EDITORIAL POINT OF VIEW

Comparing and Contrasting Guidelines for the Diagnosis of Cardiac Sarcoidosis

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The Japanese Circulation Society recently published new guidelines for the diagnosis of Cardiac Sarcoidosis (CS). In this issue of the journal they are summarized, for the first time, in English (1). There are two other guideline documents. The World Association of Sarcoidosis and Other Granulomatous Disorders Sarcoidosis Organ (WASOG) Assessment Instrument created in 1999 and updated in 2014 (2). Also in 2014, the Heart Rhythm Society (HRS) in collaboration with the American College of Cardiology, American College of Chest Physicians, American Heart Association, Asia Pacific Heart Rhythm Society and the European Heart Rhythm Association published their international guideline document (3). The HRS document was closely aligned with the WASOG publication (2, 3).

As co-chair of this HRS document I have been invited to compare and contrast our guidelines with this new publication (1, 3). There are many similarities, for example both have two diagnostic pathways, however there are a number of important differences. One major difference is the requirement for a positive extra-cardiac biopsy in the clinical diagnosis pathway. The HRS guidelines require this but the Japanese guidelines do not; specifically they state "granulomas are found in organs other than the heart, or [2] the patient shows clinical findings strongly suggestive of pulmonary or ophthalmic sarcoidosis and at least 2 of the five characteristic laboratory findings of sarcoidosis." Another difference is in the need for multiple clinical criteria to make the diagnosis via the clinical pathway (two or more major criteria or one major and two or more minor criteria). This contrasts with the HRS document which only requires one 'major' criteria (1). Hence I suspect the Japanese guidelines will result in some cases of inactive clinically silent CS being 'missed'. For example patients with small patches of delayed enhancement on MRI but no other abnormal testing will not meet the threshold for diagnosis.

Also the new Japanese guidelines tackle, for the first time, the definition of and criteria for the diagnosis for isolated cardiac sarcoidosis (ICS) (1). This is important as this is an increasingly recognized clinical scenario. The prevalence of isolated CS varies widely with reported rates from 27 to 54% (4-7). There are likely three main reasons for this variability. Firstly, the lack of an agreed definition of isolated CS. Secondly the heterogeneous nature of many of the cohorts and thirdly, the diagnostic method(s) for assessing extra-cardiac involvement. Hence a key starting point to understand ICS is for all of us to agree on a standardized definition and I agree with much of what is proposed. However I would like to suggest a minor modification to the wording of criteria number 2 to remove any possibility of ambiguity. Specifically to state that lymph nodes are considered an organ i.e." ^{67}Ga scintigraphy or ^{18}F-FDG PET reveals no abnormal tracer accumulation in any organs, ***including lymph nodes***, other than the heart. Further criteria 3 might be clearer if stated as "a chest CT scan reveals nothing to suggest pulmonary sarcoidosis and no hilar and mediastinal lymphadenopathy". I suspect with this precise definition, then the prevalence of ICS will be much less than the reported rates 27 to 54% (4-7).

The suggested diagnostic criteria for ICS is more controversial as they propose that it is possible to diagnose

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ICS without a positive cardiac biopsy (1). They suggest that having a positive PET or Gallium scan and at least three other major criteria is sufficient to make the diagnosis. I would argue that this likely will lead to an over-diagnosis of ICS. Also in most cases in my experience and others it is often possible to get a positive biopsy using a guided biopsy technique. That is, using electrophysiological (electro-anatomic mapping) (8, 9) or imaging (PET or CMR) (4) guidance. Also in some cases, waiting for a few weeks, if the patient is clinically stable, likely will reveal some lymph nodes that can be biopsied. This latter point is important as extra-cardiac involvement can develop over time; for example Adamson describe two patients initially thought to have ICS but who went on to develop extra cardiac involvement (10).

The Japanese have been leaders in many aspects of CS including in guideline development and there is much to like about the latest version, most importantly the first attempt at defining ICS. However ideally in the future we should aim to get all experts together and create one set of agreed international criteria. Finally and very importantly none of the guidelines have been validated and research should focus on this.

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Conflicts of interest

None.

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