Systemic Lupus Erythematosus Associated with Bronchiectasis*

Report of Two Cases
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Systemic Lupus Erythematosus has been described as a disease of "a variable number of visceral manifestations." Involvement of lung parenchyma and pleura have been reported frequently, but only one case of systemic lupus erythematosus presenting primarily as bronchial disease* has been published. The present report concerns two patients with systemic lupus erythematosus presenting primarily as bronchiectasis. The purpose of this paper is to report these cases and call attention to the facet of this multiple system disease which has received little notice previously.

CASE 1
A 26-year-old white woman was admitted to Presbyterian Hospital on September 28, 1961

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for pulmonary evaluation. She had had three episodes of pneumonia. She had bronchopneumonia in December, 1956 which subsided after ten days of treatment with penicillin. In 1958, she again had bronchopneumonia and was treated with penicillin for three weeks. She had lobar pneumonia and pleuritis in January, 1960. This last illness required three months for complete resolution. Since age 22, she has had episodes of acute respiratory infections every four or five months manifested by fever of 103° or 104°F., severe malaise, tightness in her chest, and a cough productive of purulent sputum. These episodes were treated with antibiotics by her family physician and they subsided in several weeks. Between these acute infections she had a chronic cough, usually dry, but occasionally productive of purulent or blood-streaked sputum. She had significant psychiatric and gynecologic problems in the recent past requiring admission for ataxia abasia in June, 1961 and dilatation and curettage of the uterus on seven occasions for functional menorrhagia or incomplete abortions. In January, 1961, she was hospitalized for evaluation of

**FIG. 1**
**FIG. 2**

**FIGURE 1:** Bronchogram, Case 2, 1949. **FIGURE 2:** Bronchogram, Case 2, 1949.
hematuria with normal cystoscopic and pyelographic findings.

When first admitted to the thoracic surgery service, she was a slender, apprehensive young woman, with moist rales at the right lung base. Routine laboratory studies, electrocardiogram and chest x-ray films were within normal limits. Bronchoscopy on October 2, 1961 revealed edematous hyperemic mucosa and markedly increased secretions. Bronchial washings were negative for tumor cells and acid-fast organisms. Bronchograms following the bronchoscopy showed early bronchiectatic change in both lower lobes. Several hours following completion of the bronchogram, her temperature rose to 102.4°F. and she had chills, malaise, increased cough with very little sputum and coarse rales at both lung bases. Chest x-ray films showed bilateral lower lobe pneumonia. She was treated with tetracycline and amphotericin B (Mysteclin), expectorants, and intermittent positive pressure breathing with isoproterenol (Isuprel). On October 6, 1961, she experienced sudden onset of severe dyspnea with shallow respirations and a very marked bright brick-red color over her entire body, most marked over the head, neck, and upper thorax. Emergency tracheostomy produced prompt relief of her acute difficulties. Adrenal corticosteroid hormones were added to her therapy and within three days she was symptom-free and had complete clearance of the pneumonitis. She had no further breathing problems or difficulty with secretions following removal of the tracheostomy tube. Additional laboratory studies showed negative tests for urinary porphyrins, porphobilinogen, five hydroxy indolacetic acid, latex tests for rheumatoid arthritis, and normal blood urea nitrogen, electrolyte and serum electrophoresis determinations. LE cell preparations were repeatedly positive. She was well when discharged on October 17, 1961. Subsequently, she was hospitalized in February and June, 1962 for uterine dilatation and curettage for incomplete abortion and menorrhagia. On her last admission, which lasted 13 days, she had such severe associated anxiety symptoms that psychiatric consultation and intravenous sodium amytal for a period of eight days were required.

CASE 2

A 46-year-old white woman was admitted to Charlotte Memorial Hospital for the 35th and final time on March 6, 1962. Conditions necessitating hospitalization since 1920 included: tonsillectomy, right oophorectomy and salpingectomy, appendectomy, peri-rectal abscess, ventral hernia repair, hematuria, functional menorrhagia,
cystitis, metronomorrhagia, submucous resection, early menopausal symptoms, sinusitis and otitis media, gastroenteritis, ethmoid and maxillary sinusitis, cephalgia, bilateral dorsal sympathectomy because of Raynaud's phenomenon with superficial gangrene of the left index finger, and acute meperidine (Demerol) intoxication. Her first admission for a pulmonary problem was in 1936 when tuberculosis was suspected on the basis of a chest x-ray film, but could not be demonstrated bacteriologically. In 1948, because of chronic productive cough bronchoscopy was performed with negative findings except for hyperemic mucosa and purulent secretions. The cough productive of foul purulent sputum persisted. Bronchoscopy, repeated in August, 1947, showed most of the secretions came from the left lower lobe. Bronchograms showed early bronchiectasis of the right and left lower lobes. In April, 1948, the persistence of this productive cough necessitated repeat bronchoscopy and bronchograms which showed the maximal bronchiectatic involvement to be in the middle lobe with little change in the previous findings in both lower lobes. Middle lobe lobectomy was performed with subsequent uneventful recovery. Microscopic study of the specimen showed fusiform bronchiectasis. In September, 1949, she had profuse hemoptysis, as well as severe productive cough with chills and fever. Hemorrhage was localized to the left lower lobe at bronchoscopy. Lower left lobectomy was performed following which she was extremely dyspneic, cyanotic, had a high fever and continued hemoptysis for more than one week before recovering. The pathologist reported marked chronic non-specific bronchitis with fusiform bronchiectasis. In August, 1953, she had a pulmonary hemorrhage and acute pneumonitis in the right lower lobe. In November, 1956, fever, acute arthritis of hands and fingers and epigastric pain were relieved only by prednisone (Meticorten). LE cell preparations were negative at this time. In December, 1956, after discontinuing the Meticorten the chills and fever recurred, accompanied by right pleuritic pain, severe headache and emesis. These symptoms cleared with Meticorten therapy. On this and subsequent admission in January, 1957, LE cells were abundant. She was bronchosced in September, 1957, because of recurrent hemoptysis which was seen coming from the right middle lobe stump and which cleared on hydrocortisone therapy. In October, 1957, she had bilateral bronchial pneumonia with severe headache, cervical myalgia, fever of 103°F., all of which responded to antibiotics and hydrocortisone. BUN determination was 44 mg. per cent on this admission. In November, 1959, left upper and lower lobar pneumonia responded to administration of adrenal corticosteroids and anti-biotics. Prior to her final admission on March 6, 1962, she had had fever for 24 hours with progressively increasing dyspnea during that time. Approximately one hour prior to admission, she became delirious and this progressed rapidly to unconsciousness. On examination, she had a fever of 106°F., was cyanotic, dyspneic, tachyneic, and gasping for breath. Rales and bronchial breathing were heard. She progressively deteriorated and expired six hours after admission. Necropsy was performed. Acute and chronic bronchitis and bronchiectasis, but no specific lesions of lupus were seen in the lungs. Kidneys, myocardium, and liver were normal, and only the spleen showed scattered arterial changes suggestive of lupus.

