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Hypertrophic Cardiomyopathy With Dominant Hypertrophy in the Right Anterobasal Region of the Ventricular Septum: A Case Report

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Abstract

A 62-year-old man was referred to our hospital for investigation of abnormal electrocardiography findings. The mean frontal plane QRS axis was directed toward the right superior quadrant(−125 °). Terminal S waves were present in all 3 bipolar standard leads and an R wave in lead aV_R. RS complex was seen in lead V_1 and deep S waves in leads V_2−V_6. Left ventricular hypertrophy associated with asymmetrical septal hypertrophy was suspected based on transthoracic echocardiography, but the echocardiographic quality was poor. Magnetic resonance imaging revealed hypertrophic cardiomyopathy with massive wall thickening involving the right anterobasal region of the ventricular septum. Magnetic resonance imaging may provide useful information about the distribution of ventricular myocardial hypertrophy in patients with hypertrophic cardiomyopathy and unusual electrocardiography findings.

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Key Words
■ **Cardiomyopathies, hypertrophic** ■ **Magnetic resonance imaging**
■ **Electrocardiography**

INTRODUCTION

Standard 12-lead electrocardiography(ECG)may show a great variety of abnormalities in patients with hypertrophic cardiomyopathy[1−3]. ECG is important for the first diagnostic screening of this disease, but new diagnostic modalities, such as magnetic resonance(MR)imaging, MR spectroscopy and positron emission computed tomography, have greatly contributed to the diagnosis of hypertrophic cardiomyopathy and to the interpretation of its morphological and pathophysiological features[4]. We describe a case of hypertrophic cardiomyopathy with unusual ECG findings, and discuss the significance of the changes in relation to the MR imaging findings.

CASE REPORT

A 62-year-old man was referred to our hospital for investigation of abnormal ECG findings(**Fig. 1**). He had no history of cardiac or pulmonary disease. Chest radiography showed the cardiac silhouette was normal. Transthoracic echocardiography could not clearly show the details of the heart in the parasternal echocardiographic window because of limitation of the window by the adjacent lung tis-

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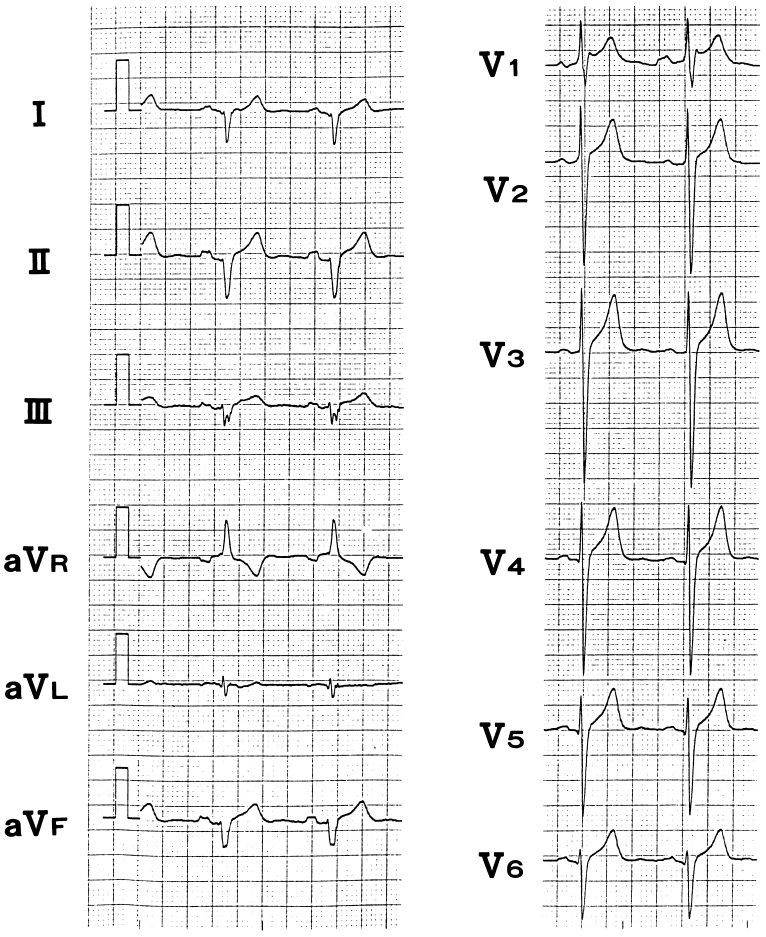

Fig. 1 Electrocardiographic findings
The mean frontal plane QRS axis is directed toward the right superior quadrant (−125 °). Terminal S waves are present in all 3 bipolar standard leads and an R wave is seen in lead aVR. An RS complex is seen in lead V_1, and deep S waves are evident in V_2–V_6.

