CASE REPORTS

Myocardial Stunning in Hypertrophic Cardiomyopathy: Recovery Predicted by Single Photon Emission Computed Tomographic Thallium-201 Scintigraphy

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A young woman with hypertrophic cardiomyopathy confirmed by echocardiography and cardiac catheterization presented with chest pain and features of a large left ventricular aneurysm. The initial diagnosis was myocardial ischemia with either an evolving or an ancient myocardial infarction. Subsequently, verapamil therapy was associated with complete resolution of the extensive left ventricular wall motion abnormalities, normalization of left ventricular ejection fraction and a minimal myocardial infarction.

Hypertrophic cardiomyopathy is characterized by asymmetric myocardial hypertrophy, myofibrillar disarray, dynamic left ventricular outflow obstruction and decreased left ventricular compliance (1). The electrocardiogram (ECG) frequently shows abnormal Q waves suggestive of myocardial infarction. Although patients with this disorder are prone to sudden death (2) and have described symptoms of dyspnea, syncope, palpitation and chest pain, myocardial infarction has been described only rarely. We describe extensive spontaneous ischemia with subsequent resolution of the ischemia and associated severe wall motion abnormalities in a young woman with hypertrophic cardiomyopathy and angiographically normal coronary arteries.

Case Report

A 32 year old white woman was admitted because of vague substernal heaviness associated with generalized weakness and exertional dyspnea lasting 12 h. She described three similar episodes in the past. The first was 1 year earlier after she received oxytocin and methylergonovine in the immediate postpartum period. That episode was associated with hypotension, bradycardia and dizziness. An ECG at that time showed small Q waves in the inferolateral leads and ST-T wave abnormalities. The patient recovered spontaneously and did well until 3 weeks before admission, when she had an episode of nonexertional chest heaviness lasting about 2 h. Another brief episode occurred the night before admission.

Many years earlier, she had been told she had a heart murmur. There was no history of hypertension, hyperlipidemia, diabetes, tobacco use, alcohol or drug abuse, oral contraceptive use, syncope or palpitation and there was no family history of cardiac disease.

Physical examination. The patient, a tall, slender woman, had a blood pressure of 92/60 mm Hg, a heart rate of 64 beats/min and a respiratory rate of 14 breaths/min. The apex beat was enlarged and dyskinetic. The jugular venous pressure was estimated to be 11 cm H2O. The first and second heart sounds were normal. A third heart sound was present. There was a grade 3/6 apical systolic murmur that radiated to the left sternal border and axilla. The remainder of the findings were normal. Sublingual nitroglycerin exacerbated the patient's chest discomfort.

Initial evaluation. The ECG showed normal sinus rhythm and evidence suggesting an acute inferolateral myocardial infarction (Fig. 1). A chest radiograph was normal. An echocardiogram (Fig. 2) revealed an apical aneurysm. Asymmetric septal hypertrophy, hyperdynamic basal left ventric-
Figure 1. Electrocardiogram on admission. Figure 3. Second electrocardiogram showing progression with possible inferolateral myocardial infarction.

Systolic function, systolic anterior motion of the mitral valve and premature aortic valve closure. Doppler examination of the left ventricular outflow tract was consistent with a dynamic obstruction with a peak gradient of 10 to 15 mm Hg. Mild mitral regurgitation was also recorded. The overall left ventricular cavity size was normal, and the ejection fraction was estimated to be approximately 45%. The findings were consistent with hypertrophic obstructive cardiomyopathy with an apical left ventricular aneurysm. Review of an echocardiogram recorded at another institution 3 years earlier showed findings of hypertrophic cardiomyopathy with no left ventricular wall motion abnormalities.

Serum enzymes. Serum creatine kinase was 79 IU/liter on admission (normal, 15 to 57) with 8% MB fraction, lactate dehydrogenase peaked at 72 IU/liter (normal, 48 to 115) with isoenzyme I greater than II (33% and 31%, respectively) and aspartate aminotransferase was 42 IU/liter (normal, 12 to 31).

Hospital course. Because of further episodes of chest discomfort and further evolution of ECG changes (Fig. 3), cardiac catheterization was performed. Left ventriculography confirmed the asymmetric septal hypertrophy and dyskinetic anterolateral, apical, apical septal and diaphragmatic segments. The left ventricular end-diastolic pressure was 26 mm Hg. A mean intraventricular gradient of 22 mm Hg at rest was found. This increased to 45 mm Hg after ventricular premature beats and after amyl nitrite. The epicardial coronary arteries were entirely normal. Marked systolic compression of septal perforator vessels was seen.

