Hypertrophic Cardiomyopathy*
Disappearance of Auscultatory, Carotid Pulse, and Echocardiographic Manifestations of Obstruction following Myocardial Infarction

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Three patients with echocardiographically documented asymmetric septal hypertrophy and findings compatible with dynamic left ventricular outflow tract obstruction suffered acute myocardial infarction in regions remote from the interventricular septum. Following infarction, echocardiographic signs of dynamic left ventricular outflow tract obstruction were no longer apparent. No resting gradient was documented in either of the two patients undergoing cardiac catheterization following myocardial infarction. Valsalva's maneuver resulted, however, in a gradient of 20 mm Hg in one patient. It appears that myocardial infarction may cause loss of or marked lessening of dynamic left ventricular outflow tract obstruction, even when the infarction involves areas other than the septum.

While acute myocardial infarction has been reported infrequently in patients with hypertrophic cardiomyopathy, myocardial necrosis may occur in the presence or absence of significant extramural coronary artery disease. In several cases, it appears that infarction involving the interventricular septum may have caused conversion from an obstructive form of hypertrophic cardiomyopathy to a nonobstructive, including even dilated, cardiomyopathy. We describe three patients with echocardiographic findings of asymmetric septal hypertrophy (ASH), systolic anterior movement of the mitral valve, and, in two cases, partial mid-systolic aortic valve closure, whose physical findings and echocardiographic signs of dynamic obstruction disappeared following acute myocardial infarction involving areas other than the septum.

CASE REPORTS

CASE 1

A 73-year-old woman with treated hypertension was evaluated for etiology of an ejection murmur audible between the apex and left sternal border. The carotid upstroke was rapid at rest and, during the Valsalva maneuver, had a spike and dome configuration. The intensity of the murmur increased dramatically during a Valsalva maneuver. M-mode and two-dimensional echocardiograms revealed no apparent intrinsic aortic or mitral valve disease. Systolic anterior mitral valve movement to abut the septum was visualized at rest (Fig 1), and prolonged systolic contact between septum and mitral valve developed during the Valsalva maneuver. In the body of the left ventricle, septal and posterior wall thicknesses were 20 mm and 15 mm, respectively. Left ventricular diastolic diameter was decreased at 3.2 cm, posterior wall excursion was increased at 14 mm, and estimated ejection fraction was 82 percent. Gated bloodpool scan demonstrated a hyperdynamic left ventricle, with an ejection fraction of 78 percent.

Five weeks later, the patient was admitted with symptoms of acute myocardial infarction, confirmed by the development of inferior Q waves and elevated serum enzymes. No murmur was apparent on admission or at any time during her subsequent course, even with the patient sitting and performing a Valsalva maneuver. Gated blood scan revealed a decrease in ejection fraction to 53 percent. Repeated echocardiography revealed a larger left ventricle, measuring 4.2 cm in diastole, a marked decrease in magnitude of posterior wall thickening and excursion, and no systolic anterior mitral valve movement. The carotid upstroke was now rounded, and no murmur could be recorded.

Cardiac catheterization, performed due to continued chest pain, revealed an ejection fraction of 49 percent, postero-basal and inferior wall hypokinesis, and no intraventricular pressure gradient, even during a Valsalva maneuver. Significant narrowing of all three coronary vessels was present. The patient underwent successful bypass grafting. No phonocardiographic or echocardiographic features of dynamic obstruction were noted in three postoperative echocardiograms performed over a two-year interval.

CASE 2

A 55-year-old man, with a six-year history of treated hypertension, underwent evaluation of a systolic murmur noted during hospitalization for acute inferolateral transmural myocardial infarction. A carotid pulse tracing was remarkable for a spike and dome configuration and for prolongation of the left ventricular ejection time, corrected for heart rate according to the method of Weissler et al., to 0.44 second. The apical ejection-quality murmur increased during the Valsalva maneuver. M-mode echocardiogram revealed prominent systolic anterior mitral valve movement to abut the septum and mid-systolic closure of otherwise.
normal-appearing aortic valve leaflets (Fig 2).

