Persistent pulmonary interstitial emphysema in a case of Langerhans cell histiocytosis

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Abstract

We present the case of a 10-month-old boy with multisystem Langerhans cell histiocytosis showing thin-walled lung cysts along with computed tomography (CT) evidence of persistent pulmonary interstitial emphysema (PPIE), in the absence of pneumothorax or pneumomediastinum. Follow-up CT performed after 6 months demonstrated complete resolution of interstitial emphysema.

Key words: Langerhans cell histiocytosis; lung cysts; persistent pulmonary interstitial emphysema

Introduction

Pulmonary interstitial emphysema (PIE) is a condition that usually affects newborn children with a history of prematurity, lung disease, and mechanical ventilation. It is typically a transient phenomenon, but may persist and form expanding air-filled cystic lesions within the pulmonary interstitium. Radiographic findings are characteristic, and computed tomography (CT) scan is usually not required for a definite diagnosis.

Langerhans cell histiocytosis (LCH) is a disease which occurs in varied clinical settings. It commonly affects young children in a disseminated form with multisystem involvement, while affecting young adults as a localized disease with isolated lung involvement. Pulmonary involvement typically results in formation of lung nodules, cavities, and thin-walled cysts.

We present a case of multisystem LCH associated with persistent PIE. To the best of our knowledge, such an association has not been previously reported in literature.

Case Report

A 10-month-old male child presented with difficulty in breathing for 15 days. His parents had also noticed a skin rash which had first developed 3 months before. There was no history of wheeze, cyanosis, any other complaints or similar episodes in the past. The child had been born after an uncomplicated pregnancy at full term by a normal vaginal delivery and had cried immediately after birth. There was no history of prior hospitalization or assisted ventilation.

On examination, the child was tachypneic, air entry was equal on both sides, and liver was palpable. A vesiculopapular rash was seen over his trunk and extremities.

Chest radiograph showed mild overinflation of bilateral lung fields and a suspicious focal lucency/air-containing cystic lesion in the right mid zone [Figure 1]. No previous radiographs were available for comparison.

High-resolution computed tomography (HRCT) of the chest was advised for further evaluation [Figure 2]. It showed presence of air within the peribronchovascular interstitium in both lungs, radiating outwards from the hila. A large, thin-walled, air-containing cyst/bulla was seen in the region of right middle lobe (also interstitial in location). In addition, multiple small (<1 cm in diameter) thin-walled...
cysts were seen scattered within the lung parenchyma bilaterally. Intervening lung parenchyma showed areas of patchy ground-glass opacities. There was no associated pneumothorax or pneumomediastinum. No lung nodules were seen. Scans through the upper abdomen revealed hepatomegaly with periportal hypodensities.

Based on the CT findings, a provisional diagnosis of (multiorgan) LCH with persistent pulmonary interstitial emphysema (PPIE) was given. A skin biopsy [Figure 3] revealed findings consistent with LCH. The child was put on chemotherapy (including injection vincristine in a dose of 1.5 mg/m² on day 1) and injection cytosinearabinoside-dose 100 mg/m²/d for 4 consecutive days). Total therapy was given for 16 weeks, and child was kept on follow-up. A repeat CT [Figure 4] was performed 6 months later, which documented complete resolution of the interstitial emphysematous changes. Numerous thin-walled lung cysts were still present, along with ground-glass opacities. The child is still on follow-up. His parents report an improvement in his general condition, including his breathlessness.

**Discussion**

LCH is a disorder with diverse clinical presentations, and may present with involvement at a single osseous site or may show multisystem involvement. Disseminated LCH is a severe multisystem disorder that commonly occurs in young children and has a poor prognosis. Among the extrarosseous sites, skin involvement has been reported in around 55% cases and lung involvement in 26% cases with multisystem disease. At the other end of the spectrum, isolated involvement of the lungs may also occur, which is known as (isolated or localized) pulmonary LCH, and commonly
affects young adults, especially cigarette smokers. The imaging appearance of pulmonary involvement is similar in both the localized and disseminated forms of the disease. When compared with lung involvement as a part of disseminated disease, isolated lung involvement has a better prognosis. However, clinical outcomes in these patients are variable – 50% show clinical and radiographic stability, 25% show spontaneous regression, and 25% may show progressive replacement of lung parenchyma by cysts, and may result in end-stage lung disease.[9]

Though the etiopathogenesis of this disease is incompletely understood, the most accepted theory suggests an underlying immunological process. The disease manifestations result from an abnormal proliferation of Langerhans cells (LCs).[3] LCs are a type of dendritic cells, which are cells of the immune system that are concentrated mostly in sites like the skin, gastrointestinal tract, and lungs, i.e. the organs which are in close contact with stimuli from the external environment.