Oral administration of verapamil was begun, and the dose was increased to tolerance (360 mg/day). Radionuclide angiography showed a large apical aneurysm and asymmetric septal hypertrophy. On the 8th hospital day, an echocardiogram showed a persistent aneurysm but improvement in global left ventricular function. Single photon emission computed tomographic thallium-201 scintigraphy performed 13 days after admission showed only slight inferoapical thinning, which was within normal limits (Fig. 4).

Posthospital course. Radionuclide angiography performed 6 weeks after hospital discharge showed an ejection fraction of 33% and no evidence of aneurysm. Serial echocardiographic examinations over 4 months demonstrated gradual resolution of all regional wall motion abnormalities (Fig. 5). The patient has remained free of symptoms for more than a year since hospital dismissal.

Discussion

Myocardial ischemia and infarction in hypertrophic cardiomyopathy. This case demonstrates that spontaneous myocardial ischemia can result in marked wall motion abnormalities mimicking extensive myocardial infarction in hypertrophic cardiomyopathy despite normal epicardial coronary arteries. The subsequent resolution of the wall motion abnormality with only a slight cardiac enzyme elevation demonstrated that the ischemia was reversible. This pa-
Possible causes of acute ischemia. The cause of acute myocardial ischemia in patients with hypertrophic cardiomyopathy and normal coronary arteries is unknown. Cases demonstrating coronary emboli (8) and coronary spasm (10) have been reported. Myocardial compression of septal perforator branches, as seen in this patient, is not uncommon in hypertrophic cardiomyopathy (11). It could result in significant ischemia in these patients, who characteristically have a prolonged systolic ejection time, so that greater dependence of myocardial perfusion is placed on the relatively shortened diastole (6, 11).

Abnormalities in diastolic relaxation could be responsible for impairment of early filling of the coronary reservoir (12–14). Inadequate capillary density in relation to the increased myocardial mass present (14) may result in further decreased oxygen extraction (5). These microcirculatory changes may be due to replacement by abnormal myocellular architecture or fibrosis (15). Increased left ventricular filling pressure during ischemia could result in further decrease in oxygen delivery secondary to microcirculatory compression (5).

In the series of Maron et al. (16), 83% of the patients with hypertrophic cardiomyopathy had disease of the small intramural coronary arteries that was strongly associated with areas of fibrosis. Such small vessel disease could cause ischemia by reducing coronary reserve (17, 18). Tillmanns et al. (19) found evidence that ergometrine constricted the microvascular prearteriolar vessels. Cannon et al. (20) showed that patients with normal coronary arteries in whom ergonovine induced chest pain experienced chest pain after infusion of dipyridamole despite a more than doubling of the transmural flow, presumably because of abnormal small coronary vessels. It could be postulated that the methylergonovine received preceding the postpartum episode of our patient resulted in coronary spasm of either the small or the large coronary vessels.

Subendocardial and possibly transmural ischemia may occur with left ventricular hypertrophy of other causes, although the mechanisms may not be identical. It has been shown that in other patients with left ventricular hypertrophy, the small vessel coronary abnormalities are not as prevalent or as marked (16) and septal perforator compression is not present (11). Additionally, the presence of an outflow tract obstruction seems to play a role in coronary reserve (6).

Therapeutic issues. Therapy for ischemia in hypertrophic cardiomyopathy is no better defined than is the etiology. The most commonly advocated therapeutic goals have been to improve ventricular filling and to avoid inotropic agents. As in other ischemic syndromes, agents affecting the autonomic nervous system have been advocated by some because of its influence on inotropy, chronotropy and circulatory tone. Studies (3, 13) in hypertrophic cardiomyopathy have shown propranolol to primarily affect heart rate and systolic ejec-

Figure 5. Echocardiogram obtained 4.5 months after hospital discharge (end-systolic frame). The aneurysm present on the initial examination (Fig. 2) has resolved. Abbreviations as in Figure 2.
tion rate. Verapamil has been advocated because of its ability to improve diastolic filling (14). One study (21) showed it to increase cardiac output in some patients with outflow tract obstruction. Recent studies have shown ejection fraction to remain the same (13) or decrease (14) when verapamil is used. Verapamil may also be beneficial if vasospsam is a factor.

Conclusions. Hypertrophic cardiomyopathy can be associated with myocardial ischemia and infarction despite normal epicardial coronary arteries. The clinical occurrence is rare, and the cause is uncertain. This is, to our knowledge, the first report of documented reversal of a large amount of ischemic myocardium in the setting of an apparent acute myocardial infarction in a patient with hypertrophic cardiomyophy and normal coronary arteries. Treatment of such patients with verapamil may alleviate ischemia and may improve regional wall motion to a greater degree than that seen in patients with typical coronary artery disease. Thallium scanning may have an important prognostic role in these patients.

References


