Hypertrophic Cardiomyopathy in Two Elderly Siblings

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SUMMARY

Hypertrophic cardiomyopathy, which is sometimes familial and genetically transmitted as an autosomal dominant trait, is generally regarded as a disease of young or middle aged and relatively few cases have been reported in elderly patients. The present communication describes the oldest known siblings with clinically diagnosed hypertrophic cardiomyopathy, 80 and 78 years of age.

Additional Indexing Words:

Hypertrophic cardiomyopathy Elderly siblings Echocardiography

HYPERTROPHIC cardiomyopathy is not an uncommon disease, but, it is generally regarded as a disease of young or middle aged and relatively few cases have been reported in elderly patients.¹⁾⁻⁸⁾ The present communication describes the oldest known siblings with clinically diagnosed hypertrophic cardiomyopathy.

CASE REPORTS

Case 1; An 80-year-old woman was first admitted to the Inami-Kousei Hospital in June 1979, with low grade fever, cough, and fatigue. Her past history and precise family history were unremarkable, but she had never experienced episode of dyspnea, syncope, or angina and was not taking any cardiac medications. The physical examination on admission revealed a blood pressure of 126/66 mmHg, regular pulse at rate of 72 per minute, brisk-

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Fig. 1. Chest roentgenogram of Case 1 shows enlarged cardiac silhouette (cardiothoracic ratio: 63%) and an infiltrate in the left lower lobe.

Fig. 2. Electrocardiogram of Case 1 shows left axis deviation and prominent Q wave in lead V₅ and V₆.

Fig. 3. Phonocardiogram before and after inhalation of amyl nitrite in Case 1. A marked increase in the intensity of murmur is demonstrated after amyl nitrite inhalation.

Fig. 4. Apexcardiogram and carotid pulse tracing before and after amyl nitrite inhalation in Case 1. A large A wave amplitude, diminished rapid filling wave and mid-systolic dip of carotid pulse tracing after amyl nitrite inhalation (indicated by arrow) are shown.

rising carotid impulse. A bifid impulse was felt at the left anterior axillary line in the 5th intercostal space. A grade 4/6 coarse systolic ejection murmur was heard loudest at the low left sternal border, radiated to the apex and faintly to the base. The chest roentgenogram showed enlarged cardiac silhouette (cardiothoracic ratio: 63%) and an infiltrate in the left lower lobe (Fig. 1). The electrocardiogram showed left axis deviation and prominent Q waves in lead V_5 and V_6 (Fig. 2). A marked increase in intensity of the murmur was demonstrated after amyl nitrite inhalation (Fig. 3).

The apexcardiogram showed large amplitude of A wave and diminished rapid filling wave, and the carotid pulse tracing following amyl nitrite inhalation revealed mid-systolic dip (Fig. 4). The ejection period and isovolumetric contraction time corrected for heart rate by dividing the square root of the R-R interval were 330 msec and 95 msec respectively, longer than those

Fig. 5. Echocardiogram and ultrasonotomocardiogram in Case 1 reveal asymmetric septal hypertrophy, systolic anterior movement of anterior mitral valve and partial mid-systolic closure of aortic valve (indicated by arrow). Abbreviation: IVS=inter-ventricular septum; PW=left ventricular posterior wall; LV=left ventricle; LA=left atrium; SAM=systolic anterior movement of anterior mitral valve; MV=mitral valve.

found in normal subjects in our laboratory.⁹⁾ The echocardiogram showed marked asymmetric hypertrophy of inter-ventricular septum, small left ventricular cavity (Dd: 3.8 cm, Ds: 2.0 cm), systolic anterior motion and slowing of the early diastolic closing motion of the anterior mitral valve.

The aortic valve echogram showed mid-systolic partial closure (Fig. 5). The thickness of inter-ventricular septum and the postero-basal region of free wall at end-diastole were 2.6 cm and 1.1 cm respectively. Two-dimensional echocardiogram revealed disproportionate thickening of the inter-ventricular septum (Fig. 5).

Case 2; A 78-year-old sister of Case 1 was admitted to the Inami-Kousei Hospital in April 1979, with epigastric pain. Her past history was uncertain, but she had never experienced dyspnea, angina, or syncope and never been under medical care elsewhere before this admission.

Physical findings disclosed a blood pressure of 122/70 mmHg, the irregular pulse at rate of 72 per minute and the brisk-rising carotid pulse. The chest was clear. The heart was enlarged with double apical impulse. A grade 2/6 systolic ejection murmur and faint diastolic murmur were heard loudest at the higher right sternal border (Fig. 6). Electrocardiogram showed left ventricular hypertrophy (Fig. 7). The chest roentgenogram (Fig. 8)

Fig. 6. Phonocardiogram in Case 2 shows systolic ejection murmur at the higher right sternal border.

