Evaluation of isolated cardiac sarcoidosis applying updated Japanese guidelines

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BACKGROUND

The overall prognosis of systemic sarcoidosis depends on the presence of cardiac involvement. This involvement is mainly diagnosed by multimodality cardiac imaging given the known difficulties with myocardial biopsies. The focus of this editorial is on the study by Okada et al. entitled ''Clinical features and prognosis of isolated cardiac sarcoidosis diagnosed using new guidelines with dedicated FDG PET/CT.''[1]

Clinical manifestation of cardiac involvement in patients with sarcoidosis is reported to be variable and can be seen in approximately 5% of patients; however, myocardial involvement has been noted in 25% of autopsies of patients with systemic sarcoidosis.[2] These numbers reflect the state of the disease mainly in western countries; however, in Japan, an autopsy study by Iwai et al. found that as high as about 70% of Japanese had sarcoid granulomas in the myocardium.[3,4]

Guidelines for the diagnosis and treatment of cardiac sarcoidosis (CS) were recently updated from the 2006 version by the Japanese Circulation Society and were published in February 2017. This update included revised major criteria for cardiac involvement of sarcoidosis and a dedicated section for isolated CS (iCS). Other guidelines for the diagnosis of Cardiac Sarcoidosis are available. The Heart Rhythm Society (HRS) published an expert consensus statement in association with the American College of Chest Physicians, American Heart Association, Asia Pacific Heart Rhythm Society, European Heart Rhythm Association, and WASOG in 2014. These guidelines included PET/CT abnormalities in the minor diagnostic criterion along with delayed gadolinium enhancement.[5,6]

The updated Japanese guidelines added abnormally high tracer accumulation in the heart with 18F-fluorodeoxyglucose (FDG) positron emission as part of the major criteria as well as a late-gadolinium enhancement (LGE) of the myocardium in gadolinium-enhanced magnetic resonance imaging (MRI). It is important to note that the guidelines mention the importance of Whole Body FDG PET/CT in the diagnostic process.

The current paper focuses on the diagnosis of iCS using the described updated guidelines and the impact that the early diagnosis of this condition has on the patient's prognosis. In summary, 306 consecutive patients with suspected CS were evaluated with FDG PET/CT after adequate patient preparation. The group evaluated a total of 82 patients after meeting inclusion and exclusion criteria. Of these patients, 55 (67.1%) were diagnosed with sCS, and 27 (32.9%) were diagnosed with isolated iCS based on the updated Japanese criteria. On imaging, they found that there was no significant difference in uptake patterns in the two groups. Patients with iCS had an increased number of affected segments, they had lower right ventricular uptake, the SUV max was lower, and the Target to background ratio was lower when compared to patients with CS and systemic involvement. In terms of prognosis, they found that 29.3% of patients experienced adverse events.

A prior study by Hideki Kawai in 2020 using the new criteria for the identification of CS evaluated 94 consecutive patients and found a higher diagnostic yield for the disease with more than 1.5-fold of patients diagnosed with CS using the updated criteria versus the previous international criteria.[7] A study by Juneau et al.

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evaluated extra-cardiac and cardiac findings in 31 patients; in their study, only one patient was found to have iCS; however, this study was not done using the updated criteria.[8] Another study by Giudicatti et al.,[9] retrospectively evaluated patients with FDG PET/CT and using the HRS criteria, and from 32 cases only 3/32 (9.4%) had iCS. Very few studies have focused on the detection of iCS with the updated Japanese criteria. In Japanese population as well as in other western countries, the application of these new criteria is increasingly important.

A limitation encountered includes, a low number of patients, which is common in studies focusing on cCS due to the difficulties in diagnosis and the low prevalence of disease. The fact that EMB was negative in 81% of patients with iCS is a limitation, not necessarily attributed to the study but to the fact that EMB has a low sensitivity, which could be explained by the patchy distribution of disease in the left ventricular walls.

Without tissue diagnosis, other etiologies that can mimic iCS have to be considered, these include infectious myocarditis (viral, bacterial, fungal), cardiac amyloidosis, arrhythmogenic cardiomyopathy, hypertrophic cardiomyopathy, metabolic causes like Fabry's disease, acute rheumatic heart disease, etc.

This study illustrates the importance of early diagnosis of iCS using FDG PET/CT and how the criteria in the updated Japanese guidelines increase the rate of detection even in the absence of a successful EMB. Early detection translates into earlier treatment which may reduce the incidence of adverse fatal cardiac events; however, more studies are needed to prove this point. Also, future studies should focus on larger groups of patients with serial FD PET examinations to further aid in the developing body of literature about this rare but deadly disease. This movement towards clinical diagnosis where histopathologic confirmation is not possible, will have future implications especially on disease definition in order to decide treatment strategies or inclusion in research studies.[10]

Disclosures

The authors have no conflicts of interest to disclose.

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