Review

SARCOIDOSIS ASSOCIATED WITH RHEUMATIC AUTOIMMUNE DISEASE

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Abstract

Sarcoidosis is a multisystem inflammatory disease of incompletely elucidated etiology. It is characterized by the formation of non-caseating granulomas which can be found in any organ or tissue. The most commonly affected organ is the lung and the typical presentation is with bilateral hilar lymphadenopathy, pulmonary infiltrates, and cutaneous and ocular symptoms. However, as the granulomatous reaction can take place in any organ and tissue, sarcoidosis can have variable clinical presentations. It is well documented that sarcoidosis may mimic a great array of autoimmune and non-autoimmune diseases. Moreover, it can also coexist with these disorders, which makes the diagnostic pathway all the more challenging.

Keywords: sarcoidosis, autoimmune disease, non-caseating granuloma

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Introduction

Sarcoidosis is a systemic inflammatory disease characterized by a granulomatous reaction leading to the formation of non-caseating granulomas in different organs and systems.

The prevalence of sarcoidosis is dependent on the geographic location and it has been observed that it is the highest in Nordic countries [1]. In 2016, studies were conducted in the USA and Sweden, estimating a prevalence as high as 160 cases per 100,000 people in Sweden and 50 cases per 100,000 people in the USA, reaching the highest rates of sarcoidosis worldwide [2,3].

Sarcoidosis is seen more frequently in females; the median age at diagnosis varies between 35 and 50 years [4].

Pulmonary involvement is seen in more than 90% of sarcoidosis patients and they usually present with dyspnea, dry cough, and chest tightness [5]. 30-50% of patients have extra-pulmonary manifestations, out of which skin and eye involvement are the most common manifestations [6]. Sarcoidosis may present acutely as Lofreg's syndrome, which is characterized by the presence of fever, bilateral hilar lymphadenopathy, erythema nodosum, and bilateral ankle arthritis, or it may present with non-specific signs and symptoms like fever, weight loss, and malaise [7].

Pathogenesis

Although the pathogenesis of sarcoidosis is yet to be fully understood, it is assumed that the formation of granulomas is the result of cellular immune activation after exposure to an environmental or infectious agent, in genetically susceptible individuals [8]. It is