Recurrent Spontaneous Pneumothoraces in Systemic Lupus Erythematosus

Laximidas A. Sawkar, M.D.** and Herman F. Easom, M.D.*

A 27-year-old Negro woman, with systemic lupus erythematosus had multiple episodes of spontaneous pneumothoraces. The pulmonary parenchymal cystic changes and rupture of the subpleural cysts which lead to recurrent spontaneous pneumothoraces were found to be secondary to systemic lupus erythematosus.

The pleural and parenchymal pulmonary changes in collagen diseases have been described extensively in the literature. Recurrent spontaneous pneumothoraces secondary to these changes have been described in histiocytosis X, scleroderma, chronic interstitial fibrosis, tuberous sclerosis, Marfan's syndrome, and a number of other related disorders. Extensive search of the literature, however, has not revealed a case of spontaneous pneumothorax complicating systemic lupus erythematosus (SLE). In the case reported here, spontaneous pneumothorax followed rupture of a pulmonary cyst in a patient with SLE.

Case Report

A 27-year-old Negro housewife was referred to us by her family physician for treatment of spontaneous pneumothorax. Recently she had become markedly short of breath, and a chest roentgenogram (Fig 1) made at that time showed collapse of the lung and pneumothorax on the left side. She had had pleuritic chest pain and recurrent bilateral pneumonia for six months. Antibiotic therapy in the past did not relieve her symptoms. There was also a history of arthralgia and myalgia, without skin rashes during this period. One year ago the patient had congestive cardiac failure of unknown etiology.

Admission physical examination revealed a moderately wasted, dyspneic and acutely ill Negro woman. Oral temperature was 101°F; the pulse was 100/minute, and the respiratory rate 44/minute. Blood pressure was 150/90 mm Hg. The jugular neck veins were prominent at 45° angle. There was prebital pitting edema. Cardiac auscultation revealed a p-
volvement of the multiple organs. The results of these investigations are as follows: hematocrit 35 vol percent; total leukocytes count 4500 per cu mm, with 61 percent segmented cells, 28 percent lymphocytes, 5 percent monocytes, 5 percent eosinophils, and 1 percent basophils. The platelet count was 140,000 per cu mm. Urine contained protein (1 plus) and microscopic examination was unremarkable. Blood urea nitrogen (BUN) rose to 50 mg per 100 ml and urinary creatinine clearance fell to 50 ml per minute while the patient was in the hospital. A lupus-cells preparation test showed numerous typical lupus cells. Latex fixation titer was less than 1:20.

Diagnostic thoracentesis on the right side revealed the pleural fluid to be exudative in nature, but otherwise negative. A chest roentgenogram made after the thoracentesis (Fig 2) exposed plate-like infiltrates and a few scattered small cysts at bases. Because the patient did not cooperate, pulmonary function studies could not be performed. An electrocardiogram recorded the changes compatible with the diagnosis of myocarditis. Echocardiogram ruled out pericardial effusion.

The diagnosis of SLE was made and she was treated with 60 mg of prednisone daily. After slow digitalization, she was maintained on digoxin 0.25 mg daily. Subsequently, the patient had complete relief from symptoms. The BUN, urinary creatinine clearance, echocardiogram and chest roentgenogram (Fig 3) all showed marked improvement. As she had developed steroid-induced psychosis, prednisone was reduced to 20 mg daily after two weeks' therapy. At present she has been maintained on this dose.

**DISCUSSION**

The pulmonary complications of collagen diseases have been described by Divertie and others. The most conspicuous radiologic manifestations of such diseases are, in the early stages, micronodular infiltrates, basal pneumonia, and pleural effusion. Basal cysts and honeycombing or fibrosis, seen in the advanced stages, are suggestive of permanent damage to the pulmonary parenchyma. Another roentgenographic finding said to be characteristic of SLE is progressive pulmonary parenchymal loss. Although no single case shows all of the changes described above, one or more of these signs are present in all patients with disorders of the connective tissue.

Shuford and his colleagues have described three principal aspects of the pathologic process exhibiting the tissue changes in the collagen group of diseases. Inflammatory reaction, degeneration of the collagen ground substance, and proliferation of collagen fibers, in alveolar walls and interalveolar septa have been described to be the main features of this process. The cysts, and honeycombings which are usually found in the lower half of the pulmonary parenchyma, are formed from degenerated alveolar walls that have ruptured and merged. The rupture of a subpleural cyst results in spontaneous pneumothorax.

Once the diagnosis of a collagen disease has been made, pneumothorax should be treated by parietal pleurectomy or pleural abrasion, in order to prevent further recurrences. Steroids and 6-mercaptopurines have been found beneficial in these disorders of connective tissue.

The present case is the only reported case of SLE in which roentgenograms revealed cystic changes in the pulmonary parenchyma. Treatment with steroids has helped to relieve the symptoms and apparently arrested further progress of the disease process in this patient.

**REFERENCES**


Reprint requests: Dr. Sawkar, Eastern North Carolina Sanatorium, Wilson 27893