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A case of hypertrophic cardiomyopathy with mid ventricular obstruction in both ventricles

Keiko Akiyama*; Michiyo Yamano; Mayumi Takeoka; Kyouko Ego; Tomoko Sakaguchi; Eiko Konisi; Nisikawa Masanobu; Tatsuya Kawasaki

***Corresponding Author: Keiko Akiyama**

Central Clinical Laboratory, Matsushita Memorial Hospital, Sotojima 5-55, Moriguchi, Osaka 570-8540, Japan.

Phone: +81-66992-1231, Fax: +81-66992-4845, Email: akiyama.keiko001@jp.panasonic.com

Abstract

Mid-ventricular obstruction (MVO) of the left ventricle is infrequently observed in patients with hypertrophic cardiomyopathy (HCM). We report a rare case of HCM with MVO in the right and left ventricles. An asymptomatic 66-year-old man was referred to our hospital for further examination of heart murmurs. On cardiac auscultation, grade 3 systolic ejection murmur was the loudest at the second left sternal border, and fourth sounds were heard at the apex and the base. Transthoracic echocardiography demonstrated myocardial hypertrophy in the middle portion of the interventricular septum, with an increased peak velocity during systole. Cardiac magnetic resonance imaging revealed a maximum wall thickness of 27 mm, in addition to small chambers in the apex of both ventricles at end-systole. A diagnosis of HCM with biventricular MVO was confirmed by transthoracic contrast echocardiography with agitated saline.

Keywords

Hypertrophic cardiomyopathy; mid-ventricular obstruction; right ventricle.

Introduction

Mid-ventricular obstruction (MVO) of the left ventricle is a rare form of hypertrophic cardiomyopathy (HCM) first reported by Falicov et al. in 1976 [1]. Several studies have investigated this condition [2,3], but little is known about MVO of the right ventricle. We report a case of HCM with both left and right ventricular MVO.

Case report

An asymptomatic 66-year-old man was referred to our hospital for further examination of heart murmurs. His previous medical history included cerebral hemorrhage 20 years earlier and idiopathic thrombocytopenia. On cardiac auscultation, grade 3 systolic ejection murmur was the loudest at the second left sternal border, and fourth sounds were heard at the apex and the base (i.e., the second right and left sternal borders) (Figure 1). The jugular venous pressure was not high and the remainder of the examination was normal. The patient was not taking medications at presentation. He did not smoke, use illicit drugs, or drink excessively, and had no known allergies. He reported that there was no family history of heart disease, including HCM, but his family history was notable for premature sudden death of his father, brother, and cousin.

Electrocardiography demonstrated a normal sinus rhythm, normal axis deviation, ST-segment elevaions in leads V1 to V2, and T wave inversions in precordial and lateral leads. No pulmonary edema or pleural effusion was present on chest radiograph. The complete blood cell counts, C-reactive protein level, and renal and liver function tests were normal, except for a platelet count of 22,000/mm^3, but this was unchanged from that measured at the annual checkup several months before this presentation. The level of brain natriuretic peptide was 177.7 pg/ml (reference ≤18.4).

Transthoracic echocardiography revealed myocardial hypertrophy in the middle portion of the interventricular septum (Figure 2). Although neither left ventricular outflow tract obstruction nor systolic anterior motion of the mitral valve was present, the peak velocity of the mid-left ventricle was 2.5 m/s. The left ventricular ejection fraction was 59%, the left ventricular end-diastolic diameter was 38 mm, the peak transmitral E wave velocity and the deceleration time of the transmitral E wave were 0.54 m/s and 238 ms, respectively, and the transmitral E/A ratio and E/early diastolic mitral annular velocity ratio of the intraventricular septum were 2.82 and 12.78, respectively. No ventricular hypertrophy of the free wall in the right ventricle was detected. A diagnosis of HCM with MVO of the left ventricle was made.

Cardiac magnetic resonance revealed myocardial hypertrophy in the mid-portion of the interventricular septum, a maximum wall thickness of 27 mm, and a small chamber in the apex of the left ventricle at end-systole (Figure 3). Of note, a small chamber was noted at the apex of the right ventricle at end-systole (Figure 3). No additional information regarding the apical chamber of the right ventricle was obtained on repeated transthoracic echocardiography because of poor imaging, but contrast echocardiography with agitated saline confirmed the presence of the small apical chamber (Figure 4). The peak velocity at the site where contrast echocardiography confirmed a MVO in the right ventricle was 2.0 m/s during systole. The patient was finally diagnosed as having HCM with biventricular MVO.

