A 3-YEAR-OLD girl had a history of recurrent cough, fever (up to 38.8°C), dyspnea, and weight loss since age 15 months and a family history of tuberculosis. An intradermal tuberculin test (purified protein derivative) had positive results (15 × 15-mm induration). Findings from chest radiograph (Figure 1) revealed a nonhomogeneous pulmonary opacity with irregular borders on the right lower lobe that did not resolve with antituberculous therapy over 1 year. On examination, she had an asymmetrical thorax and minimal intercostal retractions. Breath sounds were decreased over the right lower lobe. Immunoglobulin levels, nitroblue tetrazolium reduction test results, and sweat chloride concentration were within the reference range. Thoracic computed tomography with contrast medium revealed multiple cystic lesions in the right lower lobe, infiltration of the lower lobe, and increase in right lung volume (Figure 2). No foreign body substance was found on bronchoscopy. A right lower lobe superior segmentectomy via thoracotomy was performed. There was no systemic arterial supply to the lesion, excluding intralobar sequestration. The resected lung had a cystic mass (4.0 × 3.5 × 1.5 cm) (Figure 3 and Figure 4). One year after surgical resection, the chest radiograph appeared normal (Figure 5).

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Diagnosis and Discussion

Congenital Cystic Adenomatoid Malformation of the Lung

Figure 1. Chest radiograph obtained at age 15 months shows nonhomogeneous pulmonary opacity with irregular borders on the right side.

Figure 2. Thoracic computed tomographic scan with contrast medium shows multiple air-filled cystic lesions of various sizes and increased right lung volume.

Figure 3. A low-power photomicrograph shows bronchiolar-like multiple cystic structures lined with columnar epithelium in the pulmonary parenchyma (hematoxylin-eosin, original magnification ×32).

Figure 4. A high-power photomicrograph shows multiple cystic structures lined by ciliated columnar epithelium, surrounded by smooth muscle and scattered cartilage tissue and interstitial lymphocytic infiltrations (hematoxylin-eosin, original magnification ×125).

Figure 5. A chest radiograph shows no pathological findings 1 year after surgical resection.

Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung. First described by Ch’in and Tang in 1949,1 it has no association with race, sex, maternal age, or familial predisposition.2 It is characterized by an adenomatous overgrowth of the terminal bronchiolar-like tubular structures with subsequent suppression of alveolar growth, probably resulting from an embryologic insult before the 49th gestation day.3

Histologic features described by Kwittken and Reiner4 include an increase of terminal respiratory bronchiolar-like structures lined by respiratory epithelium and connective tissue stroma containing disorganized elastic tissue and smooth muscle. Mucous glands and cartilage plates, except as components of entrapped normal bronchi, are not found. Inflammation is not present in infants but may be found in older patients.2 Pathologically, CCAM has been classified into 5 types5: Type I is characterized by multiple large cysts; type II, multiple evenly spaced cysts no greater than 1 cm; type III, a solid lesion; type IV, peripheral cyst; and type V, fatal acinar dysplasia. The lungs are firm and small. The lesion is usually unilateral and may occur in any lobe. Unlike intrapulmonary bronchogenic cysts, CCAM is supplied by the pulmonary artery, and the cysts communicate with the tracheobronchial tree.3

Patients with CCAM can present as neonates with severe, progressive respiratory distress or as older children or adults with recurrent pulmonary infections localized to one lobe. Small lesions cause few symptoms. Cyst expansion causes respiratory distress, severe dyspnea, and cyanosis at birth or soon after. A predominately solid lung mass is usually found in stillborn or premature infants and is associated with hydrops, ascites, and polyhydramnios. On occasion, CCAM may remain asymptomatic and be discovered incidentally on a chest radiograph later in life.6,7

Definitive diagnosis is made on histologic examination.7 Radiologic findings are usually diagnostic and vary depending on the type of underlying malformation. Chest radiography may show multiple or single cysts or a solid mass. Postpneumonic pneumatoceles, congenital lobar emphysema, bronchogenic cysts, and congenital diaphragmatic hernia in the newborn should be considered in the differential diagnosis, for which thoracic computed tomography or magnetic resonance imaging is often required. Prenatal diagnosis is possible by ultrasonography after the 16th week of gestation.2,3

The perinatal prognosis depends on the size of the lung lesion and the secondary physiologic derangements. The lesion can either spontaneously regress or disappear8 and be asymptomatic at birth, or it can progress to cause either fetal death from hydrops or neonatal death owing to associated pulmonary hypoplasia. Ultrasonographic findings suggesting a poor prognosis include polyhydramnios, hydrops, ascites, mediastinal shift, and entirely adenomatous lesions.7 Pregnancy termination may be recommended in cases with hydrops, severe associated congenital malformation, and chromosomal abnormalities.8 In utero drainage of the large cysts or removal of large solid lesions has led to improvement in the fetus and normal lung tissue growth.7,9 Most infants with poor prognosis in the fetal stage die in the newborn period. In all lesions of suspected CCAM, resection is recommended to avoid respiratory distress and recurrent infection. Long-term prognosis, depending on the amount of normal lung tissue after surgical resection, is relatively good.10

Accepted for publication May 24, 1999.

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