Unilateral Hyperlucent Lung with Polycythemia and Cor Pulmonale*

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A patient with unilateral hyperlucent lung manifested severe abnormalities of pulmonary function. Hypoxemia, hypercapnia, polycythemia, and decompensated cor pulmonale occurred. It is postulated that these changes are secondary to complicating diffuse chronic bronchitis.

Seventeen years ago, Swyer and James1 and MacLeod2 described an uncommon syndrome associated with increased transradiancy of one lung. Since then, the clinical, physiologic, radiologic and pathologic aspects of this entity have been described.3-5

Severe impairment of pulmonary function, secondary polycythemia, pulmonary arterial hypertension and decompensated cor pulmonale have rarely been present in patients with this disorder.6 Recently, we had the opportunity to study a case with these features.

CASE REPORT

The patient was a 48-year-old Negro man who was hospitalized with a two year history of increasing exertional dyspnea, orthopnea, and occasional paroxysmal nocturnal dyspnea in addition to infrequent anterior chest pain unrelated to activity. There was a three-year history of cough productive of moderate amounts of mucoid sputum.

About one year prior to admission, the patient began to have worsening of the above symptoms in addition to intermittent ankle edema which was partly controlled with diuretic administration. Symptoms worsened during the two weeks before hospitalization.

The patient smoked 4 packages of cigarettes per day for 20 years. At the age of five years, the patient had pertussis. There were multiple episodes of "pneumonia" during childhood. He recalled being unable to "keep up" with other children because of shortness of breath with exertion. There was no history of industrial dust exposure. The remainder of the history was noncontributory.

On examination, the patient was well developed, moderately obese, in mild respiratory distress and non-cyanotic. The blood pressure was 120/90, heart rate 100, respiratory rate 30. The cardiac apex was palpated in the fifth left interspace just to the left of the midclavicular line; S3 and S4 were heard at the left sternal border, and there were no thrills or murmurs. There was diminished expansion of the thorax, especially on the right side, and normal resonance to percussion bilaterally. Breath sounds were diminished in intensity over the right lung and scattered rhonchi, wheezes and basal rales were heard on both sides. Neck veins were distended, the liver slightly enlarged and the ankles edematous.

Hemoglobin was 21.0 gm, hematocrit 67 percent, reticulocytes 2.9 percent, platelets 214,000, white blood cells 10,050 with a normal differential. There were slight anisocytosis, hyperchromia and toxic granulation. The bone marrow was normocellular and contained increased iron deposits. The total blood volume was normal. Following repeated phlebotomies totaling 1500 ml, the hemoglobin was 14.6 gm, hematocrit 46 percent, and evidence of congestive heart failure subsided.

Serum protein electrophoresis showed a gamma globulin of 2.3 gm per 100 ml. The CO2 content was 38 mEq/L, the chlorides were 89 mEq/L. All other blood chemistries were normal.

Sputum cultures produced no pathogens. Skin tests with tuberculin and fungal antigens gave negative results.

The electrocardiogram revealed findings interpreted as right atrial and right ventricular hypertrophy.

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FIGURE 1 A and B. Inspiration and expiration roentgenograms. Note the hyperlucency of the right lung, persistent during expiration. Note the small right hilar shadow and the poor excursion of the right hemidiaphragm and rib cage.
Chest roentgenograms (Fig 1) showed marked prominence of the main and left pulmonary arteries and increased transradiancy of the right lung. Fluoroscopic examination, and a comparison of the films in full inspiration and forced expiration, revealed normal motion of the hemidiaphragm on the left and no motion on the right. There was a shift of the mediastinum to the left with forced expiration.

Bronchoscopy was interpreted as revealing diffuse bronchitis with no obstruction in the major branches. Biopsy confirmed the diagnosis of chronic bronchitis.

Bronchography (Fig 2) demonstrated normal branching of the major bronchi bilaterally. However, there was lack of filling and bud-like pooling of many of the distal airways throughout the right lung.

