Cystic Pulmonary Cirrhosis (Bronchiolar Emphysema)

(Muscular Cirrhosis of the Lungs)*

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Progressive and relentless respiratory distress associated with x-ray film demonstration of diffuse pulmonary infiltration and fibrosis may be related to a number of disease entities. In the differential diagnosis, clinicians consider a large number of conditions, only a few of which are herein listed: malignant lymphoma, miliary tuberculosis, fungus disease, pneumoconiosis, collagen disease, eosinophilic granuloma, tuberculosis, sarcoidosis, Hamman-Rich syndrome, reticuloendotheliosis, von Wegener’s disease, lymphangitic carcinomatosis and berylliosis.

A rare cause of severe progressive pulmonary insufficiency has been reported under a variety of names referring either to the gross appearance of the lungs in an advanced stage of the disease or to the microscopic changes noted in necropsy material: muscular cirrhosis of the lungs,1,3 cystic emphysema with interstitial sclerosis,3 pulmonary muscular hyperplasia,4 bronchiolitis,5 and cystic cirrhosis of the lungs.6 While the current acceptable designation is bronchiolar emphysema, since it relates the pathogenesis to emphysema and seems to identify the main site of the morphologic changes, we prefer the designation “cystic pulmonary cirrhosis,” as it is a descriptive term that best corresponds to the micropathology. Because of its rarity, this case is being presented.

Case Report

The patient, a 40-year-old Negro woman, was allegedly well until 1957 when she developed anterior chest pain, orthopnea and exertional dyspnea. An electrocardiogram at that time was reported as showing evidence of coronary insufficiency. The symptoms continued and were in time associated with intermittent dysphagia, especially for solid food, weakness, fatigability, and peripheral edema. Despite digitalis and diuretic therapy, she became progressively worse and in addition developed a persistent, non-productive cough.

She was first seen in the Mount Sinai Hospital in February, 1959 with severe respiratory distress and marked hypotension. Norepinephrine by intravenous drip was needed to stabilize the blood pressure. Other pertinent findings included fever (101°F. orally), mucus membrane pallor, engorged neck veins, tachypnea and grunting respiration without cyanosis. Physical findings at first indicated right-sided pleural effusion and on thoracentesis, hemorrhagic exudate was aspirated. Subsequent findings were dullness on percussion over the right lower lung posteriorly with distant bronchial breath sounds, bronchophony, whispered pectoriloquy and crepitant rales. Some crepitant rales were also heard over the left base posteriorly. Similar physical findings of a consolidating lesion with a patent bronchus were heard over the right infraclavicular area and right base anteriorly. A pleural friction was audible in the right anterolateral thorax. Percussion revealed the left heart border 12 cm. from the mid-sternal line in the sixth intercostal space. The right border was obscured by the pulmonary changes. Diffuse pulsations were noted over the entire precordium. No murmurs or thrills were found, although a third heart sound was heard at the left sternal border equidistant between the first and second heart sounds. The apical rate was 100/ min. and regular. The second pulmonic sound was accentuated. The liver was palpated 5-6 cm. below the right costal margin and was tender both to palpation and percussion. The spleen was not palpated and the remainder of the physical examination was unrevealing.

Skin tests for blastomycosis, histoplasmosis and coccidioidomycosis were negative; that for tuberculosis (O.T. 1:1000) was positive. An electrocardiogram showed sinus tachycardia, right heart strain and myocardial ischemia. Chest x-ray film examination following thoracentesis revealed cardiogasty, increased bronchovascular markings in the left lower lung field and a partial right-sided pneumothorax attributed to the introduction of air following thoracentesis. An x-ray film of the chest taken ten months previously was re-

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viewed and showed diffuse, disseminated, fine granular densities.

Laboratory tests revealed persistent anemia. The leukocyte count ranged from 16,500 to 30,500 with 80 to 88 per cent polymorphonuclear leukocytes. All peripheral smears showed moderate anisocytosis, hypochromicity, macrocytosis, target cells and adequate platelets. Urinalyses were negative. Serologic tests and blood cultures were negative, as were tests for LE cells. Sputum concentrations and cultures were negative for acid-fast bacilli; in one culture Klebsiella was the predominant organism. Blood urea nitrogen values were in sequence 49, 17 and 86 mg. per cent with creatinine values of 3.7, 3.6 and 3.4 mg. per cent. A compensated acidosis was evident with a CO₂ capacity of 18 m.Eq. and blood pH 7.42. Sodium, chloride, potassium, calcium, phosphorus and alkaline phosphatase determinations were normal, but a bromsulfalein test revealed 76 per cent retention in 45 minutes. The serum protein was 6.3 gm. with albumin 2.8 gm. and globulin 3.5 gm.; protein electrophoresis showed a hypergammaglobulinemia (40.7 per cent). The sedimentation rate was normal; the C-reactive protein, 4+. Despite all therapy, chest pain and dyspnea continued unabated and she died in severe respiratory distress on the eighth hospital day.