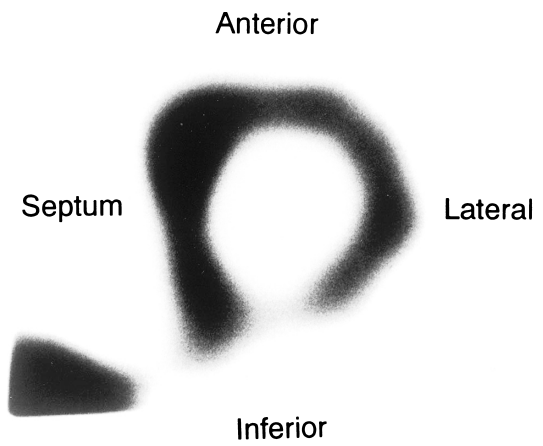

Fig. 2 Rest-gated single photon emission computed tomography scan with technetium-99m tetrofosmin
The basal short-axis slice of the left ventricle shows uptake of technetium-99m tetrofosmin is increased in the right anterobasal region of the ventricular septum.

sue, but the apical view showed slight indications of the presence of left ventricular hypertrophy associated with asymmetrical septal hypertrophy. The difficulty in the echocardiographic evaluation seemed to be due to the muscular physique of the patient. Single photon emission computed tomography with technetium-99m tetrofosmin revealed myocardial hypertrophy localized in the anterobasal portion of the ventricular septum (**Fig. 2**). MR imaging was performed to evaluate the ventricular hypertrophy accurately, and revealed hypertrophic cardiomyopathy with massive wall thickening involving the right anterobasal region of the ventricular septum (**Fig. 3**). This finding was considered to be consistent with the unusual ECG changes. The patient has remained clinically stable and asymptomatic, and his ECG findings have also remained unchanged for the past 5 years.

DISCUSSION

Standard 12-lead ECG shows abnormal findings in 85% to over 90% of patients with hypertrophic

