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RADIOLOGY IMAGES

Job's syndrome presenting with a tension pneumothorax and a lung abscess

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A 19-year-old male presented to the emergency department with severe hypoxic respiratory distress. He had a history of hyperimmunoglobulin E syndrome (HIES) diagnosed at the age of 24 months. He had recurrent skin abscesses with *Staphylococcus aureus* and multiple respiratory infections with *S. aureus* and *Pseudomonas aeruginosa*. He had also developed bronchiectasis and required left lower lobe lobectomy in early childhood. Chest X-ray (CXR) revealed a right tension pneumothorax with a large lung abscess (Fig. 1). A chest tube was placed (Fig. 2) and empiric antibiotics were initiated. The patient refused any further intervention for the abscess and the pneumatocele. The chest tube drained purulent-appearing material that suggested a possible ruptured pulmonary abscess into the pleural space. The sputum and fluid culture grew *Klebsiella oxytoca* and *Enterobacter gergoviae*. A repeat chest tomography a week later revealed resolution of the pneumothorax with improvement of the lung abscess and infiltration (Fig. 3). After a 2-week treatment with intravenous antibiotics, he

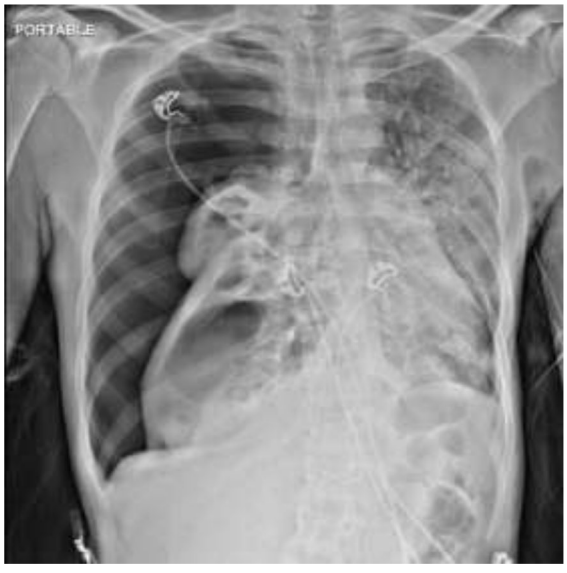

Fig. 1. Complete right pneumothorax with a collapsed right lower lung cavitary lesion and mediastinal shift to the left (Day 1).

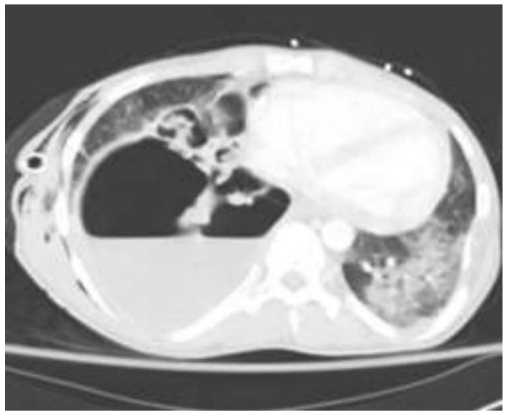

Fig. 2. Resolved pneumothorax with a chest tube. A large fluid filled cavitary lesion in the right lower lobe and infiltration in the left lung (Day 1).

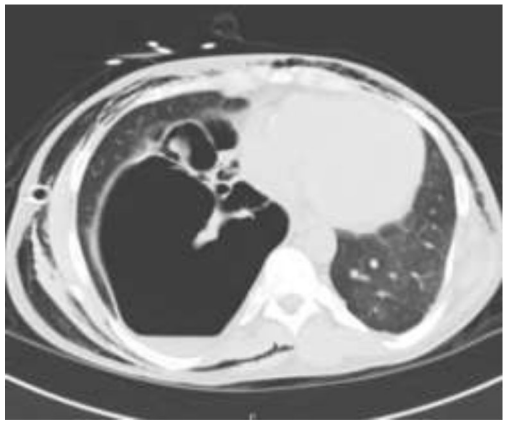

Fig. 3. Marked improvement of the fluid in the cavity and infiltration (Day 7).

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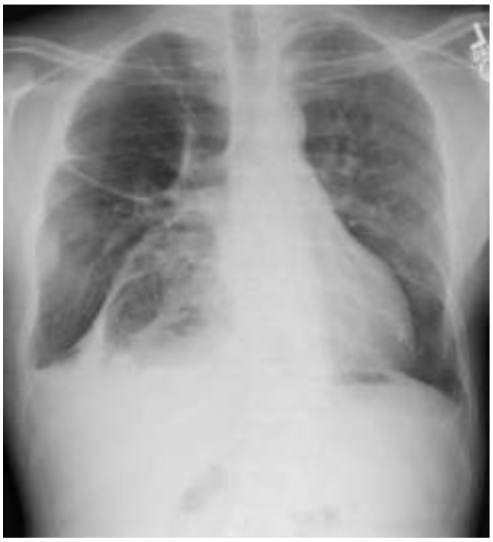

Fig. 4. Decreased fluid in the cavity with persistent right lung bulla (8 months).

was discharged on oral trimethoprim-sulfamethoxazole for prophylaxis. Eight months after the discharge, the patient was doing well saturating 97% on ambient air. The repeat CXR revealed decreased fluid in the cavity with a persistent right upper lobe bulla (Fig. 4).

HIES is a rare immunodeficiency disorder characterized by recurrent skin and sinopulmonary infections, unique facial features, skeletal and vascular abnormality in addition to high immunoglobulin E level. It is known as 'Job's syndrome' based on a description of the biblical character Job. Approximately 200 cases have been reported worldwide (1). Mutations in the signal transducer and activator of transcription 3 (STAT3) and tyrosine kinase 2 (Tyk2) genes have been found in HIES (2, 3). The abnormal cell signaling caused by the genetic defects leads to various manifestations of HIES. Most common organism for recurrent pneumonias is *S. aureus*. Pneumonias are frequently complicated by bronchiectasis, bronchopleural fistulae, and pneumatoceles (4). Secondary infections are often caused by *P. aeruginosa*, *Aspergillus*, and non-tuberculous mycobacteria. Secondary infections have been associated with more complications and mortality (5). Lung abscess and empyema require drainage. Interventions should be considered with caution for those with limited lung capacity due to preexisting pulmonary complications. Use of immunomodulating agents and hematopoietic cell transplantation has not been established (6, 7). Aggressive treatment for acute skin and pulmonary infections and preventing infectious complications is most crucial in management.

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