Prognosis of cardiac sarcoidosis in Japanese patients with implantable cardioverter defibrillator - with versus without other organ involvement -

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Introduction: Cardiac sarcoidosis (CS) is a granulomatous disorder affecting multiple organ systems including lungs, lymph nodes, a liver, skin, eyes, and gastrointestinal tract. The leading causes of death in patients with CS are cardiac arrhythmias, progressive heart failure, and progressive respiratory failure. Patients with CS are at high risk of sudden cardiac death from ventricular arrhythmias. Implantable cardioverter defibrillators (ICDs) have been used for primary and secondary prevention of sudden death in patients with CS. However, there are few reports about the clinical prognosis in CS patients with ICD. The purpose of this study was to identify the difference between prognosis of Japanese CS patients who received an ICD with and without other organ involvement.

Methods: We retrospectively analyzed the database of our ICD clinic. Of 498 consecutive ICD patients, 34 patients (6.8%) were diagnosed with CS. All of CS patients received steroid therapies. We defined CS patients without other organ involvement as isolated CS, and with other organ involvement as non-isolated CS. We examined their background, left ventricular ejection fraction before and after steroid therapies, the incidence of appropriate ICD therapies, and the incidence of hospitalization and death.

Result: Diagnosis of CS was made by cardiac magnetic resonance (CMR) (n=7), positron emission tomography (PET) (n=14), CMR and biopsy (n=4), CMR and PET (n=12), PET and biopsy (n=8), late enhanced cardiac computer tomography (CT) and CMR (n=5), and CT and PET (n=5). Figure shows the patients background and the clinical outcomes during the follow-up. Of 34 CS patients with an ICD, 13 (38%) had isolated CS. Of 13 patients, 6 (36%) were male, age was 59±12 years, follow-up period was 68±57 months, and left ventricular ejection fraction was 38±12%. During the follow-up period, appropriate therapies occurred in 4 of 13 (31%) isolated CS patients and 6 of 21 (29%) non-isolated CS patients (p=1.0). Two of 13 (16%) isolated CS patients were hospitalized for heart failure, compared with 7 of 21 (33%) non-isolated CS patients (p=0.43).

Conclusion: The prognosis Japanese patients with CS was not good. There was no significant difference between the prognosis of CS with and without other organ involvement in this population.