**DISCUSSION**

In 1939, Reifenstein, et al.* reviewed the post mortem findings in 17 cases published in the literature and three of their personal cases of systemic lupus erythematosus. Pneumonia, lobar and bronchial, was found in 15 cases, pleuritis with effusion in two cases; and pulmonary congestion with scattered areas of atelectasis in four cases. Dubois* reported an incidence of pleuritis in 59.5 per cent and effusion in 55 per cent of his series of 62 cases. He did not mention pulmonary parenchymal or bronchial involvement in his report. Israel* reviewed the pulmonary manifestations of disseminated lupus erythematosus and found pulmonary or pleural disease in 20 of 22 cases. He felt that the pneumonias were primarily bacterial in origin and that specific lupus erythematosus pneumonitis was relatively infrequent. It was his opinion "that some alteration of bronchial or pulmonary structure was likewise responsible for the extraordinary frequency of bacterial pneumonias in disseminated lupus erythematosus." He did not describe any primary disease of major bronchi. Harvey, et al.,* in a comprehensive review of systemic lupus erythematosus, listed 20 cases of definite lupus pneumonitis characterized by "hyaline membranes in the alveoli, focal necrosis of alveolar walls with capillary thrombi, areas of organized interstitial pneumonia and hemorrhage, and metaplasia of bronchiolar epithelium." He also observed pulmonary tuberculosis, pneumo-
coccic lobar pneumonia, lung abscess, aspiration pneumonitis, lobular pneumonia. Pleurisy was observed in 60 of his 138 cases. Again, no mention was made of the involvement of the tracheobronchial tree in association with systemic lupus. Cassidy and Kennedy reported a case of systemic lupus erythematosus presenting as bronchiectasis in a 16-year-old boy. His disease was characterized by a profuse foul sputum with x-ray findings suggestive of an acute bronchopneumonia which did not respond to any antibiotics, but responded very dramatically to the adrenal corticosteroids. The lupus appears documented by the persistent findings of LE cells over a period of several years and a positive LE precipitation test. Unfortunately, however, bronchiectasis was not demonstrated by either bronchograms or microscopic study of a surgical specimen.

The diagnosis of systemic lupus erythematosus was established in the present two cases by the finding of LE cells repeatedly and by the clinical courses. Multiple systems were involved with exacerbations and remissions and a favorable response to adrenal corticosteroids usually occurred. Bronchiectasis was demonstrated by bronchography in both cases. In case 2, both surgical and necropsy material were studied and bronchietastic changes seen. Bronchial arteries and arterioles were scrutinized for stigmata of lupus, but none was seen. According to the concept of Israel, the bronchiectasis was probably secondary to some alteration of bronchial structure, but the nature of this alteration could not be ascertained by usual microscopic techniques in the present case.

Systemic lupus erythematosus is known to have pleural manifestations—pleuritis, effusion; specific lung parenchymal lesions—lupus pneumonitis; and non-specific lung disease—tuberculosis, lobar and bronchopneumonia, abscess, and aspiration pneumonitis. As demonstrated in these two cases and that of Cassidy and Kennedy, bronchial disease, i.e., bronchiectasis, should be added to the pulmonary manifestations of systemic lupus erythematosus.

REFERENCES


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INHALATION THERAPY

The efficiency of humidifying devices designed to moisten (dry) oxygen during the course of inhalation therapy is limited by the development of the reduced temperatures of the water in the reservoirs of these devices. Heating elements are available that permit the maintenance of water-reservoir temperatures sufficient to provide a supply of water vapor at or near body temperature after its passage through a standard length of large-bore (1.8 cm. internal diameter) tubing. The increased water vapor content of the gas issuing from a heated nebulizer requires tubing of a sufficient diameter so that water condensation does not block or impede the flow of gas to the patient.