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CASE FOR DIAGNOSIS

A baby with cough and poor feeding

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Case history

A 9-week-old female baby presented at her local hospital with a 10-day history of cough and poor feeding. The pregnancy had been uneventful with normal antenatal scans. The baby was born at term by normal vaginal delivery weighing 3.23 kg with good American Paediatric Gross Assessment Record scores and was sent home on the second day after birth. Prior to this presentation, she had exhibited no respiratory symptoms and was maintaining good growth.

On review, the baby was afebrile with no respiratory distress. Chest auscultation revealed reduced breath sounds on the left side. The rest of the physical examination results were unremarkable. Arterial oxygen saturation was 97% in air.

Initial chest computed tomography (CT) results are shown in figure 1. Unfortunately, the initial chest radiograph was "lost".

One week later, the baby underwent thoracotomy and resection. A chest radiograph taken the day prior to surgery is shown in figure 2.

The histopathology of the lesion is shown in figure 3.

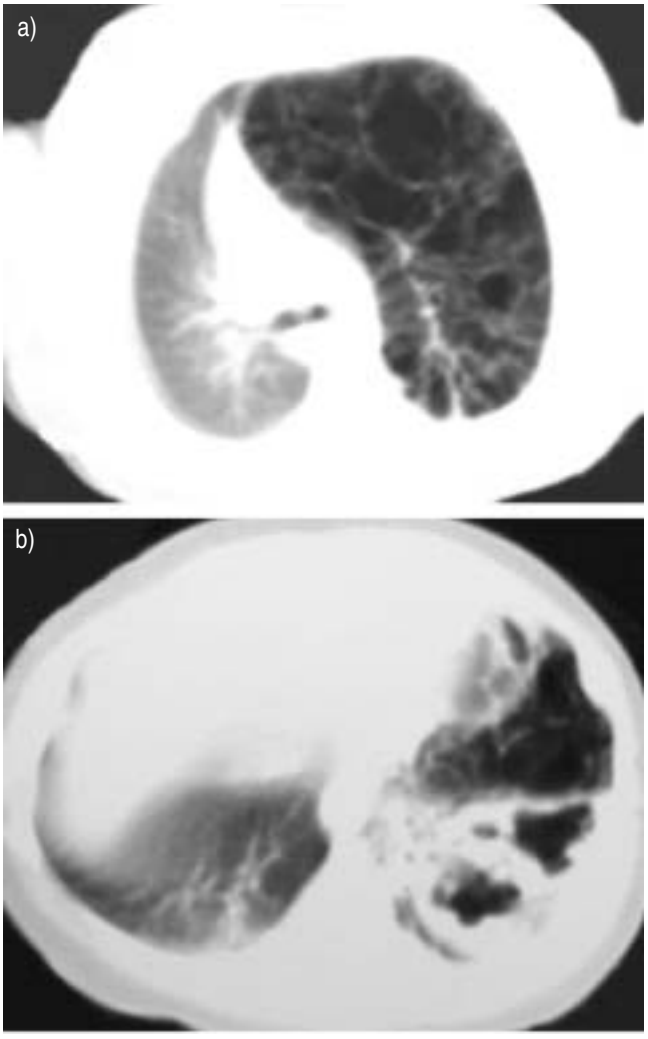

Fig. 1. – Initial computed tomography of the chest showing: a) upper and b) lower lung lobes.

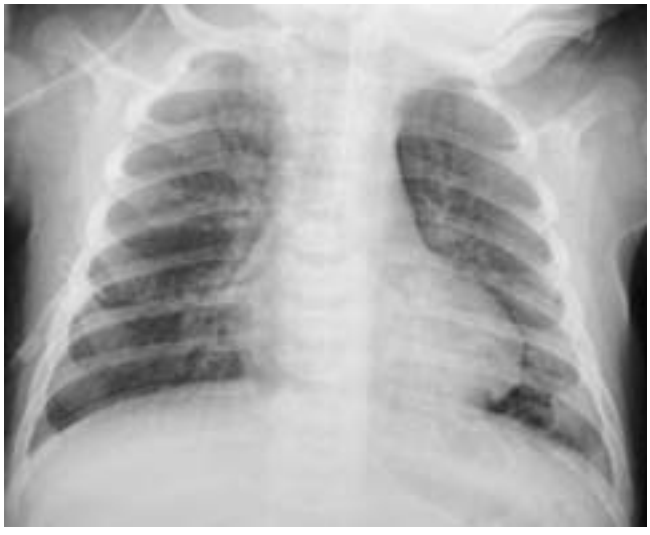

Fig. 2. – Preoperative chest radiograph.

