Inspiratory Right Ventricular Outflow Obstruction in a Patient with Hypertrophic Cardiomyopathy

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Summary

A patient with familial hypertrophic cardiomyopathy with exertional near syncope is reported. Intra-right ventricular obstruction was demonstrated by hemodynamic studies during inspiration and the Valsalva maneuver with systemic hypotension. Improvement occurred following the administration of propranolol. It was suggested that syncope might be precipitated by hemodynamic changes such as a high output state and a depressed cardiac volume in relation to intra-right ventricular obstruction in patients with hypertrophic cardiomyopathy.

Additional Indexing Words:
Familial hypertrophic cardiomyopathy Exercise induced near syncope Respiratory change Valsalva maneuver Adrenergic beta blockade

It has been recognized that sudden death occurs in patients with hypertrophic cardiomyopathy, especially those having syncope. Although various arrhythmias, myocardial infarction and acute hemodynamic changes such as increased left ventricular outflow obstruction have been reported, the cause of death remains unclear.

We present a rare case with familial hypertrophic cardiomyopathy, exercise induced near syncope and palpitations, where intra-right ventricular obstruction was demonstrated during deep inspiration and by the Valsalva maneuver.

Case Report

A 16 year old student had suffered from chest discomfort, dizziness and...
palpitation upon exercise 6 years ago and his local doctor was consulted. The diagnosis of hypertrophic cardiomyopathy was made and he took the beta blocker, propranolol 30 mg/day, following which his symptoms soon disappeared.

In 1985, he experienced general fatigue and exercise-induced near syncope 2 or 3 times a week. He was transferred to our hospital for further evaluation. It was learned that his father and uncle had died suddenly at the ages of 35 and 45, respectively. His sister and aunt also had been confirmed as having severe concentric left ventricular hypertrophy by echocardiogram. He was admitted to our hospital in December, 1986.

Clinical findings and noninvasive cardiac examination:

On admission, pulse was 72/min and regular. Blood pressure was 140/60 mmHg. A fourth heart sound was audible with a grade 3/6 systolic musical murmur along the lower left sternal border which radiated to the apex. In deep inspiration, the murmur was markedly increased and systolic blood pressure fell to 70 mmHg. The lungs were clear to auscultation and percussion. Otherwise, physical examination and laboratory tests were all normal.

Chest roentgenogram was normal except for an enlargement of the cardiac margin. Electrocardiogram at rest showed a giant negative T wave in leads I, II, III, aVF and V3-V6 with high QRS voltage. A septal q wave was not recognized.

Echocardiogram (Fig. 1) demonstrated a diffuse concentric hypertrophy of the left ventricle, but mid-systolic closure of the aortic valve and systolic anterior movement of the mitral valve were not seen. The right ventricle was also narrowed by a hypertrophied interventricular septum and muscle bands. In Doppler studies high speed flows in the right ventricular outflow tract enhanced by inspiration were recorded, but the exact pressure gradient could not be measured because of the beat to beat movement of the heart. 201Tl myocardial scintigraphy demonstrated a hypertrophied biventricle.

Cardiac catheterization and angiographic findings:

Hemodynamic data before and 5 months after the administration of propranolol 60 mg/day are shown in Fig. 2. During inspiration the right ventricular pressure was increased but pulmonary arterial pressure was decreased, so that the pressure gradient between these two points was about 18 mmHg (Fig. 2A). After the administration of propranolol, the pressure gradient across the right ventricular outflow tract disappeared (Fig. 2B). But during the Valsalva maneuver the pressure gradient reappeared and the systemic systolic blood pressure fell to 65 mmHg (Fig. 2C). Pressure values otherwise were normal with the exception of a slightly elevated left
ventricular end-diastolic pressure. Right ventriculography (Fig. 3) demonstrated a severely hypertrophied right ventricle, especially the moderator bands. It is suggested that the pressure gradient is produced by these abnormal hypertrophied structures. Left ventriculography also showed an extreme concentric hypertrophy and a thinning of the left ventricle in the systolic phase. Coronary arteriogram did not demonstrate significant organic stenosis.

Myocardial biopsy showed a typical pattern of hypertrophic cardiomyopathy with disarray and fibrosis. The patient's symptoms were controlled by propranolol, 60 mg/day and he was discharged from our hospital.

DISCUSSION

Syncope in hypertrophic cardiomyopathy has been explained on the basis of various pathophysiological mechanisms\(^1\)-\(^5\) and is closely related to sudden death. On the other hand, right ventricular obstruction in hypertrophic cardiomyopathy has been reported\(^6\)-\(^8\) but it has not been clear whether or not right ventricular obstruction is related to syncope.

In our case, right ventricular obstruction was demonstrated during deep inspiration and by the Valsalva maneuver but not at rest. The pressure gradient can be explained on the basis of the law proposed by Venturi, described below. The right ventricular outflow tract wall is anatomically thin.
Fig. 2. Right ventricular (RV), pulmonary arterial (PA) and femoral arterial (FA) pressure curves during inspiration before (A) and after (B) the administration of propranolol, and Valsalva maneuver (C) after propranolol.

Fig. 3. Right ventriculogram in diastole (a), systole (b) and its schematic representation (right) during inspiration. Hypertrophied moderator bands visualized in the systolic phase. RVOT=right ventricular outflow tract; RV=right ventricle; RA=right atrium; LAO=left anterior obliquation 40.
and may yield to a negative pressure caused by an increased flow rate in the right side of the heart at the time of inspiration. During exercise, increased cardiac output and ventricular contractility will also produce a pressure gradient in the same manner.

Pressure gradient attenuation during the Valsalva maneuver in patients with idiopathic subaortic stenosis has been reported. It has been suggested that decreased right ventricular volume during this maneuver caused an obstruction, as mentioned, on the left ventricle.

Adrenergic beta blockade should be thought of as an effective therapy for hypertrophic cardiomyopathy with right as well as left ventricular outflow obstruction because of its negative inotropisms.

In summary, we reported on a patient with hypertrophic cardiomyopathy with right ventricular obstruction at the time of deep inspiration and the Valsalva maneuver which was suspected to be related to exertional syncope.

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REFERENCES