

Role of echocardiography in suspicion of Cardiac sarcoidosis

A case of cardiac sarcoidosis

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Abstract

The echocardiogram is often abnormal in manifest cardiac sarcoidosis (CS) disease, but is usually normal in clinically silent CS. Abnormalities are variable and usually nonspecific, although interventricular septal thinning, especially basal, can be a feature of CS. Less frequently, there may be an increase in myocardial wall thickness, simulating left ventricular (LV) hypertrophy or resembling hypertrophic cardiomyopathy. Other abnormalities include LV and/or RV diastolic and systolic dysfunction, isolated wall motion abnormalities, basal septal thinning, and aneurysms. Regional wall motion abnormalities are usually seen in a non-coronary distribution. Cardiac involvement is undeniably one of the most challenging issues in sarcoidosis. Although autopsy studies reveal heart lesions in 20 to 30% of sarcoid patients, fewer than 5% suffer from clinical disease. Cardiac sarcoidosis (CS) has a predilection for the myocardium, but the pericardium and endocardium may also be affected. CS manifestations are various and most frequently include the following: aberrations of atrioventricular or intraventricular conduction, either silent or symptomatic; ventricular arrhythmias; subacute congestive heart failure; and sudden death. CS must be detected in all sarcoid patients by means of detailed medical history, physical examination, and resting electrocardiogram (ECG) at first evaluation and during follow-up. In patients with suspected CS, further investigations are aimed at evaluating diagnosis and cardiac consequences. Unfortunately, no gold standard exists that would allow CS diagnosis with a level of confidence. Endomyocardial biopsy is an invasive procedure that lacks sensitivity. Patients need, at a minimum, specialized cardiologic advice, echocardiography, and 24-hour ambulatory ECG. Other diagnostic tools include thallium, technetium, and gallium scintigraphy, and more recently, 18F-fluorodeoxyglucose positron emission tomography and cardiac magnetic resonance (CMR). The respective role of these new imaging tools in the diagnostic approach remains to be defined. CMR has the advantage of not exposing patients to radiation, but it is not feasible in those with cardiac devices. In Western countries, heart involvement accounts for 13 to 25% of sarcoidosis-related deaths, and it is the leading cause of mortality in Japan.

The main prognostic indicators are New York Heart Association functional class, left ventricular enlargement, and sustained ventricular tachycardia. Treatment is based on systemic corticosteroids with an initial dose between 30 mg/day and 1 mg/kg/day (which is usually maintained for at least 24 months), specific cardiologic agents, and the placement of a pacemaker or implantable cardiac defibrillator in case of an atrioventricular block or severe intractable ventricular arrhythmias. Cardiac transplantation is exceptionally required.

There were no restrictions about the language of publication. Two reviewers independently performed the screening of studies, selection, validation, data extraction, and the assessment of methodological quality. To be included, studies were required to state that they were randomized, double-blind, and placebo controlled, and used hawthorn extract monopreparations. Thirteen trials met all inclusion criteria. In most of the studies, hawthorn was used as an adjunct to conventional treatment. Eight trials including 632 patients with chronic heart failure (New York Heart Association classes I to III) provided data that were suitable for meta-analysis. For the physiologic outcome of maximal workload, treatment with hawthorn extract was more beneficial than placebo (weighted mean difference, 7 Watt; 95% confidence interval [CI]: 3 to 11 Watt; $P < 0.01$; $n = 310$ patients). The pressure-heart rate product also showed a beneficial decrease (weighted mean difference, -20; 95% CI: -32 to -8; $n = 264$ patients) with hawthorn treatment. Symptoms such as dyspnea and fatigue improved significantly with hawthorn treatment as compared with placebo. Reported adverse events were infrequent, mild, and transient; they included nausea, dizziness, and cardiac and gastrointestinal complaints. In conclusion, these results suggest that there is a significant benefit from hawthorn extract as an adjunctive treatment for chronic heart failure.

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