Persistent Middle Lobe Abnormality in an East African Male*

Capt Steven E. Gradwohl, M.C., U.S.A.;
Maj Robert A. Dietrich, M.C., U.S.A.;
and Maj Warren L. Whitlock, M.C., U.S.A.

A 12-year-old East African boy presented in June 1987 with fevers, cough, and malaise. His chest roentgenogram was interpreted as showing a right middle lobe infiltrate. He was treated with erythromycin for two weeks and had clinical resolution of his symptoms. The repeated chest roentgenogram showed persistent abnormalities (Fig 1 and 2).

The patient had immigrated to the United States one year earlier. He had a history of recurrent pneumonias during his childhood but an otherwise normal development. He had also had an ascaris infection two years earlier. He was thin and at the 85th percentile for height and weight. Chest examination showed scattered crackles in the right anterior axillary line and decreased breath sounds in the right upper lobe. There was no lymphadenopathy or clubbing. Laboratory data showed a hematocrit of 39.4 percent, a WBC count of $6.2 \times 10^9/L$ (7 percent eosinophils), normal serum chemistry values, and a negative intermediate-strength PPD. Pulmonary function revealed an FVC of 53 percent of predicted, a FEV1/VC of 88 percent, a TLC of 85 percent of predicted, and a Dco of 100 percent of predicted. The posteroanterior and lateral chest roentgenograms (Fig 1 and 2) showed unilateral hyperlucent lung with a nodular mass adjacent to the right hilum. Right middle lobe atelectasis was also present. An expiratory view (Fig 3) demonstrated air trapping and midline shift.

*From the Pulmonary Disease Service, Department of Medicine, Letterman Army Medical Center, Presidio of San Francisco, CA. The opinions or assertions contained herein are the private views of the authors and are not to be construed as official or as reflecting the views of the Department of the Army, the Department of Defense, or the U.S. Government. Reprint requests: Dr. Whitlock, Letterman Army Medical Center, Presidio of San Francisco 94129-6700

FIGURE 1

Roentgenogram of the Month (Gradwohl, Dietrich, Whitlock)
Diagnosis: Congenital bronchial atresia

The differential diagnosis of the unilateral hyperlucent lung can be divided into those with a radiologically demonstrable soft tissue mass and those without. The latter category includes infantile lobar emphysema, pulmonary embolus, unilateral perihilar bullous emphysema, proximal interruption of the pulmonary artery, and the Swyer-James syndrome. The diagnoses to consider in unilateral hyperlucent lung with a mass include endobronchial obstruction (benign/malignant neoplasm or foreign body), exobronchial obstruction (lymph nodes, extralobular sequestration, mediastinal bronchogenic cyst or tumor), mucoid impaction (allergic bronchopulmonary aspergillosis), and congenital bronchial atresia (CBA).

First described by Ramsey in 1953, CBA is an anomaly characterized by a blindly terminating bronchus with hyperinflation of the segment distal to the obstruction. There is a slight male preponderance, with cases coming to diagnosis from birth to age 44 years. Most are asymptomatic at presentation; however, varying clinical courses ranging from respiratory distress in the newborn to recurrent pneumonias, dyspnea, and chest pain are recognized. A static roentgenogram reveals localized hyperlucency usually of the left upper lobe, although any segmental or lobar bronchus may be involved. A variably shaped soft tissue mass extending from the hilum, occasionally with an air-fluid level, is common. Expiratory views reveal air trapping. Angiography, bronchography, and radionuclide scans are helpful in differentiating CBA from other entities; however, CT has largely replaced these techniques.

Pathologic examination of resected specimens has revealed a markedly emphysematous, hyperinflated, and nonpigmented segment often compressing adjacent normal lung. A bronchocele containing mucus and desquamated material is usually seen just distal to the point of atresia with no patent proximal connection with the bronchial tree. This filled bronchocele represents the perihilar soft tissue density seen on the roentgenogram. The observed hyperinflation is thought to occur by collateral intersegmental air drift via intra-alveolar (pores of Kohn), bronchoalveolar, and interbronchiolar channels. Normal branching bronchial generations are demonstrated distal to the obstruction; however, the alveoli are decreased in number and size, perhaps reflecting diminished ventilation and perfusion during development, which results in reduced "work hypertrophy."

The etiology of CBA remains undetermined. It is thought to arise from some insult occurring after 16 weeks of development, since the postatretic bronchial tree is normal. Several authors cite embryologic vascular instability as a possible initiating factor. The prognosis for CBA is fairly benign, and most asymptomatic cases can simply be followed up. Surgical indicators have been established and include recurrent pneumonias or limiting dyspnea.

Bronchoscopy in our patient revealed an abnormal right upper lobe takeoff with only two segmental orifices and an abnormal cartilaginous bridge at the usual location of the anterior segment. Thoracotomy with right upper lobectomy was performed, confirming the diagnosis of CBA of the anterior segment of the right upper lobe. The right middle lobe appeared normal and was not resected. The patient remains well without recurrence of pulmonary infection.

REFERENCES