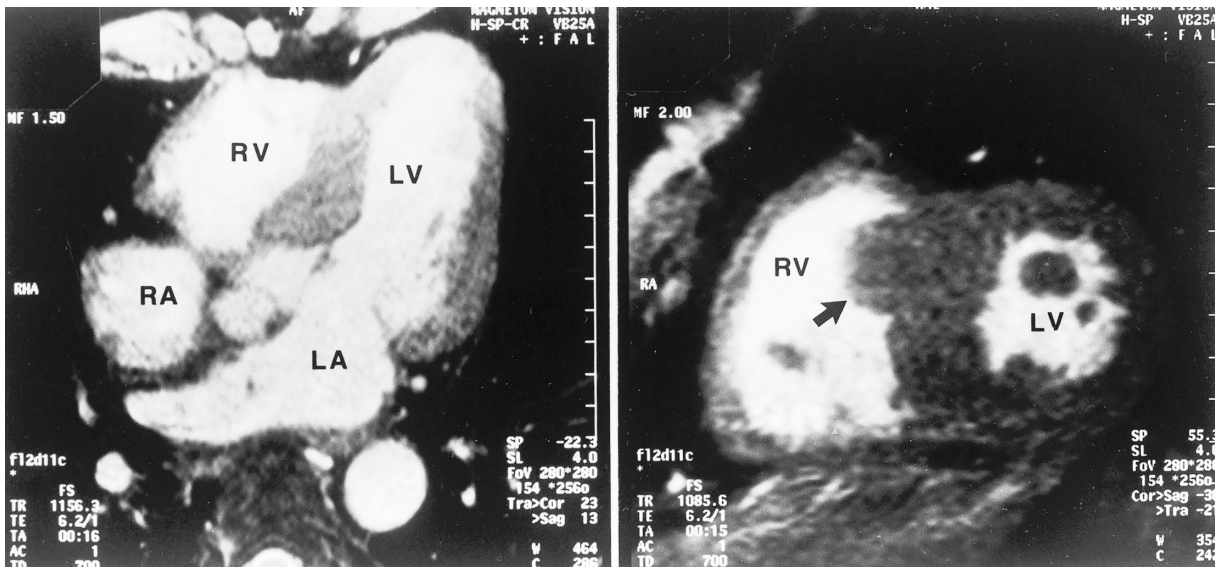

Fig. 3 Magnetic resonance image showing ventricular hypertrophy with asymmetrical septal hypertrophy
The right anterobasal region of the ventricular septum is markedly thickened and protrudes into the right ventricular outflow tract (*arrow*).
RV = right ventricle; RA = right atrium; LV = left ventricle; LA = left atrium.

cardiomyopathy[1–3]. Deviation of the mean frontal plane QRS axis superiorly and to the right is rare, but occurs in 2% of patients with hypertrophic cardiomyopathy[1]. The significance of such uncommon QRS axis deviation is still unclear. Wigle *et al.*[5] reported the effects of ventriculomyotomy in 3 patients with hypertrophic cardiomyopathy and abnormal hypertrophy was located mainly in the anterior end of the septum and the immediately adjacent left ventricular anterior wall. The septal hypertrophy may have resulted in dominance of anterosuperior over posteroinferior electrical forces that could be altered by septal incision. This patient had localized hypertrophy of the anterobasal region of the septum, so the electrical force of septal activation was spread to the right, anteriorly and inferiorly, and produced a small q wave in leads I and aV_L, a small r wave in leads II, III and aV_F, an R wave in the right precordial chest leads, and a q wave in the left precordial leads. The QRS complexes caused by the spread of excitation through the 2 ventricles were considered to be modified by the localized hypertrophy of the anterobasal septum, which would cancel the normal frontal QRS axis in the limb leads and produce deep terminal S waves in all limb leads (except aV_R) and in the precordial leads (except V_1).

Echocardiography is widely accepted as a useful tool for the diagnosis of hypertrophic cardiomyopathy, but hypertrophy of a localized region of the basal ventricular septum is an incidental echocardiographic finding in most cases[6]. Maron *et al.*[7] observed hypertrophy limited to the anterior septum (type I) in 12 (10%) of 125 patients with hypertrophic cardiomyopathy. The prevalence of normal ECG in the patients with this type I was significantly greater than in the other 3 morphologic types[2]. However, in the majority of their type I patients[7], hypertrophy was not limited to the ventricular septum and extended to the apical portion of the left ventricle. Belenkie *et al.*[6] emphasized that hypertrophy localized to the basal septum infrequently represented part of the spectrum of hypertrophic cardiomyopathy. We speculate that the unusual ECG findings in the present case reflect the massive bulge of myocardium localized in the right anterobasal region of the ventricular septum.

The advantages of MR imaging over echocardiography include a large field of view that provides more accurate diagnostic information in almost all patients[8,9]. The present case of the use of MR imaging to identify hypertrophic cardiomyopathy with dominant hypertrophy in the right anterobasal region of the ventricular septum indicates that MR

imaging can yield additional insight into the features of hypertrophic cardiomyopathy in relation to specific ECG abnormalities which might be impossible to clarify by echocardiography.

要　　約

心室中隔基部の右室側前方に著しい肥大を伴った肥大型心筋症の1例

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症例は62歳の男性で，心電図異常の精査のため当科を紹介され受診した．平均前頭面QRS電気軸は右上方に向かい（−125 °），3つすべての双極標準誘導でS波を，またaV$_R$誘導ではR波を認めた．胸部誘導V$_1$はRS型を示し，深いS波をV$_2$−V$_6$で認めた．経胸壁心エコー図で非対称性中隔肥厚を伴う左室肥大の存在が疑われたが，エコー図での描出は不良であった．磁気共鳴画像で心室中隔基部の右室側前方に，著しい限局的な肥厚を伴った肥大型心筋症であることが明らかになった．特異な心電図所見を伴う肥大型心筋症において，その心室の肥大部位に関する情報を得るうえで，磁気共鳴画像検査は有用と思われた．

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