Sparkling myocardium: a unique contrast pattern in apical hypertrophic cardiomyopathy

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A 20-year-old man was referred for an evaluation of a systolic murmur and effort dyspnoea. His electrocardiogram (Fig. 1A) showed left ventricular hypertrophy (LVH) and deep T-wave inversion. The level of high-sensitivity troponin T was within the reference range at the time of consultation. Echocardiography (Fig. 1B) revealed severe concentric LVH with highly echogenic intramyocardial longitudinal and transverse bands visible both at the septal and lateral walls. The SonoVue® (Bracco, Chicago, IL, USA) contrast agent was used for better definition of the endocardial border, and disclosed severe LVH involving all ventricular walls but more pronounced at the apex, featuring an “ace-of-spades” cavity geometry. During diastole, we also noticed multiple small echogenic foci of contrast diffusely flashing within the myocardial walls, creating an unexpected pattern that resembled moving sparkles (Fig. 1C; Suppl. Video 1, 2 — see journal website).

This striking contrast echocardiography finding has never been reported at our experienced laboratory, nor were we able to find any similar report in previous publications. Our hypothesis was that this pattern was related to the presence of prominent myocardial microvasculature that fills with contrast during diastolic myocardial relaxation [1]. Other potential explanations included hypertrabeculation, myocardial recesses, and coronary-to-left ventricular microfistulae, but there was no imagiological evidence to support these hypotheses in our case. Cardiac magnetic resonance (Fig. 1D, E) confirmed the diagnosis of apical hypertrophic cardiomyopathy (HCM), and genetic testing revealed a heterozygous mutation in the delta-sarcoglycan gene (p.Lys258del), previously unreported in association with HCM. In conclusion, this case demonstrates classic features of apical HCM as well as a unique intramyocardial contrast pattern, and truly highlights the benefit from the use of contrast echocardiography in HCM.

References


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