Beta-blockers were administered, and considering his history of cerebral hemorrhage, anticoagulants were not prescribed. He declined further invasive investigation or treatment. The patient has been closely monitored and has been doing well without event for more than two years.

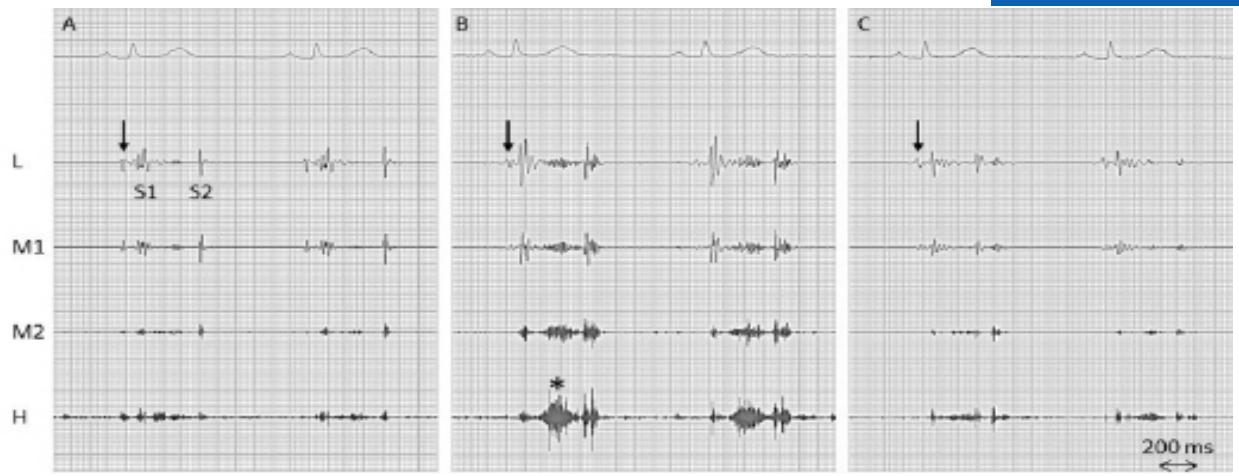

Figure 1: Phonocardiography: A phonocardiogram, obtained at the apex (A), second left sternal border (B), and second right sternal border (C) in the supine position at a paper speed of 50 mm/s, shows the fourth heart sound (arrows). A systolic ejection murmur that started after the first heart sound (S1) and ended before the second heart sound (S2) was loudest at the second left sternal border (B, asterisk). All recordings were in the same range. H: High frequency; L: Low frequency; M1: Lower-middle frequency; M2: Higher-middle frequency.

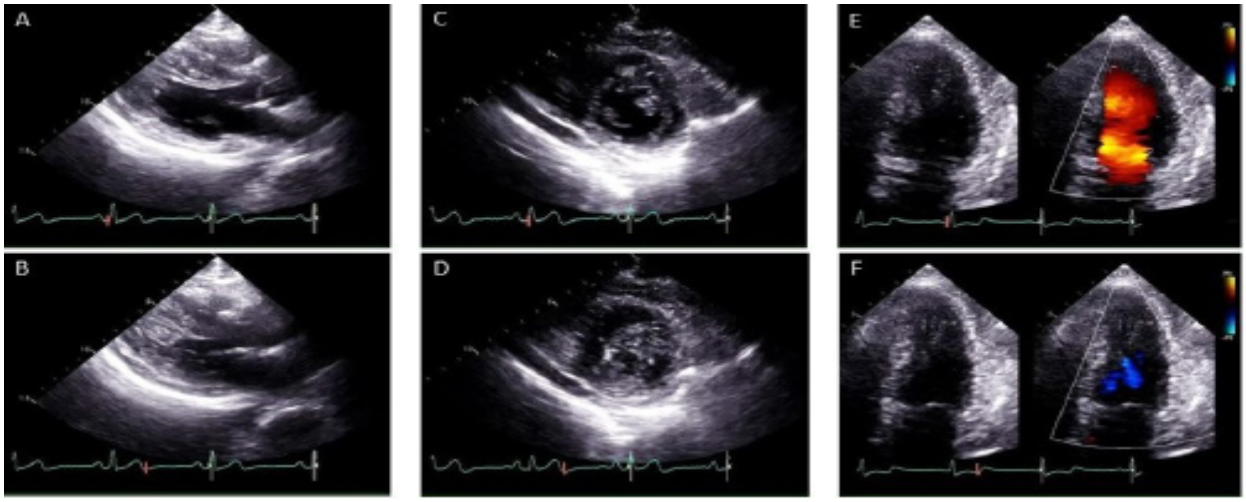

Figure 2: Transthoracic Echocardiography: Parasternal long axis views show myocardial hypertrophy in the intraventricular septum at end-diastole (A), along with mid-ventricular obstruction at end-systole (B). Myocardial hypertrophy and mid ventricular obstruction of the left ventricle are also observed in short axis images at end-diastole (C) and end-systole (D), and in four-chamber apical views with color Doppler at end-diastole (E) and end-systole (F).