Fig. 7. Electrocardiogram of Case 2 shows left ventricular hypertrophy.

showed enlarged cardiac silhouette (cardiothoracic ratio: 60%). The indirectly recorded carotid arterial pulse rose sharply and large A wave amplitude of apexcardiogram was demonstrated. The configuration of carotid pulse tracing and apexcardiogram following the premature atrial contraction did not change significantly (Fig. 9). Echocardiogram and two-dimensional echocardiogram revealed disproportionate thickening of inter-ventricular septum, however systolic anterior motion of anterior mitral valve and mid-systolic closure of aortic valve were not demonstrated. The thickness of inter-ventricular septum and postero-basal region of left ventricular free wall at end-diastole was 2.6 cm and 1.0 cm respectively (Fig. 10).

Fig. 8. Chest roentgenogram of Case 2 shows enlarged cardiac silhouette (cardiothoracic ratio: 60%).

Fig. 9. Apexcardiogram and carotid pulse tracing of Case 2 show a large A wave amplitude and diminished rapid filling wave.

DISCUSSION

Hypertrophic cardiomyopathy, which is sometimes familial and genetically transmitted as an autosomal dominant trait,¹⁰⁾⁻¹²⁾ is a disease of cardiac muscle characterized by disproportionately thickened interventricular septum compared with the postero-basal left ventricular free wall.^{10),12)-15)} Although some of these patients have obstruction to left ventricular outflow tract due to systolic anterior motion of the anterior mitral valve^{10),13),16)-18)} or markedly hypertrophied and vigorously contracting ventricular septum,¹⁹⁾ the presence or absence of obstruction to ventricular outflow may not represent a fundamental difference.^{10),13),14)} The hypertrophic cardiomyopathy in young or middle-aged patients is easily diagnosed, but that in aged patients frequently missed because of the similarity or coexistence of coronary artery disease or

Fig. 10. Echocardiogram and ultrasonotomocardiogram of Case 2 show an asymmetric septal hypertrophy but no signs that indicate the obstruction to left ventricular outflow are revealed. Abbreviation: IVS=inter-ventricular septum; LV=left ventricle; LA=left atrium; MV=mitral valve; PW=left ventricular posterior wall.

hypertensive heart disease.^{5),11),20)-22)}

However, recent studies using echocardiography have shown a characteristic sign of hypertrophic cardiomyopathy and made it possible to diagnose in elderly patients. The echocardiographic features of hypertrophic cardiomyopathy include asymmetric septal hypertrophy,^{10),12)-16)} anterior displacement of the mitral apparatus,¹⁹⁾ systolic anterior movement of mitral valve,^{10),16)-19)} reduced velocity of early diastolic closing motion of anterior mitral leaflet,¹⁴⁾ inter-ventricular septal hypokinesia,²³⁾ and mid-systolic closure of the aortic valve.²⁴⁾ Though the cardiac catheterization and left ventriculography were not performed in our 2 cases, the characteristic echocardiographic features consisting of systolic anterior motion of anterior mitral valve, mid-systolic closure of aortic valve in Case 1 and asymmetric septal hypertrophy of Case 1 and Case 2 confirmed the diagnosis of hypertrophic cardiomyo-

pathy. The systolic anterior movement of anterior mitral valve, mid-systolic closure of aortic valve, mid-systolic dip of carotid pulse tracing following the amyl nitrite inhalation and prolonged left ventricular ejection time index and isovolumetric contraction time index in Case 1 strongly suggested the presence of left ventricular outflow tract obstruction in this case.

In Case 2, asymmetric septal hypertrophy was shown by echocardiogram, but there were no signs that suggested the obstruction to left ventricular outflow tract. The etiology of diastolic murmur observed in Case 2 was uncertain, but Frank reported that this type of murmur was present in 7 of 126 patients¹¹⁾ and Hara reported early diastolic murmur of unknown etiology in 1 of 5 patients with idiopathic hypertrophic cardiomyopathy.²⁵⁾ Although the cause of hypertrophic cardiomyopathy is still unknown, the occurrence of obstructive and non-obstructive cardiomyopathies in two elderly siblings suggest that the disease is familial and the presence or absence of obstruction is merely a different manifestation of the same basic cardiac disease.^{10), 12)–14), 26)}

In recent years, the reports of hypertrophic cardiomyopathy in the advanced age group have appeared in the literature with increasing frequency, but there are few reports concerning elderly siblings with hypertrophic cardiomyopathy. Sanders reported 2 sisters with hypertrophic obstructive cardiomyopathy, 68 and 63 years of age.¹⁾ These 2 cases in siblings unusually lived long as patients with hypertrophic obstructive cardiomyopathy, but the present 2 cases are the oldest known siblings with clinically demonstrated hypertrophic cardiomyopathy.

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