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Case Report

A natural history of apical hypertrophic cardiomyopathy with development of an apical aneurysm formation: A case report following a quarter century

Keisuke Kawai (MD)[a,*], Akiko Taji (RMS)[b], Akihiro Takahashi (MD)[a], Tatsuya Kuniyoshi (MD)[a], Toshiyuki Yamatani (MD)[a], Ken-ichi Hirata (MD)[c]

[a] Department of Internal Medicine, Kasai City Hospital, Kasai, Hyogo, Japan
[b] Department of Clinical Laboratory, Kasai City Hospital, Kasai, Hyogo, Japan
[c] Division of Cardiovascular Medicine, Department of Internal Medicine, Kobe University Graduate School of Medicine, Hyogo, Japan

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ABSTRACT

We report the definite long-term natural history of a man with apical hypertrophic cardiomyopathy (ApHCM), who developed an apical aneurysm by the age of 60 years. At 33 years, his electrocardiogram (ECG) was nearly normal. T-wave inversions were first identified in leads V5–6 of an ECG at 36 years of age. Echocardiography first demonstrated a left ventricular (LV) lateral wall hypertrophy at 37 years of age. At 42 years, echocardiography showed the pattern of diastolic spade-like appearance of the LV cavity, confirming the ApHCM diagnosis. At 54 years, a late-peaking apical systolic gradient of 10 mmHg (velocity: 1.6 m/s) emerged. At 60 years, he had developed an apical aneurysm. Cardiac computed tomography revealed no significant major coronary artery stenosis. Thus, in our case, LV hypertrophy and apical high pressure in ApHCM advanced the formation of an apical aneurysm in the absence of coronary artery disease.

<Learning objective: High apical systolic flow velocity of ≥1.5 m/s in apical hypertrophic cardiomyopathy (ApHCM) using echocardiography may be predictive of the development of apical aneurysm, and make us consider medical intervention by beta-blockers or verapamil to delay or prevent aneurysm formation. Because recent studies have demonstrated that ApHCM may be less benign than previously suspected, observation without medical attention for ApHCM might be hazardous.>

Introduction

Apical hypertrophic cardiomyopathy (ApHCM) is a variant of hypertrophic cardiomyopathy. ApHCM is characterized by myocardial hypertrophy occurring predominantly in the left ventricular (LV) apical portion, with giant negative T waves (>10 mm) in the left precordial leads of the electrocardiogram (ECG) [1]. Although patients with ApHCM sometimes develop apical aneurysm, the definite long-term natural history of ApHCM has not been clarified.

Case report

Here, we report the case of a 61-year-old man without a history of essential hypertension and with a history of diabetes, bronchial asthma, and duodenal ulcer. He had no family history of sudden death or hypertrophic cardiomyopathy. The patient measured 163 cm in height and 53 kg in weight. His blood pressure was 106/80 mmHg and the heart rate was 55 beats/min.

The natural history of the patient from 33 years of age to 61 years follows:

At 33 years of age, the patient's ECG was nearly normal (Fig. 1).

At 36 years of age, T-wave inversions were first identified in the left precordial leads V5–6 of the ECG during a routine examination of the patient. No abnormality was documented at that time by echocardiography.

At 37 years of age, with T-wave inversions in leads V4–6 of the ECG (Figs. 1 and 2), the echocardiography first demonstrated

* Corresponding author at: Department of Internal Medicine, Kasai City Hospital, 1-13, Yoko-o, Hojo-cho, Kasai, Hyogo 675-2311, Japan. Tel.: +81 790 42 2200; fax: +81 790 42 3460.
E-mail address: kkawai@river.sannet.ne.jp (K. Kawai).

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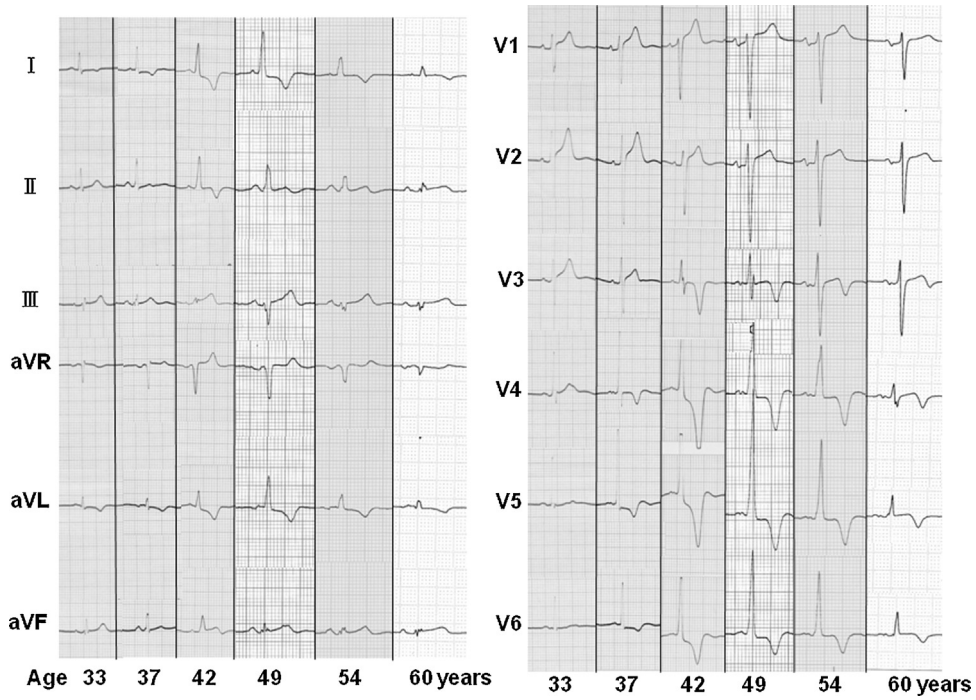

