Unusual cardiac magnetic resonance findings in a young patient, years after the diagnosis of hypertrophic cardiomyopathy

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A twenty-four-year-old man who had been diagnosed with hypertrophic cardiomyopathy (HCM) at the age of 12, based on echocardiography findings of massive hypertrophy of the septum, anterior and inferior wall (maximum wall thickness of 31 mm) with typical electrocardiographic (ECG) features (Supplementary material, *Figure S1*), was intermittently followed up for over 12 years before cardiac magnetic resonance was performed. The family history was negative, the patient was asymptomatic, and at the age of 18, the 5-year sudden cardiac death risk score assessed by the online tool recommended by 2014 European Society of Cardiology (ESC) guidelines on HCM [1] was calculated at 2.6%. Hence, implantable cardioverter-defibrillator (ICD) placement was not indicated. Subsequently, the patient had been lost to follow-up for several years. At the age of 24, he contacted his cardiologist again and a cardiac magnetic resonance exam was scheduled for risk assessment. The study showed good biventricular function with normal chamber size. However, the images presented in Figure 1 and Supplementary material, *Videos S1–S3*, unexpectedly changed the long-established diagnosis of HCM. These findings were consistent with primary cardiac tumors — fibromas.

Primary cardiac tumors are rare (0.0017%––0.019% in the autopsy series), and fibromas constituted only 3.2 % of a large pathology study of 533 cardiac tumors [2]. They present as solitary tumors in most cases, and the initial

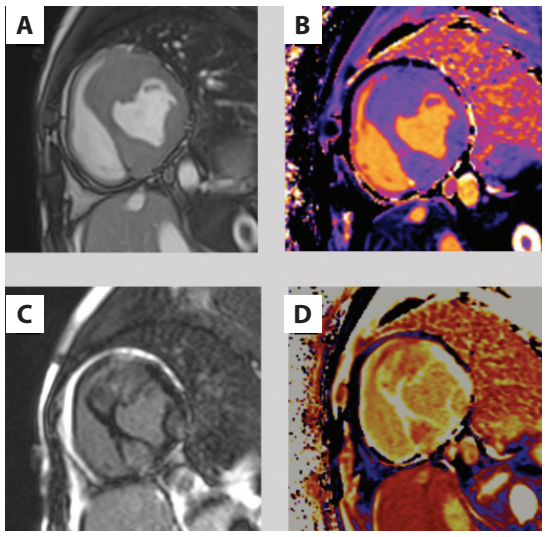

Figure 1. A. Mid-ventricular SAX bSSFP cine still-frame showing marked hypertrophy of the anterior, septal, inferior, and inferolateral segments. **B.** Native myocardial T1 mapping (MOLLI sequence) at the same slice location; no regional variation was visually apparent and the mean T1 time was 998 ms (the institutional reference range: 951-1035 ms). **C.** LGE image acquired at the same slice location at 10 min post-contrast administration (gadobutrol 0,1 mmol/kg), showing multiple highly enhanced lump tumors in the anterior, septal, inferior, and inferolateral segments. **D.** Post-contrast T1 mapping (MOLLI sequence) at the same slice location, showing marked homogenous shortening of post-contrast T1 within the tumors (Siemens Aera 1.5 T, Erlangen, Germany)

Abbreviations: bSSFP, balanced steady-state free precession; LGE, late gadolinium enhancement; MOLLI, modified Look-Locker inversion recovery; SAX, short axis

diagnosis of HCM is not uncommon based on asymmetric hypertrophy in echocardiography [3]. Conversely, multiple fibromas are an extremely rare finding. A single case of multiple fibromas in the left ventricle was found in a series of 18 symptomatic fibroma patients referred for surgical excision [4]. According to a large literature review, multiple fibromas can be found in one out of ten fibroma cases [5]. Cardiac fibromas are thought to be generally benign tumors with the greatest prevalence in the left ventricle followed by the right ventricle or the septum, and extremely rare in the atrial walls. However, younger age at diagnosis and larger relative tumor size have been linked to poor prognosis [5]. Moreover, tumors that involve the septum are more likely to cause arrhythmia and sudden cardiac death [5].

The management tends to be limited to watchful waiting in asymptomatic cases, and the pace of growth is generally very low after the age of 20 [5]. Conversely, surgical excision is the treatment of choice when either ventricular or valvular function is impaired or if significant arrhythmia occurs, with excellent early and late results in most cases [4]. Sudden death was reported to occur in a proportion of previously asymptomatic patients, and, therefore, the optimal management in these patients is still a matter of debate. Considering multiple tumor locations with massive septal involvement in this case, management options, including surgery, were discussed despite the asymptomatic course, good exercise tolerance, and uneventful 24-hour ECG monitoring. As the patient refused to consider an ICD as a treatment option, watchful waiting was finally recommended.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

Conflict of interest: KG is currently employed by Siemens Healthineers. The remaining authors declare no conflict of interest concerning this publication

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