ELSEVIER
SAUNDERS

Clin Chest Med 29 (2008) xiii–xiv

CLINICS
IN CHEST
MEDICINE

Preface

Robert P. Baughman, MD Marjolein Drent, MD, PhD
Guest Editors

The fascination with sarcoidosis as a disease lies in the failure to identify the cause, its variable presentation, and its unpredictable clinical course. The features of this disease have been known for more than 100 years, which Dr. Sharma discusses in his article. While no specific answer regarding the cause of the disease has been made, we have learned a great deal about sarcoidosis.

One hypothesis is that sarcoidosis is a multi-organ disease representing a specific immune reaction that is triggered by one or more environmental agents (either living or dead). This immune response appears to be modified by genetic predisposition. While this is a fairly broad definition, one can delineate various components of this hypothesis.

From its original description, sarcoidosis has been assumed to be an infection. The granulomatous response found in samples appeared similar to a very common disease of the time, tuberculosis. As tuberculosis has become less frequent, sarcoidosis has become more clearly recognized as a separate disease. Drs. Chen and Moller summarize the work to date that examines the various possible infectious and noninfectious causes of sarcoidosis.

The immune response of sarcoidosis includes the formation of granulomas. This immune response frequently is characterized as Th-1 response. Drs. Gerke and Hunninghake analyze the immune response in terms of its similarity to other Th-1 responses (such as tuberculosis) and differences from other diseases. The variability of the outcome of sarcoidosis seems to be a reflection of the immune response, either in the initial reaction or in its subsequent modification.

Genetic predisposition has been proposed to modify almost every disease encountered by mankind. A rhinovirus that infects a group of office workers ranges from a few who have a runny nose for 2 days to the unfortunate ones who are wheezing and coughing for weeks. Sarcoidosis affects different ethnic groups in different ways, suggesting the influence of genetic background. By using a more homogenous background of patients or families who have the disease, researchers are recognizing that certain genetic factors are increase the risk of the disease, influence organ involvement, or modify the duration of disease. These issues are summarized by Dr. Müller-Quernheim and his colleagues.

One of the challenges of sarcoidosis remains its diagnosis and evaluation. Dr. Judson provides a comprehensive approach to the possible cause of sarcoidosis. He focuses on the multi-organ nature of the disease, emphasizing how this helps to diagnose the disease and make the physician

0272-5231/08/$ - see front matter
doi:10.1016/j.ccm.2008.04.001

aware of possible extra-thoracic complications of the disease. Dr. Akbar and colleagues provide examples of the information the radiologist can provide about the disease. While the focus is on lung manifestation, imaging of other organs is also highlighted. Dr. Rottoli's group summarizes markers of inflammation as measured in various tissue samples, including bronchoalveolar lavage.

Specific organ involvement is discussed in several articles. Drs. Vucinic and Jovanovic summarize pulmonary involvement in sarcoidosis. Sarcoidosis less frequently affects the heart or central nervous system; however, when these areas are affected, the consequences can be severe and long lasting. Drs. Lower and Weiss provide information about the diagnosis and management of neurosarcoidosis. Dr. Padilla's group focuses on cardiac sarcoidosis, including information about how to screen for potential cardiac disease. Dr. Knox's group highlights the presentation in the three most common extra thoracic organs: liver, skin, and eye.

For the patient who has sarcoidosis, the disease can have a devastating effect on the overall quality of life. Recent instruments for the general quality of life and specific instruments related to the effect of the disease on health have given important insights into sarcoidosis. Drs. De Vries and Drent provide a state of the art summary of this challenging area.

The treatment of sarcoidosis has to be tailored to the individual patient. As new drugs become more widely used in sarcoidosis, clinicians are left with decisions about when to use the older agents (such as prednisone and hydroxychloroquine) versus the newer drugs (such as infliximab or thalidomide). Drs. Baughman, Costabel, and du Bois provide one approach to the patient, which is based on the clinical outcome of a patient: acute, chronic, or refractory. For pulmonary patients, it has been emphasized recently that one reason for a patient failing to respond to anti-inflammatory drugs is the secondary complication of pulmonary hypertension. Dr. Culver and his group summarize the current knowledge of sarcoidosis associated pulmonary arterial hypertension.

The cause for the variable clinical outcome in sarcoidosis remains a mystery. Dr. Nagai and her colleagues discuss the differences in outcome. One can only assume that genetic background, handling of the etiologic agent by the host, and possibly treatment all have an influence on the clinical outcome.

Although there are still many questions, the practicing physician has plenty of information to diagnose and manage patients who have sarcoidosis. In this issue, we have tried to provide a ready summary of this information.

We thank Elsevier overall, and we thank Sarah Barth in particular for all of her help and encouragement throughout the process of preparing this issue.

Robert P. Baughman, MD
Professor of Medicine
Department of Medicine
University of Cincinnati Medical Center
1001 Holmes, Eden Avenue
Cincinnati, OH 45267-0565, USA

E-mail address: baughmrp@ucmail.uc.edu

Marjolein Drent, MD, PhD
Professor of Interstitial Lung Diseases
Department of Respiratory Medicine
Sarcoidosis Management Center
University Hospital Maastricht
PO Box 5800, 6202 AZ
Maastricht, The Netherlands

E-mail address: m.drent@lung.azm.nl