ulcerations of the larynx. Recently, Gaillard et al\textsuperscript{5} described a 34-year-old man who, after three years of chemotherapy for chronic lymphocytic leukemia, developed a subglottic lymphomatous infiltrate. The tumor rapidly shrank in response to local radiotherapy.

Localized lymphomas at this site are also rare. In 5,319 consecutive patients with malignant lymphoma seen at the Mayo Clinic, DeSanto and Weiland\textsuperscript{4} found only nine who had primary laryngeal tumor—six with lymphosarcoma, three of reticulum sarcoma.

Climie et al\textsuperscript{6} have described a unique lymphoid hamartoma arising in the pyriform sinus. Pathologically, it was characterized by prominent lymph follicles, but absent lymph sinuses. Al-Saleem and associates\textsuperscript{7} reviewed five cases in the world literature of tumor-like hyperplasia of the reticuloendothelial system localized in the larynx. The same authors reported an additional patient with supraglottic pseudolymphoma. None of the six patients had systemic blood abnormality or regional node involvement.

Studies of disseminated lymphomas have rarely shown laryngeal involvement. Sugarbaker and Craver\textsuperscript{8} found the larynx affected in 0.5% of patients with hemagenous or lymphomatous spread of lymphosarcoma. Shilling, et al\textsuperscript{9} autopsied 19 patients who died of malignant lymphoma or leukemia. Sixty percent of these patients had histologic involvement of the larynx. Granulocytic leukemia was the usual cause of clinically apparent tumor characterized by stridor. The authors associated these larger tumors with the hemorrhage and necrosis more common in granulocytic infiltrates. None of the three patients with lymphocytic leukemia had more than microscopic involvement.

\textbf{REFERENCES}

7. Sugarbaker ED, Craver LF: Lymphosarcoma, a study of 196 cases with biopsy. JAMA 115:17, 112, 1940

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\section*{Congenital Diffuse Bronchio-Alveoliectasis: A Variant of Congenital Cystic Disease of the Lungs*}

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This report presents the clinical and pathologic features of diffuse cystic transformation of a lobe of the lung in a newborn. Because of the rather unusual but interesting histologic picture which distinctly segregated it from the so-called congenital cystic adenomatoid malformation of the lungs, the name congenital diffuse bronchio-alveoliectasis was suggested. This apparently rare anomaly manifested as respiratory distress since the patient's birth. The patient underwent lobectomy with an uneventful postoperative course.

Congenital diffuse cystic disease of the lungs of infants is rare. It has been known for decades under several names such as cystic fetal bronchial adenoma, idiopathic diffuse bronchiectasis, congenital cystic adenomatoid malformation of the lungs, congenital lobar emphysema, congenital pulmonary lymphangetiasis, congenital hamartoma and many more. This diversity of names reflects the various gross and histologic types of what are probably the same condition.

\section*{Case Report}

The patient, a nine-day-old white boy was admitted to the Jewish Hospital and Medical Center of Brooklyn because of severe respiratory distress since birth. Physical examination on admission disclosed marked xiphoid retractions. Breath sounds were absent on the left side and diminished on the right side on auscultation. Complete blood count (CBC), blood chemistries and electrolytes, and results of urinalysis were within normal limits. Chest roentgenograms disclosed increase in volume of the left upper lobe of the lung, which was ridged with multiple densities alternating with irregular radiolucent areas (Fig 1). There was a density at the left cardiac border thought most likely to represent a collapsed left lower lobe. The heart and mediastinum were shifted to the right. An emergency thoracotomy was performed. A huge, voluminous, dark pink, emphysematous, noncollapsible left upper lobe of the lung was delivered into the operating field. A left upper lobectomy was carried out. The left lower lobe, which was collapsed, expanded satisfactorily when positive pressure was applied. The postoperative course was uneventful. Postoperative roentgenograms showed good expansion of the right lung and remaining left lower lobe, normal shift of the heart and mediastinum and normal sternal depression. The child was discharged in satisfactory condition.

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increase supported

Surgical Specimen

The specimen consisted of a left upper lobe of the lung that was greatly increased in mass, measuring 10 x 7 x 5 cm and weighing 140 gm. The lobe was semifirm and presented an irregularly bosselated but smooth and glistening pleural surface that lacked the normal lobular pattern. Visible primary bronchi were demonstrated some of which directly communicated with the multicystic spaces. The pulmonary arteries and veins were normal in size, caliber and distribution. The cut surface of the fixed mass revealed numerous irregular cystic channels varying in size from 0.2 cm to 1.3 cm in diameter, the spaces being smaller and less prominent at the periphery (Fig 2). Most of the cystic spaces intercommunicated with one another.

Microscopic Findings

Histologic examination revealed diffuse cystic dilatation of bronchi, bronchioles and corresponding alveolar ducts (Fig 3). With the exception of the main, entering bronchus, no cartilage-supported bronchi were found, nor were there any branching mucous glands found in any bronchial and bronchial structures. There appeared to be a great increase in the number of the respiratory bronchioles lined by single rows of ciliated to nonciliated low columnar epithelium supported by a thin layer of smooth muscle cells (Fig 4). Elastic stain (Weigert's method) demonstrated no unusual increase in elastic tissue. The dilated terminal bronchioles communicated freely with the adjacent alveolar structures in a branching pattern, the latter exhibiting the overall picture of exaggerated, uniformly diffuse, cystic emphysema. The lining of the alveoli was made up of low cuboidal cells with fairly abundant cytoplasm and centrally placed nuclei. No inflammation was noted but a few aspirated squamous cells presumably derived from the amniotic sac, were present in the larger alveolar spaces. The venous and arterial supply at the hilus, except for moderate engorgement, was unremarkable.