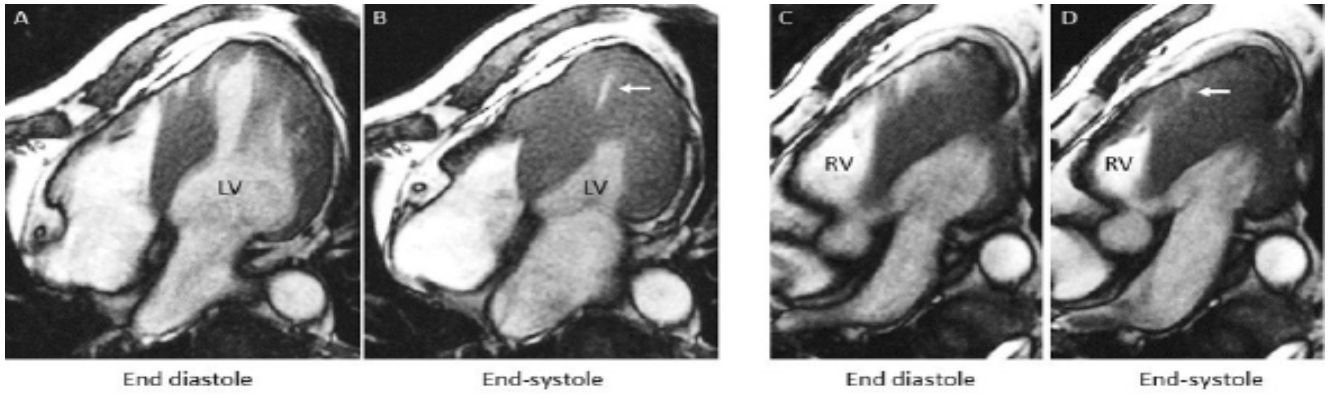

Figure 3: Magnetic Resonance Imaging: Four-chamber apical cine images at the end-diastolic phase show myocardial hypertrophy of the interventricular septum of the mid left ventricle (A), together with an apical chamber at the end-systolic phase (B, arrow). On the cine image focused on the right ventricular apex (C), a small apical chamber is also present at the end-systolic phase (D, arrows).

