitium. Such leakage leads to the development of air trapping cyst clusters which compress the perivascular bundles, generating an even greater decrease in the pulmonary complacency(3,4). Early diagnosis and management of such a condition are fundamental, since simple measures may lead to the resolution of the condition at early stages, while delay in its recognition may imply the necessity of more invasive treatments likely to acute and chronic complications.

In the present report, the authors describe the case of a preterm newborn that developed persistent PIE and, even after a delayed diagnosis, responded favorably to the treatment. The main characteristics of this entity are approached with emphasis on the imaging findings.

CASE REPORT

A male infant prematurely born at 28 gestational weeks, weighting 1,385 grams, developed respiratory distress syndrome. The child was submitted to orotracheal intubation with tracheal instillation of exogenous pulmonary surfactant. Infectious markers were negative. At the fourth day of life, the patients progressed with a small pneumothorax at left which was spontaneously reabsorbed. At the sixth day, still under mechanical ventilation, chest radiography demonstrated a bilateral, diffuse reduction of pulmonary transparency in association with hypoattenuating, serpentine, tubular images different from the air bronchogram pattern (Figure 1). On the following days, worsening was observed in the imaging pattern, with gradual and per-
characterized at left. The set of findings allowed the diagnosis of PIE.

The treatment was immediately instituted with selective intubation of the right main bronchus (Figure 4A) and exogenous pulmonary surfactant reinstillation in association with high frequency and low pressure ventilation. After three days, a Fogarty catheter was inserted and a balloon was inflated in the left main bronchus (Figure 4A), in an attempt to isolate the most affected lung. At the 25th day of treatment, the patient already presented clinical and radiological improvement, with reexpansion of the right lung, and was extubated and referred to the infirmary. Figure 4B shows anteroposterior chest radiographic image acquired three months after the diagnosis and treatment.

DISCUSSION

PIE represents rupture of bronchoalveolar junctions under high pressure, with air leakage into the perivascular and peribronchovascular interstitium. It should not be confused with chronic smokers’ pulmonary emphysema, which is characterized by a permanent expansion of the air space distal to the terminal bronchiole and destruction of alveolar septa in the absence of fibrosis.

Three types of air leakage are described in cases of PIE, as follows: 1) centrifugal, with development of subpleural bubbles and eventual occurrence of pneumothorax, many times presenting as a first sign of the disease, like in the present case; 2) centripetal migration, which may produce pneumomediastinum or pneumopericardium; 3) systemic air embolism, rarely found and almost always lethal, where the air disseminates through lymphatic channels and or alveolo-vascular fistulae into heart chambers, arterial and/or venous vessels.

The risk factors for development of PIE include all the conditions typically found in two situations: 1) those causing reduced pulmonary complacency [prematurity, low Apgar score, respiratory distress syndrome, meconium/amniotic fluid aspiration, infection (neonatal sepsis, pneumonia)]; 2) those determining high intra-alveolar pressure (positive pressure ventilation in the delivery room, use of high peak pressure during...
PIE is classified according the time of disease evolution. Acute PIE is transient and does not last more than one week otherwise it is referred to as persistent PIE, a much rarer presentation. Both presentations may be either focal or diffuse, as well as uni- or bilateral.(1,2,4)

The radiological characteristics of PIE at chest radiography include the presence of hypoattenuating, serpiginous tubular, and sometimes cystic images, which do not fit the air bronchogram pattern(3,4). A peculiar type of focal PIE manifests as single or multiple cysts with regular borders called pseudocysts, most frequently found in the right para-hilar region(3). The tomographic pattern described as “lines and dots” intermingled with large gaseous inclusions is typical, representing peribronchovascular bundles compressed by the air-filled interstitium(1,7), with sensitivity of up to 82% in some studies(1).

The differential diagnosis is made with conditions which present with “cystic chest” in the neonate and include cystic adenomatoid malformation, congenital lobar emphysema, diaphragmatic hernia, pneumatoceles, bronchogenic cyst (in cases of isolated pseudocyst) and aspiration pneumonia(1,2,3,7). The differentiation should be based on the correlation between the onset of clinical and radiological signs and the previous history of tracheal intubation, in a context of reduced pulmonary complacency (although such characteristics are not essential to define the condition).

The treatment is performed in an attempt to reduce the air leakage, and includes lateral decubitus, selective intubation of the contralateral main bronchus, occlusion of the affected main bronchus (with a Fogarty catheter, for example), high frequency and low-pressure ventilation, intermittent physiotherapy with 100% oxygen (in cases of focal and persistent compressive PIE), even iatrogenic pneumothorax/multiple pleurotomies and lobectomy in refractory cases(1,3,8).

Follow-up studies are scarcely found in the literature, but it is known that birth weight < 1,600 grams and development of PIE at the first 48 hours of life represent worst prognosis factors. Survivors present increased risk for developing pulmonary bronchodyplasia and chronic lobar emphysema(3).

CONCLUSION

PIE represents a rare complication affecting principally low birth weight newborns submitted to high pressure mechanical ventilation. The radiologist’s role in the early recognition and diagnosis of such an entity is fundamental, considering that the early treatment is effective in most cases, avoiding surgical interventions and later complications.

REFERENCES