Discussion

Diffuse congenital cystic disease of the lungs, in which cystic bronchiectasis and cystic emphysema form major subgroups, is an apparently rare condition found almost exclusively in infants. Its pathogenesis, up to the present time, is still not fully understood. It appears, however, that the condition is a true developmental defect which is initiated when the lung anlage has already undergone lobulation but at a time when structures such as cartilage, mucous glands, alveoli and arteries are not yet differentiated.
CONGENITAL DIFFUSE BRONCHIO-ALVEOLECTASIS

The malformation generally affects one or two lobes of the lung or occasionally a portion of a lobe. The condition usually manifests itself in the first few hours or days of life as respiratory distress which is related to the pressure effects of the lung on the mediastinum. Survey of early reports shows that there is a high incidence of prematurity in infants with congenital pulmonary cysts. The condition is usually associated with anasarca of the infant as well as polyhydramnios as a feature of pregnancy. However, it is apparent in these reports that most of the instances are in stillborn and in premature infants. It is postulated that the most likely etiology of the anasarca appears to lie in the increased intrathoracic pressure resulting from the enlarged bulk of the anomalous lung inasmuch as cardiac hypertrophy or dilatation is conspicuously absent in the published cases. In recent reports, in contrast with the earlier ones, anasarca and polyhydramnios are stated to be absent, as in the current case.

Several classifications of congenital cystic disease of the lungs are in the literature. Stowens preferred to subdivide it into five categories: 1) embryonal, 2) cystic bronchiectasis, 3) alveolar (cystic emphysema), 4) adenomatoid or hamartomatous and 5) lymphangiectatic. Cooke and Blades have offered a simplified classification: 1) bronchogenic cells, solitary or multiple, 2) alveolar cells, solitary (pneumatocele, balloon cyst), or multiple and 3) combined.

It must be emphasized at this point that mixtures of the different varieties may be encountered in a single case although they are relatively rare and many variations from the described pattern may be encountered.

Our case illustrates a mixture of the bronchiolar (bronchogenic) and the alveolar patterns with a predominance of the former. The cysts were visible grossly and histologically appear to be composed mainly of widely dilated bronchioles and alveolar spaces diffusely involving the left upper lobe of the lung. Cartilaginous plates and bronchial adnexa, except at the hilum in the region of the entering bronchus, were conspicuously absent. These histologic findings place this case under the category of diffuse congenital cystic disease of the lung. To emphasize the striking morphologic changes practically confined to the bronchio-alveolar segments, the name, congenital diffuse bronchio-alveol ectasis was suggested.

The current case can be segregated from the so-called congenital cystic adenomatoid malformation of the lung, on the basis of the absence of the following features considered characteristic of the adenomatoid lesion: 1) the polypoid formation of bronchial and bronchiolar mucosa in the cystic parenchyma; 2) the increased amounts of elastic tissue in the walls of the cystically dilated bronchial and bronchiolar structures; and 3)
alveoli lined with mucus-producing cells (mucogenic cells).

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REFERENCES
1 Stowens D: Pediatric Pathology, 2nd ed. Baltimore, The Williams & Wilkins Co., 1969
2 Chin KY, Tang MY: Congenital adenomatoid malformation of one lobe of the lung with generalized anasarca. Arch Path 48:221, 1949

Combined Aortic and Pulmonic Valvular Stenosis with an Ostium Primum Atrial Septal Defect*

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The fifth reported case of combined aortic and pulmonic valvular stenosis (the first known case with ostium primum atrial septal defect) had clinical findings suggestive of the diagnosis. Combined valvular stenosis is important to recognize clinically and confirm by catheterization because surgery of one of the lesions resulted in a fatal outcome in the previously reported cases.

Although aortic and pulmonic valvular stenosis are among the most common congenital heart defects, the combination of both aortic and pulmonic valvular stenosis in the same patient surprisingly appears to be very uncommon. This report describes the clinical findings in the fifth reported case of combined aortic and pulmonic valvular stenosis and the first known case of combined valvular stenosis with an ostium primum atrial septal defect. Accurate diagnosis of combined valvular stenosis is imperative prior to consideration of surgical correction.

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CASE REPORT

A four-year-old boy had many episodes of respiratory infections and easy fatigability. A murmur was first noted at six weeks of age. There was a family history of murmur in his maternal grandfather and a paternal uncle.

Physical examination revealed a hyperactive boy with very little scalp hair, prominence of the frontal areas of the skull and some occipital flattening. The carotid pulse had a definite shudder. A lift was palpable along the left sternal border and at the apex. There was a striking thrill in the suprasternal notch and at both the right and left second intercostal spaces, an ejection click along the lower left sternal border, and fixed splitting of the second heart sound. A grade IV/VI long, rough systolic murmur was audible at the right second intercostal space and a similar murmur at the left second intercostal space (Fig 1A and B). The murmurs were transmitted both into the neck and to the upper back.

Electrocardiogram (Fig 2) showed left axis deviation and was suggestive of biventricular hypertrophy. A vectorcardiogram confirmed that there was counterclockwise rotation in the frontal plane.

Roentgenograms of the chest showed a slightly enlarged left ventricle, a prominent ascending aorta and increased pulmonary vascularity.

Right and left heart catheterization: The catheter passed from the right to the left atrium through a low-lying atrial septal defect with the same mean pressure in both atria. Cardiogreen dye curves and oxygen saturation data revealed

FIGURE 1A (upper). Phonocardiogram in right second intercostal space with carotid tracing. 1B (lower). Phonocardiogram in left second intercostal space with carotid tracing. Fixed splitting of second sound, systolic ejection murmurs in both right and left second intercostal space, carotid shudder.