Best Practice & Research Clinical Rheumatology 30 (2016) 334–356

Contents lists available at ScienceDirect

Best Practice & Research Clinical Rheumatology

journal homepage: www.elsevierhealth.com/berh

9

Sarcoidosis: Rheumatology perspective

Arthur M.F. Yee, MD PhD *

Hospital for Special Surgery, Weill Cornell Medicine, 535 East 70th Street, New York, NY 10021, USA

Keywords:
Sarcoidosis
Rheumatology

A B S T R A C T

Sarcoidosis is a systemic inflammatory granulomatous disease for which rheumatologists are uniquely trained and qualified to treat. Historically, sarcoidosis has been managed within silos of medical subspecialties, but with increased appreciation of the systemic nature of this disorder and the availability of more therapeutic options, it is clear that a multidisciplinary approach, with the rheumatologist as a key component, can offer more optimal care. This manuscript reviews clinically relevant immunology and pathophysiology, diagnostic issues, management decision-making, and therapeutics in the care of patients with sarcoidosis. Issues particularly relevant to rheumatologists are highlighted. These include aiding in establishing diagnosis; recognition of disease manifestations involving bone, joint, and muscle; management of calcium metabolism and metabolic bone disease; and formulation and implementation of anti-inflammatory and immunomodulatory therapies.

Introduction

Sarcoidosis is a systemic inflammatory disorder that can potentially affect any organ system, notable for great variability in clinical presentation and clinical course. Since its first description, attributed to Norwegian dermatologist Caesar Boeck in the late 19th century, sarcoidosis remains a fascinating and enigmatic condition, which by its very nature of unpredictability can be unsettling for the afflicted patient and the treating physicians. Cases can range from asymptomatic individuals diagnosed after incidental findings of radiologic abnormalities that require no specific treatment to

* Tel.: +1 212 606 1171; fax: +1 212 774 2770.
E-mail address: yeea@hss.edu.

http://dx.doi.org/10.1016/j.berh.2016.07.001
1521-6942/

severely affected individuals with potentially life-threatening situations that demand aggressive therapy from the outset.

Historically, the care of sarcoidosis patients has understandably tended to be compartmentalized according to the specific organ systems that are affected. Hence, for example, patients with isolated cutaneous sarcoidosis will often be seen solely by dermatologists. This compartmentalization quite naturally extends into research as well, in which therapeutic trials will often be restricted to relatively narrow spectrums of patients with isolated organ system involvement such as lung, eye, or skin.

There are estimates that lung involvement is present in up to 90% of patients with sarcoidosis, and as a result, much of the important work in the understanding and treatment of this disease is credited to the efforts of the pulmonary medicine community. For example, the classic staging of sarcoidosis devised by Scadding half a century ago assesses pulmonary and intrathoracic involvement and is still widely cited today [1]. However, the Scadding staging system all but ignores extra-thoracic disease, and so today, with increasing acknowledgment of the systemic nature of sarcoidosis and the development of newer approaches to treatment, interdisciplinary approaches to the management of sarcoidosis have aptly become more standard.

From the standpoint of sarcoidosis, the production of this volume is timely. The role of the rheumatologist in the management of this disease has been less fully defined historically. However, it takes little stretch of imagination to recognize that sarcoidosis bears striking similarities to the many inflammatory diseases such as systemic lupus erythematosus (SLE) for which rheumatologists are relied on for their expertise. It was not surprising, then, that when one widely respected veteran pulmonologist was asked to give a presentation at the 2015 Annual Conference of the American Association of Sarcoidosis and Other Granulomatous Disorders on novel approaches to the treatment of sarcoidosis, he said that the first thing he did to prepare was “to talk to my colleagues over at the Division of Rheumatology.”

Preliminary data from an ongoing study of rheumatology trainees at Hospital for Special Surgery/Weill Cornell Medicine show that advice and assistance from the rheumatology community are regularly being sought in the care of patients with sarcoidosis. While this monograph offers a broad overview of the management of this disorder, special attention is given to topics that may be of particular relevance in the involvement of the rheumatologist.

Immunology and pathogenesis

The pathologic signature of sarcoidosis is noncaseating granulomatous inflammation (Fig. 1). The compact and highly organized sarcoid granuloma is formed by lymphocytes, macrophages,

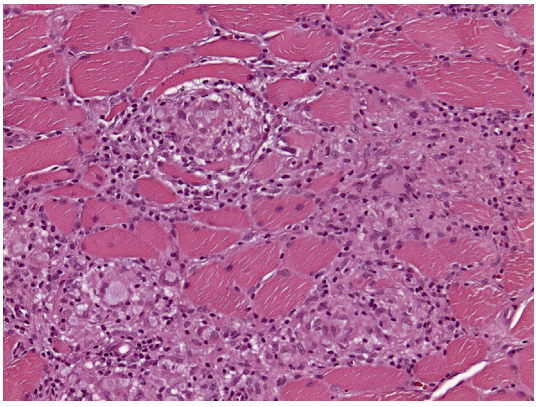

Fig. 1. Muscle biopsy of a patient with sarcoid myositis demonstrates characteristic granulomas containing macrophages and multinucleated giant cells within the endomyseal connective tissue, adjacent to muscle fibers. A thin ring of lymphocytes can be seen around the granuloma at the upper left part of the photomicrograph.