Recurrences of disease activity in patients with cardiac sarcoidosis under corticosteroid therapy: prevalence, clinical background and prognosis

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Background: Corticosteroid therapy (CTx) has been widely accepted as first-line therapy for cardiac sarcoidosis (CS), but there are very limited data regarding recurrence of disease activity of CS. We retrospectively investigated the prevalence, patient characteristics and prognostic significance in patients with recurrences of CS.

Methods: We identified 102 consecutive patients who were clinically diagnosed CS (admission: 2012 and 2019) and whose disease activity was diminished clinically at least once. Recurrence of CS was defined as detection of increased uptake of 18F-fluoro-2-deoxyglucose or gallium-67 in the follow-up examination. Composite adverse events (events) were defined as all-cause of death or hospitalization for heart failure. Echocardiographic and laboratory data before initiation of CTx were obtained.

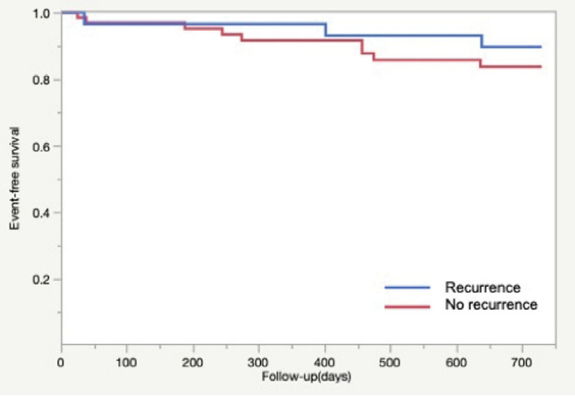
Results: During the follow-up term (median: 4.9 years), the recurrences of CS occurred in 28 patients at 30 months (median) after the initiation of CTx. In patients with recurrence, left ventricular (LV) ejection fraction before initiation of CTx was lower than in those without recurrences (median: 31% vs. 39%, $p<0.05$). After the detection of CS recurrences, 17 patients were treated with only increases of PSL and remaining 11 patients were treated with adding other immunosuppressive therapies to CTx. The univariate analysis demonstrated that there were no parameters in echocardiographic and laboratory examinations to predict the recurrences of CS. The results of univariate analysis for event occurrences (n=12) are shown in the Table. Additionally, the Kaplan-Meier analysis showed that there were no differences in event free survival rate in the patients with and without CS recurrences (Figure).

Conclusion: This study showed that the recurrences of disease activity were observed in a substantial number of patients with CS even under the CTx. All patients received intensification of CTx or additional immunosuppressive therapy, and LV systolic function, rather than the recurrence itself, was associated with clinical outcomes in this study.

Univariate Cox hazard analysis

	Composite adverse events	
	HR (95% CI)	P value
LVEF, 1%	0.927 (0.871–0.976)	0.003
LA volume, 1ml/m^2	1.029 (1.007–1.051)	0.012
BNP	1.002 (1.001–1.003)	0.009

CI, confidence interval; EF, ejection fraction; HF, heart failure; HR, hazard ratio; LA, left atrium; LV, left ventricle



Kaplan-Meier analysis for events