Fig. 1. Serial 12-lead electrocardiogram recording, showing the development and attenuation of deep T-wave inversions in leads I, aVL, and V3–6 with sinus rhythm.

LV lateral wall hypertrophy, with a wall thickness of 13 mm (Fig. 3).

At 42 years of age, the left precordial R waves and negative T waves reached their maximal amplitude (Figs. 1 and 2). Echocardiography showed the pattern of diastolic spade-like appearance of the LV cavity with severe apical hypertrophy, confirming the diagnosis of ApHCM. The early diagnosis of ApHCM can be achieved by identifying the hypertrophy that is frequently confined to the lateral wall at the apical level using magnetic resonance imaging [2]. However, in this case, due to the limited image quality of early echocardiographic studies, the severity of the LV lateral wall hypertrophy was assessed in the parasternal short-axis plane at the level of the papillary muscles using echocardiography (Fig. 3). As shown in Fig. 3, his lateral wall hypertrophy at the papillary muscles level developed over a 25-year period, but he did not demonstrate basal interventricular septal hypertrophy of ≥15 mm. Therefore, he was classified as having the apical phenotype with the "distal-dominant" form [3]: apical hypertrophy extended to the lateral wall and interventricular septum beyond the papillary muscles level without basal septal hypertrophy.

At 54 years of age, a late-peaking apical systolic gradient of 10 mmHg (velocity: 1.6 m/s) and a diastolic gradient from apex to base, which generated a paradoxical jet flow [4], developed between the apical and basal chambers (Figs. 3 and 4).

At 55 years of age, a non-sustained ventricular tachycardia of four consecutive extrasystoles was revealed in the Holter recording.

At 60 years of age, a follow-up echocardiogram demonstrated that the patient had developed an apical aneurysm. Enhanced computed tomography (CT) also showed an apical aneurysm formation

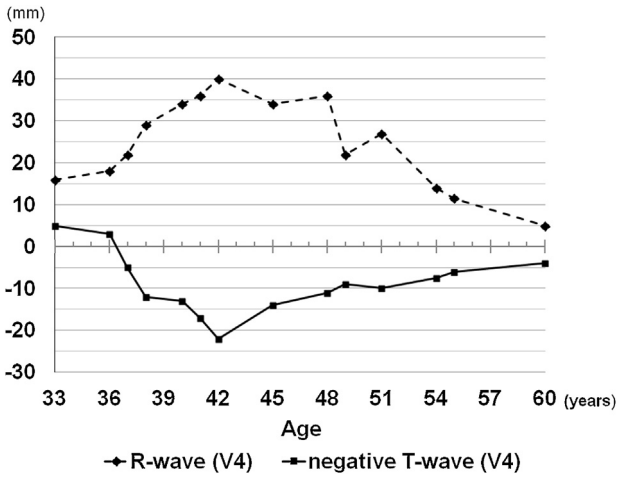

Fig. 2. Changes in electrocardiogram in lead V4 with age. Depth of the negative T-wave in lead V4 initially increased and then decreased. The presence of giant negative T-waves (>10 mm) was shown for the first time at 38 years of age. R-wave voltage also progressively increased and then decreased.

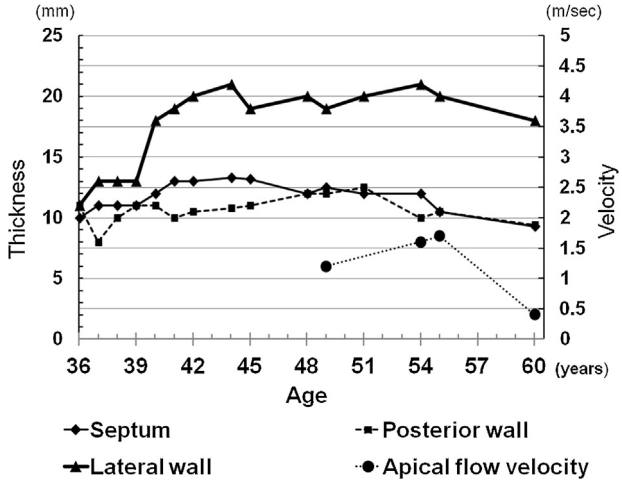

Fig. 3. Changes in echocardiogram measurements with age, showing mild thickening of the basal interventricular septum (♦) and the basal left ventricular posterior wall (■). Lateral wall hypertrophy at the level of the papillary muscles (▲) developed over a 25-year period. Using Doppler measurement, the apical systolic flow velocity (●) was up to 1.7 m/s (pressure gradient: 12 mmHg) at 55 years of age.