Cardiac Sarcoidosis treated with non-steroidal immunosuppressive therapy

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Background: Non-steroidal immunosuppressive therapy is a potential treatment strategy for cardiac sarcoidosis. However, due to the lack of robust evidence, it is not recommended as an established treatment option in the guideline.

Purpose: The purpose of the study is to demonstrate the clinical outcome of the patients with cardiac sarcoidosis using non-steroidal immunosuppressive therapy in a retrospective registry entitled ILLUMINATE-CS (ILLUstration of the Management and prognosis of JapaNese pATiEnts with Cardiac Sarcoidosis).

Methods: Of an entire cohort of 512 patients, 26 who received immunosuppressive therapy other than steroids were recruited for the analysis. The clinical outcome included all-cause death, fatal ventricular arrhythmic event (FVAE), and worsening heart failure with hospitalization.

Results: The reasons to use non-steroidal immunosuppressant were increased fluorodeoxyglucose (FDG) accumulation on the heart suggesting worsened inflammation (n = 13), side effects of steroid (n = 7), ventricular tachycardia or frequent ventricular premature contraction (n = 4), complete atrioventricular block (n = 2), worsened symptom relating to non-cardiac sarcoidosis including neuro and pulmonary sarcoidosis (n = 2), and the other reason (n = 2), with some overlapping. Used non-steroidal immunosuppressants were comprised of methotrexate (n = 20, 6.9 ± 2.1 mg/week), cyclosporine (n = 2, 75 or 100 mg/day), cyclophosphamide (n = 2, 25 or 50 mg/day), and azathioprine (n = 3, 100 mg/day in each). Side effects of non-steroidal immunosuppressant were observed in six cases and included one with methotrexate-related interstitial pneumonia, one with a methotrexate-associated lymphoproliferative disorder, one with septic coxitis with methotrexate, one with methotrexate-induced acute kidney injury, one with methotrexate-induced hepatic dysfunction, and one with azathioprine-induced anemia. After the addition of a non-steroidal immunosuppressant, steroids could be reduced in 14 cases of 26 cases with a median maintenance dose of 5 (interquartile range of 5–17) mg, although no cases stopped steroids. Of 13 cases who started non-steroidal immunosuppressant for the increased FDG uptake, decreased FDG uptake was observed in seven cases at the follow-up scan. Clinical outcome events were observed in 11 cases (42.3%). Detected events included all-cause death in five cases (19.2%), FVAE in four cases (15.4%), and worsening heart failure with hospitalization in five cases (19.2%), with some overlapping.

Conclusions: Non-steroidal immunosuppressive therapy may be a possible treatment option for patients who are not stabilized using sole steroid therapy or who suffer from the side effects of steroids.