Lung lesions in LCH are centered around bronchioles and have been described to extend both proximally and distally along the airways.[41] Temporal evolution of these lesions occurs, which is reflected in pathological as well as radiological appearances. Initial infiltration of bronchiolar epithelium and walls by LCH cells is followed by development of cellular nodules containing LCH cells along with variable numbers of lymphocytes, neutrophils, plasma cells, macrophages, and eosinophils. These are gradually replaced by fibrotic scars. Cysts may develop in and around these LCH lesions – they are postulated to represent ectatic airway lumina resulting from inflammation of bronchiolar walls, as well as areas of paracatricial traction emphysema.

HRCT chest is the gold standard for radiological diagnosis of lung involvement in LCH. Typical CT findings reflect the above described temporal evolution of disease, and progress from lung nodules (typically less than 1 cm in diameter) to cavitating nodules, eventually giving rise to thick-walled cavities, then thin-walled lung cysts, and finally end-stage lung disease. As the disease progresses, lung cysts predominate and nodules occur less frequently.[5] No lung nodules were demonstrable in our case.

Lung cysts in LCH have been described to be thin walled (wall thickness upto 2 mm) in a greater percentage of cases (as compared to thick-walled cysts).[10] Cysts are typically small (<10 mm in size), though larger cysts (10-20 mm and even > 20 mm) may also be seen.[7] Our case also showed numerous thin-walled small (<1 cm) cysts in the lung fields bilaterally. Though more commonly round or ovoid in shape, cysts have also been described to have bilobed, cloverleaf, or branching morphologies, and it is postulated that such appearances may result from coalescence of multiple cysts.[9]

Pneumothorax is a well-recognized complication of LCH and may occur as the presenting manifestation of disease, and may also be recurrent.[10] The increased predisposition to developing pneumothorax is thought to result from destructive changes in the lung parenchyma and presence of multiple cysts. Pneumomediastinum may also be seen in association. However, to the best of our knowledge, the occurrence of PIE, as seen in our case, has not been previously described in literature. We postulate that the likely mechanism of development of interstitial emphysema was air leak into the interstitium facilitated by the bronchiolocentric location of the disease. As a consequence of inflammation and possible rupture of the small airways, there was entry of air into the lung interstitium, which then propagated proximally and distally within the peribronchovascular interstitium giving rise to the typical “line and dot pattern” described on HRCT in cases of PIE.[10] This finding is attributed to the bronchovascular bundles which are surrounded by air in interstitial spaces, and appear as central lines or dots depending on their orientation to the plane of the CT section.

In addition to this line and dot pattern, our case showed one large cystic lesion which was also located within the interstitium. Such large bullous masses have been described in cases of PPIE.[11] However, cases of PPIE are typically described in neonates born prematurely who have a history of lung disease and mechanical ventilation. Persistent interstitial emphysema may be unilateral or bilateral, involving a single lobe or may be multilobar. Spontaneous resolution (both partial and complete) of these lesions has been described in cases kept on follow-up.[12]

The resolution of the interstitial emphysema in our case was documented on HRCT done while on follow-up after chemotherapy. As spontaneous resolution of this condition is known,[13] the role of chemotherapy itself contributing toward this resolution cannot be conjectured. The other HRCT findings of lung cysts and ground glass attenuation were found to persist on the follow-up scan.

Our case is unique because of the occurrence of bilateral PPIE in a case of LCH (in association with other imaging features), demonstrating a characteristic CT appearance. The importance of diagnosing PIE lies foremost in differentiating it radiologically from other luencies and cystic lesions. Bullous lesions associated with PPIE may expand to large sizes and cause compression of the lungs leading to respiratory compromise.[11] Also, PPIE may be a precursor for pneumothorax or pneumomediastinum, which may be life-threatening.[12] HRCT is helpful in making a definite radiological diagnosis.
Abbey, et al.: Pulmonary interstitial emphysema in LCH

References


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