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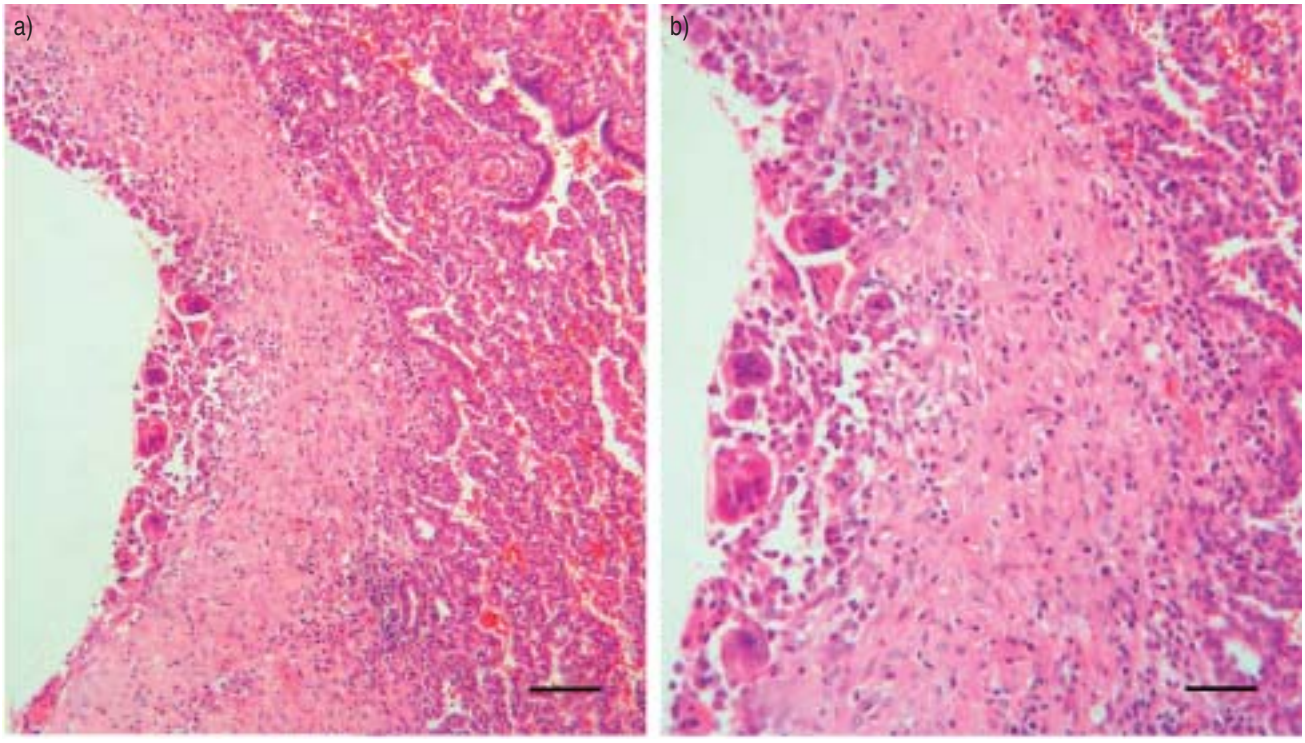

Fig. 3. – Histopathology of lung lesion (haematoxylin and eosin stain). Scale bars a) 200 μm and b) 100 μm.

BEFORE TURNING THE PAGE, INTERPRET THE RADIOGRAPHS, COMPUTED TOMOGRAM OF THE CHEST AND HISTOPATHOLOGICAL RESULTS AND SUGGEST DIAGNOSIS AND FURTHER MANAGEMENT OF THE PATIENT.

Interpretation

Chest computed tomography on presentation

There was marked hyperexpansion of the left upper lobe with multiple cystic lesions and a mediastinal shift to the right (fig. 1a). The left lower lobe was compressed with multiple cysts within (fig. 1b).

Preoperative chest radiograph

The preoperative chest radiograph was taken 9 days after initial presentation (fig. 2). There were cystic changes in the left lower lobe. The changes seen previously in the left upper lobe had completely resolved.

