SECTION EDITOR: ENID GILBERT-BARNESS, MD

Pathological Case of the Month

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A 45-DAY-OLD, full-term, male infant weighing 3420 g at birth was admitted to the hospital with a 2-day history of vomiting and a clinical diagnosis of pyloric stenosis. He had been asymptomatic until the presenting complaint. Findings from radiological examination of the thorax revealed the presence of a large lucent cyst in the left upper lobe with mild deviation of the mediastinal structures (**Figure 1**). Routine laboratory analysis findings were normal. Pyloroplasty was performed on the third hospital day. The patient was discharged from the hospital 4 days after surgery and readmitted 1 week after discharge. A computed tomographic scan of the thorax showed a large cystic lesion with smooth borders in the upper lobe of the left lung. A lobectomy was performed.

The 8 × 6 × 4-cm resected lobe contained a 5-cm-diameter cyst involving the upper two thirds of the lobe. The inner surface of the cyst appeared to be crossed by small septa. The cyst contained small amounts of clear fluid. Microscopic sections are seen in **Figure 2** and **Figure 3**.

From the Department of Pathology, Children's Hospital, La Plata, Argentina.

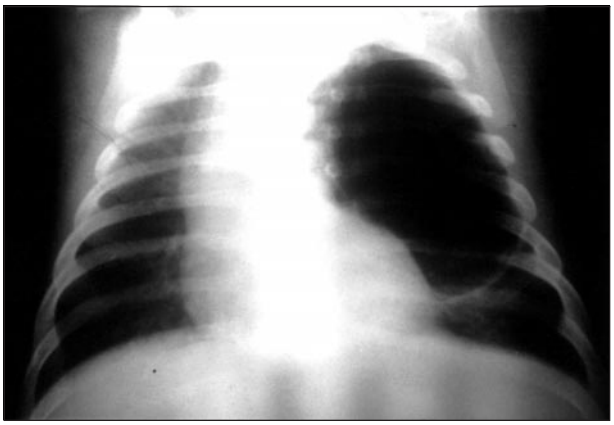

Figure 1.

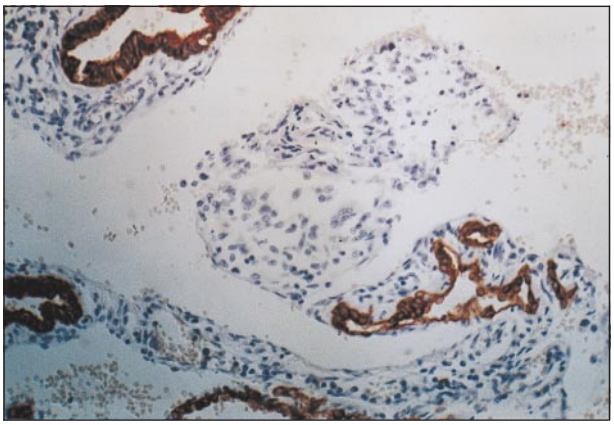

Figure 2.

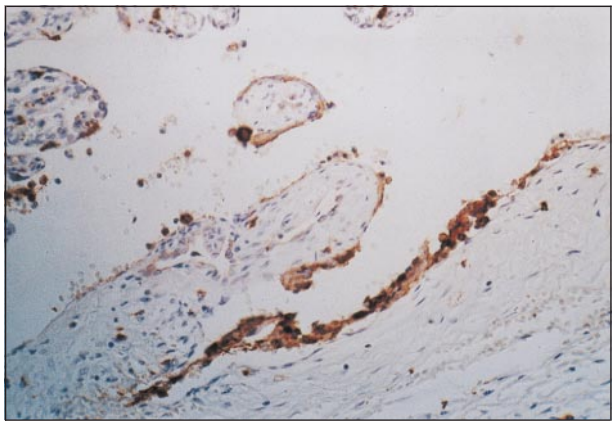

Figure 3.

Diagnosis and Discussion

Persistent Interstitial Pulmonary Emphysema Presenting as a Large Unilocular Cyst in an Asymptomatic Infant

Figure 1. *A chest x-ray film showing a large lucent cyst in the left upper lobe with a mild mediastinal deviation.*

Figure 2. *Keratin-positive alveolar lining cells in the core of the pseudopapillary projections. Nonreactive multinucleated macrophages float in the interstitial spaces (immunoperoxidase for keratin mixture [AE1 and AE3 monoclonal antibodies], original magnification ×40).*

Figure 3. *HAM 56 immunostaining of macrophages in the dissected air space. Intra-alveolar macrophages serve as a positive internal control (immunoperoxidase, original magnification ×40).*

Interstitial pulmonary emphysema in the newborn is a frequent complication of respiratory distress syndrome. It most commonly occurs with assisted ventilation. Occasionally, it is spontaneous. In most cases the interstitially leaked air is reabsorbed. Rarely it persists and becomes progressive, leading to life-threatening situations that may require lobectomy. This progressive form of interstitial pulmonary emphysema may be localized to 1 lobe or diffusely involve the lungs. In both situations pathological examination findings commonly show several confluent small cysts histologically lined by single and multinucleated histiocytes in a foreign-body–type reaction. Bronchi and vessels commonly project in the cavities owing to the dissection of the interstitial tissue produced by the air. Rarely the cysts may reach 1 to 3 cm in diameter.[1-8]

Although a diagnosis of localized persistent interstitial pulmonary emphysema was considered for this infant, he had not been treated with assisted ventilation, and the presence of just 1 large cyst made the diagnosis somewhat questionable. Several signs that are common to localized persistent interstitial pulmonary emphysema were found in this patient (left upper lobe localization, walls with histiocytes, and projections with bronchi and vessels), but many other features appear disparate (full-term infant, no antecedents of assisted ventilation, no symptoms, a unilocular large cyst, and papillary projections within the alveoli).

The images of pseudopapillary projections formed by peripheral lung tissue suggested a certain relationship with the recently described placentoid bullous lesion of the lung,[9] also called placental transmogrification of the lung.[10] However, the immunohistochemical findings showing the absence of an epithelial covering of the pseudopapillations and the presence of numerous HAM 56–positive histiocytes convincingly ruled out such a possibility.

Although some of the histiocytes were positive for S100 protein, we do not believe this case represents an example of histiocytosis X from the lung-induced interstitial emphysema. This histiocytic lesion is distinctively unusual for an infant of this age, and its initial lesion tends to be localized in small bronchi and bronchioli.[11] Eosinophils were absent in our patient.

This patient was an asymptomatic, 45-day-old, full-term infant with no history of assisted ventilation who developed a large unilocular localized persistent interstitial pulmonary emphysema. This example further expands the clinical and pathological spectrum of the complications of pulmonary interstitial air at this age.[6]

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