Case report

Lupus cor pulmonale with electron microscope and immunofluorescent antibody studies

P. P. B. YEO AND R. SINNIAH
From the Departments of Medicine and Pathology, University of Singapore, Outram Road General Hospital, Singapore, 3

Yeo, P. P. B., and Sinniah, R. (1975). Annals of the Rheumatic Diseases, 34, 457–460. Lupus cor pulmonale with electron microscope and immunofluorescent antibody studies. A case of systemic lupus erythematosus with an unusual complication of pulmonary hypertension leading to cor pulmonale is reported. Lung biopsy showed an interstitial pneumonitis with pulmonary vascular narrowing causing pulmonary hypertension. These changes appeared to be immunologically mediated by the utilization of IgG and C3. Microtubular virus-like particles were present in the endothelial cell cytoplasm of both lung and kidney.

Pleopulmonary involvement in systemic lupus erythematosus (SLE) is well known (Dubois, 1974). Arteritis of the smaller pulmonary vessels leading to pulmonary hypertension is a recognized, albeit rare, complication of SLE (Wood, 1952). A case is recorded here of a patient with SLE who developed severe pulmonary hypertension in less than 3 years during the course of her disease, culminating in cor pulmonale.

FIG. 1 Normal chest x-ray, taken on August 30, 1971, showed no cardiac enlargement or lung pathology

Accepted for publication March 3, 1975.
Clinical history

A 23-year-old Chinese housewife presented in June 1974 with a 7-month history of malaise and fatigue, followed by palpitations with exertional dyspnoea a month before admission. There was associated progressive bilateral ankle swelling and abdominal distension. In August 1971, the patient was hospitalized at another hospital with the nephrotic syndrome. Tests for rheumatoid and antinuclear factors were positive but LE cells were repeatedly negative. The patient was treated with steroids and diuretics but failed to attend for follow-up in December 1971. In the cardiovascular system there was then no detectable abnormality and chest x-ray (Fig. 1) was normal. Clinical examination on her present admission showed ankle and sacral oedema; blood pressure 110/80 mmHg; regular pulse rate 130/min; jugular venous pressure 2 cm with prominent a waves. Left parasternal heave with atrial gallop and accentuated pulmonary second sound; a soft systolic murmur in the pulmonary area, together with an early diastolic murmur. Lungs were clinically clear. The liver was palpable 1 cm below the costal margin.

Investigations

Haemoglobin was 11.1 g/dl; mean corpuscular haemoglobin count 33 g/dl (33%); red blood cells normocytic,
cytoplasm. The patient's diuretics, digitalization, were deposited were seen, proliferation of virus-like microtubular, were there steroids. In both lower and midzones bilaterally, consistent with interstitial fibrosis. There was cardiac enlargement with a large main pulmonary artery trunk. ECG showed sinus tachycardia and right ventricular hypertrophy with strain pattern. Respiratory function tests were those of restrictive lung disease with impaired diffusing capacity. Lung biopsy showed the features of interstitial pneumonitis with narrowing of alveolar septal vessels. There was muscular medial hypertrophy and concentric connective tissue proliferation of the larger vessels. On electron microscope examination clumps of microtubular virus-like particles were seen, mainly in a subnuclear position (Fig. 3) in the endothelial cell cytoplasm of blood vessels. IgG and C3 were deposited in the vascular endothelium and walls of alveolar septae (Fig. 4). Renal biopsy showed similar microtubular, virus-like particles in the endothelial cell cytoplasm.

Treatment and progress
The patient's lupus cor pulmonale improved with digitalization, diuretics, and steroids. Radiologically there was significant reduction in cardiac size (15 cm–13.3 cm over 6 months). Basal lung shadows were less prominent. The patient is now on maintenance therapy.

Discussion
There are few reports of pulmonary hypertension in SLE giving rise to cor pulmonale (Aitchison and Williams, 1956; Larson 1961). The patient described here developed severe pulmonary hypertension during the 3-year interim period from when she first had the nephrotic syndrome until her present admission with cardiac failure. The pulmonary pathology of interstitial pneumonitis and vasculopathy causing cor pulmonale were apparently mediated through the utilization of IgG and C3. It may be that circulating antigen–antibody complexes formed during the course of SLE were deposited in the lungs. Microtubular virus-like particles have been described in lupus nephritis (Györkey, Sinkovics, Min, and Györkey, 1972), but there is no report in the literature of the presence of these structures in the lungs of patients with SLE. The nature of these structures has not been resolved, though suggestions have been put forward that they may be of viral origin (Györkey and others, 1972) while Schaff, Barry, and Grimeley (1973) could find no evidence for a relationship to similarly sized cores of a known paramyxovirus.

We thank Professor P. K. Wong for encouragement; Mr. N. C. Tan for the lung biopsy; T. C. Tan, S. L. Fung, H. Y. Heng, C. Y. Chia, J. Chia, and C. K. Ow for technical assistance; and Miss L. Quek for typing the manuscript.
References


SCHAFF, Z., BARRY, D. W., and GRIMLEY, P. M. (1973) *Lab. Invest.*, 29, 577 (Cytochemistry of tubuloreticular structures in lymphocytes from patients with systemic lupus erythematosus and in cultured human lymphoid cells: comparison to a paramyxovirus)