Extensive Myocardial Fibrosis in a Patient With Hypertrophic Cardiomyopathy and Ventricular Tachycardia Without Traditional High-Risk Features

Sergio Bongioanni, MD; Paolo Spirito, MD; Andrea Sibona Masi, MD; Amedeo Chiribiri, MD; Rodolfo Bonamini, MD; Maria Rosa Conte, MD

Although patients with hypertrophic cardiomyopathy (HCM) and without conventional risk factors for sudden death are generally considered at low risk, the magnitude of risk in such patients remains unknown, and additional markers of prognosis may permit more accurate risk stratification.[1–3] We report a patient with HCM and without conventional risk factors who had a sustained life-threatening ventricular tachyarrhythmia 6 months after identification of extensive myocardial fibrosis at contrast-enhanced cardiovascular magnetic resonance (CMR).

Case Description

A 61-year-old asymptomatic patient with HCM had been followed at our institution for 12 years. The patient had none of the conventional risk factors for sudden death. There was no family history of premature HCM-related sudden death, unexplained syncope, or abnormal blood pressure response during exercise. No episode of nonsustained ventricular tachycardia (≥3 beat run) had been documented in any of the 6 ambulatory Holter ECGs recorded during follow-up. The echocardiogram showed a nondilated and hypertrophied left ventricle (LV) with a maximal LV wall thickness of 22 mm at the level of the anterior septum, which had remained unchanged since initial evaluation. The LV outflow gradient was 35 mm Hg at rest. The patient was treated with 50 mg/d atenolol. Recently, a contrast-enhanced CMR was performed as part of routine clinical evaluation. After gadolinium infusion, multiple and extensive areas of myocardial late enhancement were demonstrated, involving 80% of the septum and 45% of the free wall (Figure 1). Six months after CMR, the patient was admitted to our emergency department for intense palpitations associated with lightheadness. A 12-lead ECG showed sustained ventricular tachycardia with a 320-ms cycle and a heart rate of 190 bpm (Figure 2). Blood pressure was 100/70 mm Hg. Given the impending hemodynamic instability, electric cardioversion was performed with prompt restoration of sinus rhythm. Neither release of cardiac enzymes nor electrolyte imbalance was detected. A coronary arteriogram was performed and showed no significant coronary artery stenoses. Because of the episode of sustained ventricular tachycardia, a cardioverter-defibrillator (ICD) was implanted before hospital discharge.

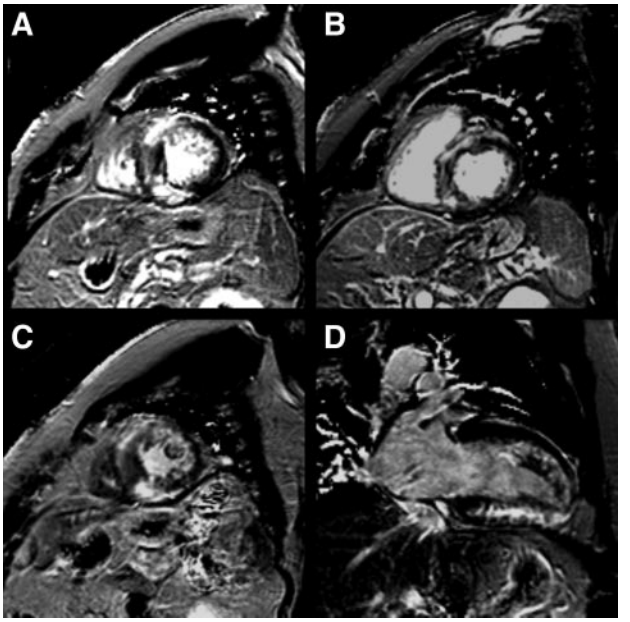

Figure 1. Contrast-enhanced CMR images of the left ventricle in short axis at equatorial (A), basal (B), and apical level (C) and in 2-chamber long-axis view (D). Images show areas of delayed enhancement at the level of the interventricular septum and the free wall, indicating the presence of extensive and transmural fibrosis.

Comments

The patient reported here shows that the current risk markers may be insufficient to identify patients with HCM at increased risk of sudden death who may be candidates for the ICD. In the absence of any of the acknowledged risk factors, this patient developed a sustained life-threatening ventricular tachyarrhythmia that required electric cardioversion. However, a recent CMR had identified extensive and potentially arrhythmogenic myocardial scarring. Therefore, our experience in this patient suggests that severe myocardial fibrosis

From the Ospedale degli Infermi (S.B., A.S.M., M.R.C.), Rivoli, Torino, Italy; Ente Ospedaliero Ospedali Galliera (P.S.), Genova, Italy; and Ospedale Molinette (A.C., R.B.), Torino, Italy.

Correspondence to Sergio Bongioanni, MD, Ospedale degli Infermi, Strada Rivalta 47, 10098, Rivoli, Torino, Italy. E-mail sergio.bongioanni@virgilio.it

(*Circ Cardiovasc Imaging*. 2009;2:349-350.)

