Imaging of Idiopathic Restrictive Cardiomyopathy: A Multimodality Approach

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A 20-year-old man presented to our hospital with a 1-month history of shortness of breath, fatigue, and syncope. His mother had died suddenly at 40 years old (Picture 1). His medical history included persistent atrial fibrillation (Picture 2A) and anaplastic large-cell lymphoma in childhood, for which he received a total doxorubicin dose of 150 mg/m². Chest X-ray showed cardiomegaly (Picture 2B); his B-type natriuretic peptide level was elevated at 1,088.8 pg/mL (normal range, ≤18.4 pg/mL), his troponin I level was elevated at 41.7 ng/L (normal range, ≤14 ng/L), and echocardiography demonstrated biatrial enlargement with a normal left ventricular (LV) ejection fraction (Picture 2C). Chest computed tomography revealed no mediastinal or hilar lymphadenopathy. Transesophageal echocardiography revealed a thrombus in the left atrial appendage. His indexed LV end-diastolic volume, LV ejection fraction, and indexed LV mass measured by cine-cardiovascular magnetic resonance imaging were 70.8 ml/m² (normal range, 60-110 ml/m²), 44.4% (normal range, 48-69%), and 44.2 g/m² (normal range, 35-70 g/m²), respectively (1), precluding a possibility of hypertrophic heart diseases, including hypertrophic cardiomyopathy, cardiac amyloidosis, and Anderson-Fabry disease. Late gadolinium-enhanced imaging showed circumferential midwall hyperenhancement (Picture 3, arrows). Pericardial thickening and delayed hyperenhancement were absent. Cardiac catheterization showed a prominent "y" descent in the right atrial pressure tracing (Picture 2D, red-arrow) and a typical "square root" sign in the right ventricular pressure tracing (Picture 2D, blue-arrow). A right ventricular endomyocardial biopsy revealed extensive myocardial fibrosis (Picture 4A-B) and myofibrillar disarray (Picture 4C) without amyloid deposition, granuloma, inflamma-

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tory cell infiltration, or vacuolar degeneration of cardiomyocytes, which were consistent with idiopathic restrictive cardiomyopathy. Multimodality imaging is needed to differentiate idiopathic restrictive cardiomyopathy from other cardiomyopathies. This case highlights the importance of a multimodality approach for the differential diagnosis and comorbidity assessment in restrictive cardiomyopathy.

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cal Systems and also received payment for lectures from Daiichi-Sankyo, Philips Medical Systems, Eisai, Bayer Healthcare, GE Healthcare, and Canon Medical Systems. The other authors have no financial conflicts of interest.

Reference


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