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Letters to the Editor

Cardiac Sarcoidosis: Indications for Implantable Cardioverter-Defibrillator Therapy

To the Editor:

In clinical practice, cardiac sarcoidosis (CS) has been an underdiagnosed phenomenon and is potentially associated with adverse clinical outcomes, including malignant arrhythmias.[1,2] The recent article by Powell et al. describes an atypical case of CS presenting with multiple myocardial masses in a young female patient.[1] The authors placed an implantable cardioverter-defibrillator (ICD) in the patient.[1] Within this context, we would like to know more about the indication(s) for an ICD in this patient, and we highlight the general indications for ICD therapy in the setting of CS.

ICD therapy is well known to be associated with a variety of complications in the long term.[3] Thus, proper evaluation of ICD indications is important, particularly for relatively young patients.[3] In patients with CS, the Heart Rhythm Society guidelines[2] strongly recommend ICD implantation in the presence of one or more of the following conditions: (i) a history of cardiac arrest or sustained spontaneous ventricular arrhythmias (VAs) (class I); (ii) a left ventricular ejection fraction (LVEF) value of $\leq$ 35% (class I); (iii) stimulated VAs on electrophysiological study (EPS; class IIa); (iv) planned permanent pacemaker implantation (class IIa); and (v) near-syncope or syncope considered to have an arrhythmic origin (class IIa).[2] However, the level of ICD recommendation is relatively weak (class IIb) in patients with mild-to-moderate left ventricular systolic dyfunction (LVEF: 36%-49%) and /or right ventricular systolic dysfunction.[2] In those with any form of systolic dysfunction on admission, decision-making for ICD implantation should be carried out following a period of immunosuppression (in case of active myocardial inflammation) along with optimal heart failure (HF) therapy, all of which might lead to a significant improvement in systolic functions.[2] Therefore, the Heart Rhythm Society guideline recommends a period of at least 3 months in this context.[2] In patients without an overt or strong indication for ICD implantation (including those with normal systolic functions on admission), detection of late gadolinium enhancement on cardiac magnetic resonance imaging potentially can suggest further strategies, including EPS (for VA stimulation).[2]

Notably, ventricular aneurysm formation and high levels of oxidative stress (and systemic inflammation) markers recently have been suggested to be important predictors of a first episode of sustained VAs/sudden cardiac death in patients with CS.[4] Therefore, these novel findings may lead to an expansion of ICD indications in the setting of CS.[4] Based on the aforementioned information, we wonder about the specific indication(s) for ICD implantation in this patient (including low LVEF).[1] Did the patient also undergo EPS for VA stimulation and evaluation of atrioventricular node functions? Palpitations reported by the patient[1] might have been due to a variety of relatively benign arrhythmias (including sinus tachycardia and extrasystoles); hence, these possibilities could have been further investigated to justify ICD implantation. In particular, we also wonder whether the patient[1] received appropriate ICD shocks on follow-up.

In summary, the decision-making for ICD implantation (for primary prevention) is generally based on the evaluation of LVEF values (along with late gadolinium enhancement evaluation and EPS in certain settings) in patients with CS.[2] However, this decision-making should be carried out following a period of optimal therapy in those with left ventricular systolic dysfunction on admission.[2] During this period, a reasonable approach is to encourage the use of temporary strategies, including wearable cardioverter-defibrillator therapy, in patients with high-risk features on initial evaluation (including those with very low LVEF values or severe systemic inflammation on admission). Finally, further strategies (evaluation of specific biomarkers, etc.)[4] are still needed to better identify patients with CS requiring ICD therapy.

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