Surgical findings

On surgery, the left upper lobe looked normal, whereas the left lower lobe was congested with signs of infection. The lower lobe was totally adherent to the diaphragm with signs of hyperaemic granulation tissue. Left lower lobe resection was performed.

Histopathology

A 5×4×4-cm lung lobe was received. On cut section, approximately one-half of the normal lung parenchyma was replaced by variously sized cystic spaces. Microscopic examination showed patchy atelectasis and focal mild interstitial fibrosis together with numerous air-filled cystic spaces (fig. 3). These spaces exhibited a fibrous wall and a lining composed of numerous multinucleate foreign-body-type macrophages. There was no evidence of cystic adenomatoid malformation and the picture was that of persistent pulmonary interstitial emphysema (PIE).

Diagnosis: "Localised persistent pulmonary interstitial emphysema of the left lung".

Clinical course

The baby progressed well following lobectomy and made an uneventful recovery. She was asymptomatic 6 months after surgery and chest CT results were normal except for a small area of hyperlucency on the left side.

Discussion

PIE in the newborn is a frequent complication of assisted mechanical ventilation. It most commonly occurs in premature infants with respiratory distress syndrome. Occasionally, it occurs spontaneously with minimal or no respiratory symptoms, as in the present patient. PIE may be acute (<7 days duration) or chronic (or persistent) and may be localised to one or more lobes, or distributed diffusely throughout all lobes. The three forms of PIE, *i.e.* acute PIE, localised persistent PIE (LPPIE) and diffuse persistent PIE (DPPIE), have different clinical, radiographical and pathological features [1–4].

LPPIE usually develops in infants with hyaline membrane disease but can also occur spontaneously with no underlying lung disease, as in the present case. LPPIE is a rare form of interstitial emphysema compared with the diffuse form, with a propensity for localised persistent progressive accumulation of air in the interstitium. This accumulation results in cystic air spaces that are typically associated with mediastinal shift and progressive respiratory distress.

On plain radiography, LPPIE can be differentiated from acute PIE and DPPIE by its expansile multicystic appearance. The cystic air spaces are localised to one or more lobes and are associated with a mediastinal shift and mass effect. Chest CT is helpful in differentiating LPPIE from other lesions, such as congenital cystic adenomatoid malformation and congenital lobar emphysema, as the abnormal air collection is in the interstitium and surrounds the bronchovascular bundles. On CT, the bronchovascular bundle may appear as a soft tissue attenuation nodule or line in the centre of the air-filled cyst (central lines/dots surrounded by lucency) but may be absent or difficult to see [5], as in the present case. Emphysematous distension of the alveoli and air-containing congenital masses do not show this appearance. In equivocal cases, it may be helpful to administer intravenous contrast material that results in enhancement of the vascular component [6].

Pathologically, persistent PIE is characterised by distortion of the pulmonary parenchyma by cysts of various size. These cysts are abnormal air-filled spaces in the interstitium and typically show foreign-body giant cells at their periphery. The postulated pathogenesis is a break in the integrity of the bronchoalveolar system that allows air to escape into the interstitium, where it excites a giant cell and fibroblastic response [7, 8].

Although some cases require surgical resection, in most cases the interstitially leaked air is resorbed, as seen in the present case. Some authors recommend conservative management in the first instance (selective intubation and selective bronchial obstruction and/or decubital positioning) [9–11]. Resection of the LPPIE may be beneficial when there is: 1) significant reduction in effective lung volume producing ventilator dependence; 2) atelectasis and recurrent infections; or 3) recurrent pneumothoraces. This is usually indicated in a small fraction of patients (<2%). These patients can be expected to improve significantly after resection and the long-term outcome is generally good [12].

It is important to differentiate localised persistent pulmonary interstitial emphysema from other radiolucent cystic lesions, such as congenital cystic adenomatoid malformation and congenital lobar emphysema, as this can avert unnecessary surgery in some cases. Careful attention to the round or linear soft tissue component seen in the wall or within air-containing spaces, although not present in all cases, is key to making the correct diagnosis on computed tomography of the chest. In retrospect, surgery was probably unnecessary in the present case but was undertaken because of difficulty in making the diagnosis of localised persistent pulmonary interstitial emphysema in the absence of classical computed tomographic changes. However, the diagnosis should be entertained in cystic lesions that resolve spontaneously over a short period of time.

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