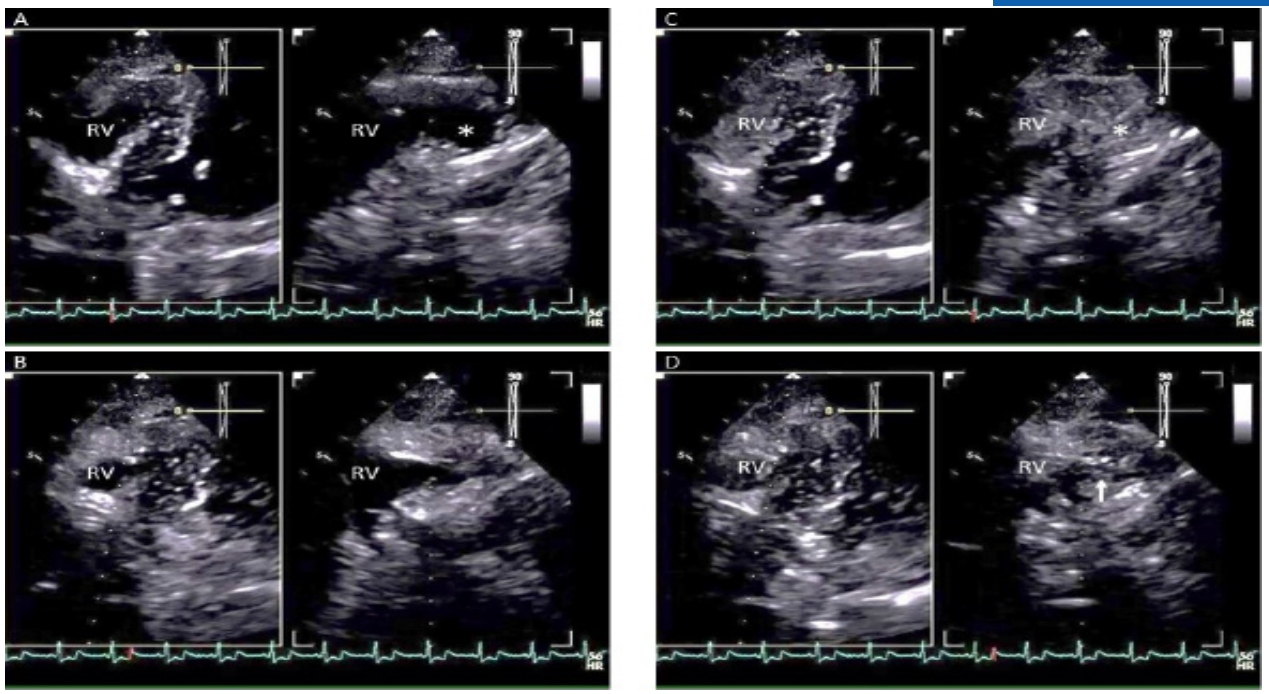

Figure 4: Contrast Transthoracic Echocardiography: Biplane images based on right ventricular modified apical four-chamber view suggest an apical chamber at end-diastole (A, asterisk), which is difficult to assess at end-systole (B). On contrast echocardiography with agitated saline, the small apical chamber is filled with micro bubbles at end-diastole (C, asterisk), and some bubbles stayed in the chamber even at the end-systole (D, arrow).

Discussion

In the current case of HCM with biventricular MVO, the diagnosis of MVO of the right ventricle was not made on initial echocardiography. Right ventricular MVO was suspected on cardiac magnetic resonance, and finally confirmed on transthoracic contrast echocardiography with agitated saline.

Left ventricular MVO, diagnosed when the peak midcavitary gradient is ≥30 mm Hg, was found in 46 (9.4%) of 490 patients with HCM [3], but the incidence of right ventricular MVO remains unclear. In a large cohort of 2,650 patients with HCM, the prevalence of right ventricular hypertrophy defined as a maximum right ventricular wall thickness of ≥10 mm was only 1.3% [4]. In another study, approximately 3% of patients with HCM exhibited right ventricular obstruction, defined as a high flow velocity signal of greater than 2.0 m/s on continuous wave Doppler echocardiography [5]. Taken together, right ventricular MVO is considered markedly rare, but it is important to note that the incidence may be underestimated because diagnosing right ventricular MVO on routine transthoracic echocardiography is difficult, as in our case.

As MVO of the left ventricle has been reported to be an independent predictor of adverse outcomes [3], early recognition of this condition is needed, although the effects of right ventricular MVO on the prognosis remain uncertain in patients with HCM. It is well known that apical aneurysms in the left ventricle, which are often caused by left ventricular MVO, are associated with significant cardiovascular morbidity and mortality [6]. Considering the weakness of the right ventricle against pressure overload, right ventricular MVO may more frequently lead to the development of apical aneurysms in the right ventricle than left ventricular MVO, although the present patient had no aneurysm in either ventricle. Further studies are required to examine the clinical significance of right ventricular MVO in patients with HCM.

S4 is commonly heard in HCM patients with sinus rhythm; in our previous report, S4 assessed by phonocardiography was observed in 75% of patients [7]. Of note, S4 is usually heard best at the apex [8] because it is considered as a result of the rapid inflow of blood by atrial contractions [9]. In our case, S4 was audible not only at the apex but also at the base, even at the second right sternal border, which was clearly recorded on phonocardiography, consistent with right-sided S4 reflecting right ventricular diastolic dysfunction [10]. Greater attention to cardiac auscultation – the distribution of S4 – may have led to the early identification of right ventricular MVO. Furthermore, a systolic ejection murmur along the high left sternal border, as observed in our case, often indicates subpulmonic or MVO to the right ventricular outflow [11,12]. As diastolic murmurs are not uncommon in HCM patients with left ventricular MVO [13-15], the presence and distribution of a diastolic murmur may also suggest right ventricular MVO, although no diastolic murmurs were heard in our case.

We reported a case of HCM with biventricular MVO, in which the diagnosis of right ventricular MVO was suspected on cardiac magnetic resonance and confirmed by contrast echocardiography. Careful physical examination may help in the diagnosis of this rare condition even in the era of advanced imaging technologies.

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Authors Information: Keiko Akiyama[1]*; Michiyo Yamano[2]; Mayumi Takeoka[1]; Kyouko Ego[1]; Tomoko Sakaguchi[1]; Eiko Konisi[1]; Nisikawa Masanobu[1]; Tatsuya Kawasaki[2]

[1]Central Clinical Laboratory, Matsushita Memorial Hospital, Osaka, Japan.

[2]Department of Cardiology, Matsushita Memorial Hospital, Osaka, Japan.

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