Interstitial lung diseases and sarcoidosis: a historical note

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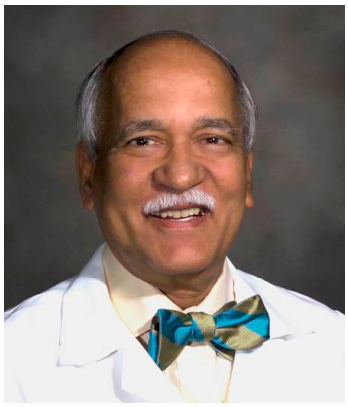

Om Prakash Sharma (MD, FRCP, DTM&H) was born in India. Currently, he is Professor of Medicine at the Keck School of Medicine at the University of Southern California, Los Angeles, USA. He is president of the WASOG. Prof. dr. Sharma has authored or edited five books, eight monographs, and written about 400 articles, short papers, editorials and reviews on the topics of sarcoidosis, tropical medicine, hypersensitivity pneumonitis, interstitial lung disease, other pulmonary disorders, and history of medicine.

The first description of an interstitial lung disease appears to have been by Bernardino Ramazzini da Capri (1633-1714). The author, in the chapter on the Diseases of Sifters, Measurers, and Handlers of Grain, of his famous book De morbis artificium diatriba, described the occurrence of dry cough, weight loss, breathing difficulty, and dropsy in these workers. 1868, Austin Flint described a nondescript lung disease that was characterized by florid inflammatory exudation, without pus, causing solidification and fibrosis of the lungs. Flint alluded that Rokitansky had observed a similar condition in which exudation had occurred into the interlobular tissue. A few years earlier, Dominic Corrigan, an Irish cardiologist, had called the similar entity cirrhosis of the lung as it was analogous, not identical, to cirrhosis of the liver. Corrigan also described the occurrence of traction bronchiectasis and Flint observed that the association of clubbing and interstitial pneumonitis/fibrosis. Wilson Fox, professor of pathological anatomy at the University College, London, recorded the microscopic changes of capillary edema; accumulation of pigmented epithelium in the alveoli; and thickening of the walls of the alveoli, and veins in lungs with interstitial pneumonitis. .

In 1944, Louis Hamman and Arnold Rich, both at the Johns Hopkins University School of Medicine, described four young patients who died of progressive dyspnea within 6 months of onset. The condition was similar to that described earlier by Flint, Corrigan, Fox, Charcot, and Osler. The term Hamman-Rich syndrome, however, became a synonym for an interstitial pneumonia of unknown cause followed by fulminating pulmonary fibrosis. It was soon apparent that the course of this new disease was not always acute, progressive, or fatal. Averil Liebow, on the basis of his extensive clinical and pathologic sciences, classified interstitial pneumonitis into five different histologic types: usual interstitial pneumonitis (UIP), desquamative interstitial pneumonia (DIP), bronchiolitis obliterans interstitial pneumonia (BIP), lymphoid interstitial pneumonia (LIP), and giant cell interstitial pneumonia (GIP). The new classification of idiopathic interstitial pneumonia includes UIP and DIP from Liebow's original classification and two new entities, acute interstitial pneumonia (AIP, or Hamman Rich syndrome) and NSIP. Bronchiolitis obliterans organizing pneumonia (BOOP) is not included because it is primarily an intra-luminal disease.

Jonathan Hutchinson, a surgeon-dermatologist, identified the first case of sarcoidosis at the King's College London. In the decades before and following the turn of the nineteenth century, several publications independently drew attention to the multisystem nature of the disease. This trend continued to the rest of the twentieth century and has persisted into the present century. Although the clinical, radiological, physiological, biochemical, and immunological aspects of the granuloma formation are well investigated, etiology of sarcoidosis needs to be uncovered by developments in the field of genomics and proteomics.