Circ Cardiovasc Imaging **is available at http://circimaging.ahajournals.org** **DOI: 10.1161/CIRCIMAGING.108.824839**

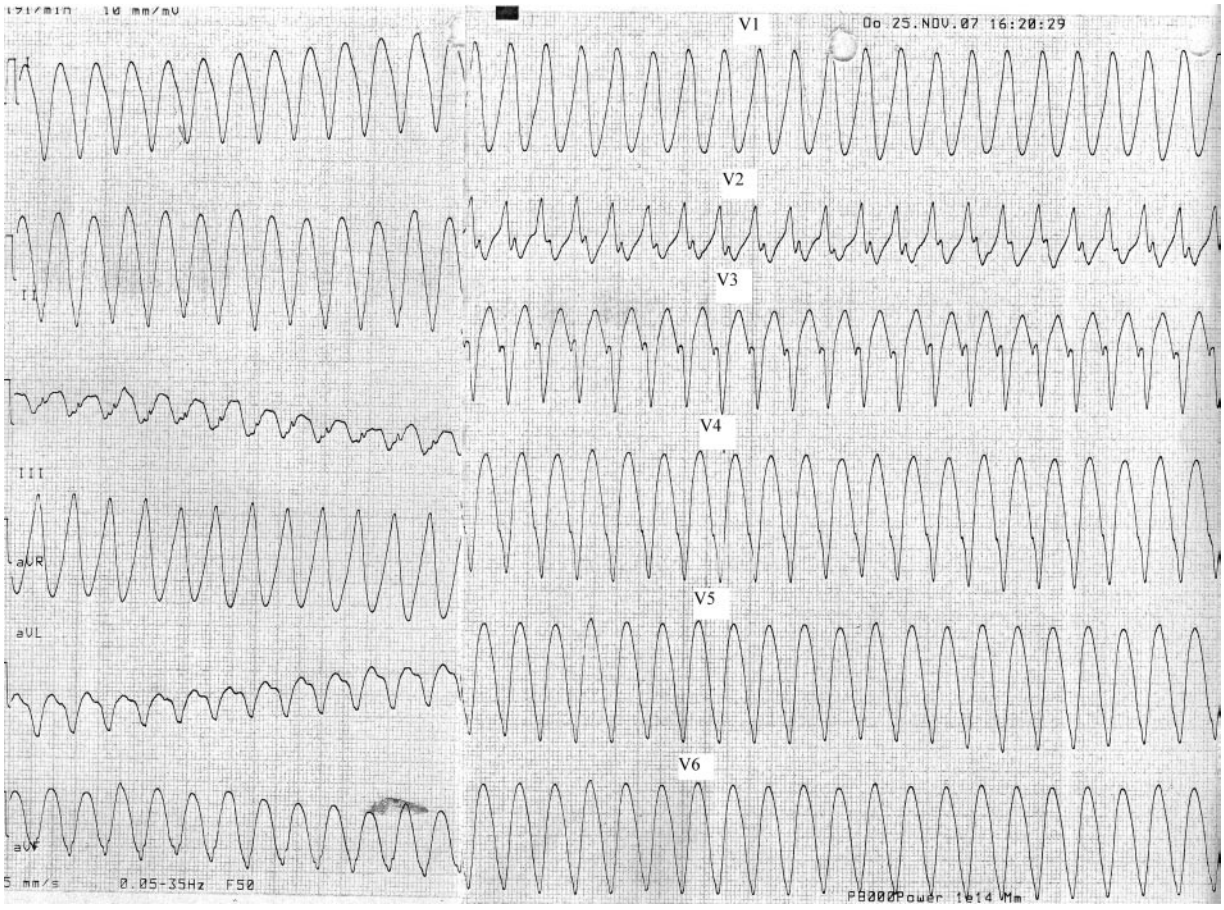

Figure 2. Twelve-lead ECG shows monomorphic ventricular tachycardia with a 320-ms cycle and a heart rate of 190 bpm.

detected in vivo by contrast-enhanced CMR could represent an additional marker of increased risk in patients with HCM.

Disclosures

None.

References

1. Maron BJ, McKenna WJ, Danielson GK, Kappenberger LJ, Kuhn HJ, Seidman CE, Shah PM, Spencer WH III, Spirito P, Ten Cate FJ, Wigle ED. Task Force on Clinical Expert Consensus Documents: American College of Cardiology; Committee for Practice Guidelines European Society of Cardiology. *J Am Coll Cardiol*. 2003;42:1687–1713.
2. Maron BJ, Spirito P, Shen WK, Haas TS, Formisano F, Link MS, Epstein AE, Almquist AK, Daubert JP, Lawrenz T, Boriani G, Estes NA 3rd, Favale S, Piccininno M, Winters SL, Santini M, Betocchi S, Arribas F, Sherrid MV, Buja G, Semsarian C, Bruzzi P. Outcomes of patients with implantable cardioverter-defibrillator for prevention of sudden cardiac death in hypertrophic cardiomyopathy. *JAMA*. 2007;298:405–412.
3. Adabag AS, Maron BJ, Appelbaum E, Harrigan CJ, Buros JL, Gibson CM, Lesser JR, Hanna CA, Udelson JE, Manning WJ, Maron MS. Occurrence and frequency of arrhythmias in hypertrophic cardiomyopathy in relation to delayed enhancement on cardiovascular magnetic resonance. *J Am Coll Cardiol*. 2008;51:1369–1374.

Extensive Myocardial Fibrosis in a Patient With Hypertrophic Cardiomyopathy and Ventricular Tachycardia Without Traditional High-Risk Features

Sergio Bongioanni, Paolo Spirito, Andrea Sibona Masi, Amedeo Chiribiri, Rodolfo Bonamini and Maria Rosa Conte

Circ Cardiovasc Imaging. 2009;2:349-350
doi: 10.1161/CIRCIMAGING.108.824839
Circulation: Cardiovascular Imaging is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 1941-9651. Online ISSN: 1942-0080