Shortness of breath and cardiac conduction abnormality caused by infiltrative cardiomyopathy

Promporn Suksaranjit1, John J. Ryan1, Frederick T. Han1, Jose Nativi-Nicolau1, Monica P. Revelo2, and Brent D. Wilson1*

1Division of Cardiovascular Medicine, University of Utah, 30 North 1900 East, Room 4A100, Salt Lake City, UT 84132, USA; and 2Department of Pathology, University of Utah, Salt Lake City, UT, USA

* Corresponding author. Tel: +801 585 2341, Fax: +801 587 5874, Email: brent.wilson@hsc.utah.edu

A 43-year-old man presented with dyspnoea on exertion. Electrocardiography (ECG) showed sinus bradycardia with second degree atrioventricular block, and right bundle branch block (Panel A). An event monitor revealed symptomatic sinus arrest with a junctional escape rhythm. Echocardiography and cardiac magnetic resonance (CMR) demonstrated right ventricle (RV) enlargement and reduced RV function (Panel B; Supplementary material online, Movies S1 and S2). Late gadolinium enhancement (LGE) was present in the RV free wall, the interventricular septum, and subepicardium of the left ventricle (LV) (Panel C). Right heart catheterization revealed normal right-sided filling pressures and decreased cardiac output. The patient underwent electroanatomical mapping-guided RV endomyocardial biopsy (EMB) where regions of low voltage with fractionated electrograms and late potentials were targeted for biopsy (Panel D, Supplementary material online, Movie S3). Biopsy samples showed non-necrotizing granulomatous inflammation involving the myocardium consistent with sarcoidosis (Panel E).

Sarcoidosis is a heterogeneous, non-caseating, granulomatous disorder. Non-invasive and invasive tests are required to establish a diagnosis of cardiac sarcoidosis (CS). Cardiac magnetic resonance and positron emission tomography provide the highest sensitivity for detection of CS, but CMR findings of mid-myocardial or sub-epicardial LGE appear to be more specific. Endomyocardial biopsy is recommended to obtain histological diagnosis when there is no histological confirmation from another source. Endomyocardial biopsy has low sensitivity with high false-negative results from sampling error. In this case, EMB guided by anatomical information from CMR and electrical voltage mapping localized pathologic myocardial tissue and increased the diagnostic yield of the procedure.

Authors’ contributions

Drs Suksaranjit, Ryan, and Wilson performed the initial review of the literature, manuscript drafting, and review. Drs Han, Revelo, and Nativi-Nicolau were involved with image acquisition and manuscript review process. Corresponding author had full access to all the data and final responsibility for the decision to submit for publication.

Supplementary material is available at European Heart Journal online.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2015. For permissions please email: journals.permissions@oup.com.