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EDITORIAL
PULMONARY FINDINGS IN THE COLLAGEN DISEASES*
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A number of systemic diseases which have been classed together in spite of considerable diversity of manifestations were first called "diffuse collagen disease" by Klemperer and his co-workers in 1942. They applied this term to disseminated lupus erythematosus, scleroderma, and dermatomyositis. Since then rheumatoid arthritis, acute rheumatic fever, periarteritis nodosa, and thrombotic thrombocytopenic purpura have been added. Hamman-Rich syndrome, Whipple's disease, and hypersensitivity angiitis are not included, though it has been suggested that they should be.

This group of diseases involves soft parts generally and blood vessels specifically in many instances. Some mechanisms have been discovered. In the case of lupus erythematosus there is little question about the presence of abnormal proteins in the blood, disturbance of desoxyribosenucleic acid metabolism of the mesenchyme, and the L. E. cell phenomenon. In all of the group, except possibly scleroderma, there must be consideration of hypersensitivity as a pathogenetic factor.

The relationship of the histopathologic changes in these diseases is emphasized by the occurrence of findings characteristic of more than one of the disorders in a single patient; the clinical overlapping of symptom complexes is considerable. Five per cent of patients with scleroderma, for example, show glomerular lesions similar to those of lupus erythematosus. Geschickter and others report that the application of a simple irritant amine to experimental animals is capable of causing the histopathologic changes of all of the more common collagen diseases. In this experimental work, the animals presumably respond in these several fashions to a single injurious substance, which, however, may have many sites of action. Other forms of trauma, even as simple as pinching the skin, can cause the fibrinoid necrosis typical of the group.

The acute manifestations of disseminated lupus erythematosus which may appear on films of the chest include pleural effusions of any size, either unilateral or bilateral, pulmonary edema, and pneumonitis of several types with some predilection for localization in the subpleural areas (Thorell). Patients in this group are also more prone than the general population to ordinary bacterial pneumonias. These probably follow pulmonary edema. Sante and Wyatt have pointed out that edema, congestion, and hemorrhage occur in the lungs when the collagen disease involves the kidneys — the so-called "antigenic (azotemic) pneumonitis." Purnell, Baggenstoss, and Olsen described basophilic mucinous edema, interstitial pneumonitis, and alveolar hemorrhage in some of their cases. The acute pulmonary edema of lupus erythematosus is not always associated with severe cardiac or renal lesions; in this event the edema may be due to the direct action of the disease in the lungs or to protein disturbances. The pulmonary abnormalities in lupus may change quite rapidly. Fibrosis may follow acute lesions or appear without known preceding involvement; it produces a fine linear and nodular pattern, narrowed vessels, and at times cor pulmonale. This may progress to a reticular or fine cystic appearance.

Periarteritis nodosa may also show "antigenic (azotemic) pneumonitis" on the

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basis of involvement of the general body economy secondary to renal lesions. In addition, changes can result from a specific local vascular lesion in the pulmonary or bronchial arteries or both (Doub et al., Braunstein). When there is a localized arterial lesion, pulmonary infarction, frequently resulting in cavitatin, may be seen. Changing patches of pulmonary edema and hemorrhage are present and may be followed by a linear and fibrotic pattern as in lupus erythematosus. Enlargement of the lung roots is a common roentgen observation. The localized vascular lesions of periarteritis may also cause widespread arterial destruction throughout the lungs. Death usually ensues in a short time. Many lesions of Wegener's granulomatosis may for all practical purposes be considered a localized periarteritis causing an infarct; others are "granulomas" with necrotic centers.

Although in some cases of scleroderma there is a diffuse and patchy infiltrative process in the lungs, indistinguishable from the changes seen in other members of this group of diseases, there is also a typical "sclerocystic" appearance with a very fine honeycomb pattern which is thought to be characteristic. This is the result of coalescence of alveoli and formation of "broncho-alveolar cysts." There is associated marked interstitial fibrous thickening following subepithelial myxomatous change. In some of our material there has been a severe bronchiectasis in the lower lobes.

The nature of the transient densities appearing in the lungs in acute rheumatic fever is not established. Some are probably the result of congestive failure, incomplete infarction, and hemorrhage. However, a highly fatal type of pneumonia has been found in a number of autopsy studies of patients who have had extensive hyaline membrane formation, Masson bodies, and mononuclear cell infiltrates. The roentgen appearance in these cases has varied from diffuse mottling to homogeneous consolidation of large portions of both lungs.

Much has been written recently concerning the pulmonary lesions in patients with rheumatoid arthritis. Ellman and Ball and Christie reported complete autopsy study of cases of rheumatoid arthritis with pleural lesions and with lesions in the parenchyma, vessels, bronchi, and lymph nodes of the lungs. These lesions were in all stages of development and of varying degrees of severity. Small foci of necrotizing pneumonia with fibrinoid changes were followed by the development of collagenous tissue. Small necrotic nodules were formed from fibrinous pneumonia, and in one case there was progression to the formation of a massive rheumatoid nodule, 7 cm. in diameter. Fibrotic nodules and strands as well as fibrosis of vessels and bronchi were found. Gresham and Kellaway published a case of rheumatoid arthritis showing cystic changes in the subpleural areas, with reproduction of sections containing both the cystic changes and active rheumatoid nodules. They were unable to establish the origin of the cysts from the granulomatous destruction of lung. Histochemical observations of the lung lesions were similar to those of a juxta-articular rheumatoid nodule. The patient also had cardiac lesions with aortic incompetence, thickening of cusp margins, and fusion of the commissures. The valve showed fibrinoid necrosis bordered by palisading histiocytes. Similar cystic and granulomatous lesions are present in our material. Fine fibrotic lesions like the Hamman-Rich syndrome are reported by Read.
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Coal miners with rheumatoid disease have been found by Caplan to have lung lesions of a particular type, somewhat resembling massive fibrosis, but more numerous, round, and more uniformly distributed through the lungs. These lesions were observed in the presence of slight or no generalized radiographic changes of pneumoconiosis. They developed rapidly and then could remain unchanged or appear in successive waves. Sometimes they disappeared or became fibrotic after cavitation. Some calcified or became incorporated in progressive massive fibrosis. Caplan and others have described this process in a foundry worker, and it has been seen in pottery workers, sand blasters, boiler scalers, and brass and iron foundry workers. Microscopically the border of the lesion shows a rheumatoid type of inflammation with formation of collagen and variable amounts of necrosis.

Typical foci of rheumatoid nodules localized mainly to the subpleural portions of the lower lungs are described in association with asbestosis by Rickards and Barrett. They recognized 26 cases of necrobiotic foci of typical rheumatoid lesions and stated that 15 of them were associated with pneumoconiosis. These cases show the altered response of a patient with a particular collagen disease to a given type of injury.

Pulmonary lesions are frequent in lupus erythematosus, periarteritis nodosa, and scleroderma, but infrequent in rheumatic fever, rheumatoid arthritis, and dermatomyositis.

REFERENCES