A 20-MONTH-OLD infant had a 4-week history of bilaterally bulging neck masses that were apparent while crying. The masses were initially noted after an upper respiratory infection, and they became progressively more prominent. The infant had several episodes of difficulty swallowing, mild hoarseness, and occasional perioral cyanosis during a 5-day hospitalization to evaluate the neck masses. The infant's medical history was notable for respiratory syncytial virus bronchiolitis at age 6 months and several episodes of otitis media. There was no history of trauma or surgery.

Physical examination showed a playful child in no respiratory distress but with prominent bulging of the supraclavicular fossae bilaterally (Figure 1) on crying. The swelling disappeared when the infant was consoled (Figure 2), and the remainder of the physical examination was normal.

A flexible fiberoptic bronchoscopy showed no laryngeal or tracheal abnormality during quiet breathing, crying, or coughing. A barium esophagogram and chest computed tomography scan were normal. Chest and neck radiographs were obtained under fluoroscopy while the child was crying (Figure 3) and quiet (Figure 4).
Denouement and Discussion

Bilateral Congenital Apical Lung Herniation

Figure 1. The infant while crying showing a lower mass with obliteration of the supraclavicular fossa.

Figure 2. The infant while quiet with disappearance of the neck mass.

Figure 3. Radiograph of the neck and upper chest at fluoroscopy while the infant is crying. There is protrusion of both lung apices (right more than left) into the neck (arrows). Slight deviation of the trachea is also apparent.

Figure 4. Radiograph of the neck and upper chest at fluoroscopy while the infant is quiet. The lung apices returned to their normal location in the chest, and the tracheal deviation disappeared.

Lung herniation is a protrusion of the lung parenchyma beyond the musculoskeletal thorax. It was initially described by Roland in 1499 and later by Hildanus and Loysean in 1606 and 1617, respectively. Fewer than 300 cases are reported in the literature.

Lung hernias are classified by location and etiology. Categories are intercostal, cervical, and diaphragmatic. Sixty-five percent of lung hernias occur through the intercostal spaces, 33% are cervical through the thoracic inlet, and 1% are diaphragmatic. Herniation of 1 lung across the mediastinum occurs because of overinflation of the ipsilateral lungs and small or absent contralateral lung. Lung hernias are mostly acquired from penetrating injuries or blunt trauma to the chest wall, multiple rib fractures, and tear of intercostal muscles. Only 18% of lung hernias are congenital. Congenital herniation of the lung occurs mainly in the cervical region due to weakness of the paratracheal fascia. Congenital intercostal herniation through defects of the anterior thoracic wall has been associated with costal cartilage malformation. Pathological hernias can occur when breast or chest wall malignancy, tuberculous osteitis, or empyema weaken the thoracic wall.

Congenital cervical lung hernia usually manifests during the first year of life but may not be apparent at birth. Familial cases are associated with hernias in other locations. Cervical lung herniation occurs through the apex of the hemithorax at the thoracic inlet. The trachea, esophagus, and neurovascular structures are flanked dorsally by the trapezius and the 3 scalenus muscles. However, a gap is present ventrally between the scalenus anterior and the sternocleidomastoid muscles.

The ascent of the lung beyond the thoracic inlet is prevented by Sibson fascia, an opposed 2-layered fascia in the neck, the parietal pleura, and the neck muscles. Cervical lung hernia occurs with partial absence, laxity, or attenuation of Sibson fascia or with an increase in space between the sternocleidomastoid and scalenus anterior muscles.

Unilateral or bilateral lung herniation is usually noted during the first year of life as a bulging mass with a Valsalva maneuver and increased intrathoracic pressure that is apparent during crying. Lung apex herniation is temporary, and the lung apices return to the thoracic cavity spontaneously. Cervical hernia may produce hoarseness, chronic cough, or stridor from external compression of the trachea or transient impairment of jugular venous drainage. Pain or tenderness is rarely present at the hernia site. The lung apices feel like an elastic, compressible mass with crepitus, and cough can be elicited by palpation. Acute respiratory distress from incarceration with an ischemic or perforated lung has been reported in isolated cases.

Congenital cervical hernias are apparent on chest and neck radiographs. With accompanying tracheal deviation, chest computed tomography scans determine the location and size of the cervical defect and exclude other masses. The radiological differential diagnosis of apical lung hernias includes pharyngocele, laryngocele, and esophageal diverticulum. Contrast esophagogram helps to differentiate esophageal diverticula. Recognition of lung hernia helps to avoid accidental iatrogenic lung trauma when placing a central venous catheter.

Adults with cervical lung hernia may have chronic cough or emphysema or occupations with repetitive respiratory maneuvers and increased intrathoracic pressure, such as playing wind instruments, glass blowing, or lifting weights. The combination of increased intrathoracic pressure and focal weakening of the thoracic wall leads to herniation.

Apical hernias in infants and children usually resolve spontaneously. Treatment is warranted if incarceration or significant compression of nearby structures occurs. Surgical reduction and repair using tissues such as peristomeum, muscle, or synthetic materials can be performed. Video-assisted thoracoscopic repair of lung hernias has also been reported.

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