Only the pertinent postmortem findings, gross and microscopic, are presented: each lung occupied about 4/5 of its respective pleural cavity. The left lung weighed 420 gm., the right 440 gm. The pleural surfaces were uniformly studded with numerous small projections closely resembling the surface pattern of the liver in Laennec’s cirrhosis (Fig. 1). There was a slight decrease in crepitation of all lung tissue. The cut surfaces of all lobes revealed replacement of the normal architecture by a multitude of tiny air-containing cysts, measuring from 1-4 mm. in diameter, diffusely scattered throughout the entire pulmonary substance (Fig. 2). These cysts were somewhat larger near the pleural margins and thus accounted for the projections seen on the external surfaces. Towards the hilum this pattern was somewhat less conspicuous and towards the per-

**Figure 1:** The pleural surfaces studded with numerous small projections closely resembling the surface pattern of the liver in Laennec’s cirrhosis.

**Figure 2:** Normal architecture of the lung replaced by a multitude of air-containing cysts.
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Thromboembolic occlusion of the smaller pulmonary artery branches was noted in the right middle lobe and lower lobes. Tracheobronchial lymph nodes were moderately enlarged.

Microscopically, the essential changes were alternating areas of distended cystic spaces and irregular but markedly fibrous thickening of the intervening tissue (Fig. 3). Parts of many of the cystic spaces were lined by cuboidal or columnar epithelial cells and some spaces were actually continuous with the terminal bronchioles, indicating that most of these cystic spaces were respiratory or alveolar ducts rather than alveolar sacs (Fig. 4). Some alveolar spaces were relatively well preserved. Elastic Van Gieson stain revealed irregularly aggregated, frayed and fragmented collagenous and elastic fibers. Purulent and organizing exudate, as well as small loculated abscesses, were found in some areas. In several sections emboli were noted in secondary or tertiary branches of pulmonary arteries without appreciable inflammatory reaction in the walls of the vessels.

The heart revealed a markedly dilated and hypertrophied right ventricle, measuring 8 mm. in thickness. Marked widespread fibrous thickening was found in the endocardium of the right ventricle. The coronary arteries were patent with minimal atherosclerotic changes. The liver showed marked congestion, both chronic and acute.

The anatomic diagnoses were: bilateral bronchiolar emphysema (so-called muscular cirrhosis of the lungs), superimposed right-sided bronchopneumonia, multiple thromboembolic occlusions of the small branches of the pulmonary artery, and cor pulmonale.

**Comment**

In an extensive review of the literature, Siebert and Fisher were able to document only ten authenticated cases and they added two of their own. Of the ten cases, only two had been recorded in the American literature. Since then, another case was presented at the Clinical-Pathological Conference, Mount Sinai Hospital, New York. The term “muscular cirrhosis of the lungs” was introduced by von Buhl in 1872. However, the first detailed report was made by Rindfleisch in 1897 who called this condition “cystic cirrhosis of the lungs.” Whatever the terminology, the outstanding clinical features are progressive dyspnea, chronic non-productive cough, cor pulmonale and congestive failure. Grossly the lungs in the well-established case resemble Laennec’s cirrhosis. The cut surface pre-
sents numerous minute cysts, measuring 1-3 mm. in diameter, surrounded by dense poorly aerated lung tissue. Histologically, the characteristic features are marked dilatation of the respiratory bronchioles with hypertrophy of the muscular elements (Fig. 5).

In their detailed morphologic study, Seibert and Fisher6 point out that hypertrophic emphysema is a diffuse disease characterized by acquired ectasis of the respiratory units with segmental enlargement and dilatation, focal elastic tissue damage at the site of the dilatation, and muscular hypertrophy. Two varieties of hypertrophic emphysema are recognized. When the process affects only the proximal or bronchiolar segments of the respiratory unit, this disease is called bronchiolar emphysema; when the distal or ductal segment is involved, it is called vesicular emphysema.

Invariably diagnostic procedures are directed toward definition and clarification of the disseminated pulmonary infiltrate evident on x-ray film. This patient was too acutely distressed for physiologic determinants including pulmonary function tests.

Although physiologic and structural changes can be elucidated by such function studies, they do not necessarily aid in defining etiology. We believe that thoracotomy and open lung biopsy, a well-tried and low-morbidity operative procedure, should be the eventual diagnostic tool of disciplined chest investigation. Obviously, this would be the most rewarding and revealing means of identification of occult pulmonary processes.

BIBLIOGRAPHY

EFFECTS OF SURGICAL PNEUMOTHORAX

The oxygen saturation, carbon dioxide tension and pH of arterial blood were measured in patients undergoing intrathoracic operations. These data indicate that partial and complete collapse of the operated lung caused a ventilation-perfusion abnormality so that blood flowing through the deflated lung could not adequately transfer gases.

Under the conditions of this study, there was a small but statistically significant difference between partial and complete collapse of the lung on arterial oxygen, carbon dioxide and pH. These biochemical parameters were not significantly influenced by the duration of the collapse of the lung.

Hypercventilation effectively minimized accumulation of carbon dioxide. With the technique used in this study, the carbon dioxide content and the pH of the blood remained within normal limits throughout the period of investigation. Hyperventilation with mixtures containing 25 per cent oxygen was not effective in eliminating oxygen desaturation of hemoglobin. However, the magnitude of desaturation was significantly decreased by hyperventilating with 100 per cent oxygen.

In four patients with extensive disease of the operated lung and minimal or no disease of the contralateral lung, arterial oxygen desaturation was not observed.