Everything you always wanted to know about sarcoidosis... but were afraid to ask

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Despite better understanding of this complex systemic disease, sarcoidosis remains a mystery. Its cause is still unknown while displaying varying clinical expression and severity. This issue of the Quarterly Medical Review aims at delivering an accurate state of the art on the pathogenesis of sarcoidosis and to describe its major clinical manifestations. Müller-Quernheim et al. have depicted sarcoidosis as an exaggerated immune response of an individual susceptible background against pathogen-associated molecular patterns of harmless commensals [1]. Genetic contribution to the pathogenesis of sarcoidosis is well recognized but still in its infancy despite recent advances. Moreover, environmental exposure to causes is believed to interact with genetic factors in the pattern of presentation and evolution. There is a need to better categorize clinical phenotypes of the disease in order to improve genetic/phenotypic correlations and eventually contribute to a more personalized care of patients. Baughman et al. have dealt with the many respiratory manifestations of the disease [2]. In sarcoidosis, respiratory symptoms stem not only from interstitial lung disease but also from involvement of upper or large airways, as well as bronchioles, pulmonary vessels, heart, and muscle. In many cases, chest X-ray and pulmonary function tests are most useful at baseline and follow-up. However, pulmonary involvement can be multifactorial, and several tests are needed to better understand the respiratory component of the disease. Computed tomography of the chest appears useful in difficult cases. The article by Nunes et al. has focused on pulmonary hypertension, one of the most severe manifestations of sarcoidosis [3]. Pulmonary hypertension can be difficult to diagnose and classify properly and its management is challenging. Pulmonary hypertension may be due to congestive heart disease (post-capillary pulmonary hypertension), thrombo-embolic events or specific pre-capillary pulmonary arterial remodeling. Right heart catheterisation is the gold standard to confirm diagnosis. Pulmonary hemodynamics are warranted in patients who are candidates for lung transplantation, when pulmonary vascular disease appears disproportionate with interstitial lung involvement or when there is a clinical suspicion of pulmonary hypertension (dyspnea, chest pain, right heart failure...). Randomized controlled trials are needed to assess the interest of medical therapies approved for pulmonary arterial hypertension in well-phenotyped pulmonary hypertensive patients. Chapelon-Abric has underscored the difficulties to confirm with a high degree of confidence a diagnosis of cardiac sarcoidosis and the significant impact of such diagnosis on prognosis and treatment [4].
various presentations of cardiac sarcoidosis and the interests and limitations of cardiac investigations used for diagnosis are well discussed. In particular, magnetic resonance imaging and FDG-PET/CT appear to be two very promising tools in that setting. Nozaki and Judson have described the many faces of neurosarcoidosis and how to confirm diagnosis in various contexts [5]. Differential diagnosis (particularly multiple sclerosis) are well discussed as well as ways to better recognize small nerve fibre neuropathy which can severely impact quality of life. Bodaghi et al. have presented imaging tools and diagnostic criteria for ocular sarcoidosis [6]. In their review, they have described clinical findings and how to monitor and treat them. Last, Mañá and Marcoval gave a complete description of the cutaneous manifestations of sarcoidosis even those less usual and more difficult to diagnose and manage. This paper is well supported by well-chosen pedagogic illustrations [7]. We hope that this issue of the Quarterly Medical Review will help our readership to better understand and manage this complex condition.

Disclosure of interest: the authors have not supplied their declaration of conflict of interest.

References