A lung scan (Fig 3) failed to reveal perfusion of the right side. Pulmonary arteriography (Fig 4) showed marked diminution in size and number of vessels in the right lung.

Pulmonary function studies, including differential bronchospirometry, arterial blood gases and pulmonary artery pressures are summarized in Table 1. These showed decreased lung volume, decreased expiratory flow, impaired diffusing capacity, hypoxemia, right-to-left shunting and a moderate degree of hypercapnia. The moderately elevated pulmonary artery pressure did not change significantly following balloon occlusion of the right pulmonary artery. Bronchospirometry demonstrated functional preponderance of the left lung.

**DISCUSSION**

The syndrome of increased unilateral or unilobar transradiancy has been well described in the literature. This entity is characterized by childhood respiratory infection and, in most cases, a benign clinical course. Roentgenographically there are persistent increased transradiancy of one or more lobes, a small pulmonary artery shadow on the diseased side, and oligemia of the affected parenchyma. The involved area manifests either normal or decreased volume. Air-trapping and mediastinal shift can usually be demonstrated. The bronchogram reveals poor peripheral filling, dilation, irregular endings, pooling, or any combination of these findings. The absence of obstruction of the large, proximal bronchi helps differentiate these patients from those with anomalous development of the bronchial tree. Decreased vascular perfusion is observed with standard chest film, tomograms, pulmonary arteriograms and lung scanning. These features were demonstrated in the case described in this report.

The pathologic lesion in six resected specimens, studied in detail by Reid, is a patchy bronchiolitis and bronchitis obliterans, with emphysema resulting from hypoplasia. The small arteries are fewer than normal in number, the capillary bed is reduced in volume and
alveolar development is impaired; however, the number of bronchial branches is normal. Since alveolar development is usually complete by the age of eight years, the lesions are probably caused by bronchial obliteration secondary to infection prior to this age.

Physiologic abnormalities are variable, but there is usually only mild to moderate ventilatory impairment and mild arterial hypoxemia in most patients with localized increased lung transradiance. Differential bronchospiratory studies support the clinical and roentgenographic data. Diminished ventilation and little or no oxygen uptake or carbon dioxide excretion are found in the affected lobe or side.

In the patient described in this report, hypoxemia, hypercapnia, and polycythemia were present, with ensuing pulmonary hypertension and right-sided heart failure. The pulmonary hypertension at rest and exercise can be ascribed to the existence of the blood gas abnormality in the presence of a restricted pulmonary vascular bed. The failure of the pulmonary artery pressure to rise with right-sided balloon occlusion reflects the already markedly deficient right pulmonary circulation.

The clinical, radiographic and spirometric manifestations encountered in the case reported herein conform to the pattern previously described. The additional features mentioned in the preceding paragraph are rarely seen in the unilateral hyperlucent lung syndrome. Reid described two such patients. In her patients, and in the one described in this report, it is probable that an obliterative bronchiolitis early in life produced a hypoplastic lung with increased transradiancy of one side. Function was maintained at or near normal levels in the uninvolved areas until the development of diffuse chronic bronchitis in adult life. It is postulated that the resulting airway obstruction and ventilation-perfusion abnormalities led to the derangements of gas exchange, and to polycythemia, pulmonary hypertension and heart failure.

**References**


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**Abdominal Sarcoidosis with Ascites**

A. Joel Papowitz, M.D. and John K. H. Li, M.D.

A patient with sarcoidosis limited to the abdomen and productive of massive ascites is reported. The patient presented in her last month of pregnancy with a markedly distended abdomen. Laparotomy was performed when abdominal swelling did not subside after delivery of a normal infant. Pathologic findings indicated active sarcoidosis. This is the third such case reported in the literature and the only one further substantiated with a positive Kveim test.

As ascites as a manifestation of sarcoidosis is a distinct rarity. When present, it is usually the result of either congestive heart failure or portal hypertension, both of which are the end stages of severe organ involvement with sarcoidosis. We have been treating a patient