Following discharge, an ejection murmur was repeatedly noted. The patient was readmitted to the hospital 19 months later with recurrent infarction. Compared with his old ECG, there were changes compatible with a new transmural anterior wall infarction and with further inferior wall injury. The initial r wave was preserved in lead V₁. Only a faint systolic murmur, not changing with the Valsalva maneuver, was audible at the apex. Repeated echocardiography revealed no apparent midystolic aortic valve closure, and no anterior systolic mitral valve movement was evident in the region of the left ventricular outflow tract. Both M-mode and two-dimensional echocardiography demonstrated ASH, with the septum measuring 20 mm and the posterior wall measuring 11-13 mm. Carotid pulse tracing at rest showed no spike and dome configuration and a normal ejection time of 0.39-0.40 second. During the Valsalva maneuver, there was no change in murmur intensity, but the carotid pulse developed a spike and dome configuration.

Recurrent pain prompted cardiac catheterization, which revealed akinesis of a localized segment of the inferior wall and significant three-vessel coronary disease. No intraventricular pressure gradient was present at rest, but one of 20 mm Hg developed during the Valsalva maneuver, and there was a positive Brockenbrough sign. The patient underwent bypass grafting. His murmur remained faint, not increasing with the Valsalva maneuver.

Figure 2. Echocardiograms (patient 2) before and after his second myocardial infarction (MI). Before myocardial infarction, early to midystolic preclusion of the aortic valve leaflets (AoV) is apparent. Following myocardial infarction, presystolic closure no longer is apparent, and no systolic anterior movement of the mitral valve (MV) is recorded. Septum is thickened. EKG = electrocardiogram; LA = left atrium.
Figure 3. M-mode echocardiogram (patient 3) before myocardial infarction (MI). Systolic anterior movement (SAM) of the mitral valve (MV) abuts septum (SEP). EF slope of mitral valve is markedly diminished. M-mode echocardiogram after MI reveals no SAM and no mid-systolic closure of aortic leaflets (AoV). Thickening of mitral annulus is seen below the posterior leaflet of mitral valve. Both M-mode and long-axis cross-sectional echocardiograms after MI demonstrate asymmetric septal hypertrophy and reduction of left ventricular outflow tract diameter. EKG = electrocardiogram; AV = aortic valve; LA = left atrium; PW = posterior wall.

Case 3

A 77-year-old woman without hypertension underwent evaluation of a harsh, grade 4/6 apical murmur, variably described as ejection or holosystolic. M-mode echocardiogram revealed slight aortic valve thickening but preserved systolic excursion. Systolic anterior movement of the mitral valve (Fig 3), seen to abut the septum in some areas, and mid-systolic notchings of the aortic valve suggested dynamic obstruction. Septal thickness could not be optimally defined, but left ventricular outflow tract diameter was decreased at 1.6 cm.

Four years later, the patient was admitted with an anterolateral subendocardial myocardial infarction. The previously noted long, prominent apical murmur was no longer detected. A grade 2-3/6 brief ejection murmur was heard in the left third intercostal space, with only faint radiation to the apex. M-mode and twodimensional echocardiograms showed mild aortic valve thickening with systolic excursion of 1.4 cm. The upper septum was remarkably hypertrophied, measuring up to 21 mm in diameter, and narrowed the left ventricular outflow tract to approximately 12 mm in the sub-aortic region. Posterior wall measured 12 mm. Neither mid-systolic aortic valve closure nor systolic anterior movement of the mitral valve were present. Carotid pulse tracing appeared normal, with a normal ejection time of 0.42 second. An ejection murmur was recorded best at the aortic area, with faint radiation to the apex. It decreased with sitting and did not increase with the Valsalva maneuver.

Discussion

Although progression from obstructive hypertrophic cardiomyopathy to dilated, congestive cardiomyopathy has been considered rare, it is not known how often the obstructive form of hypertrophic cardiomyopathy progresses to a nonobstructive form. A loss of or marked reduction in the magnitude of dynamic obstruction is noted, however, in most patients following myotomy and myectomy. A change in the anatomic narrowing of the outflow tract is likely responsible, since left ventricular performance, assessed by rest and exercise ejection fractions, is generally maintained. Uncommonly, postoperative conversion to a dilated, congestive cardiomyopathy occurs.

It appears that myocardial infarction may also be a cause of progression from obstructive to nonobstructive hypertrophic cardiomyopathy. In the few reported cases, all with documented infarction of the interventricular septum, physical findings or echocardiographic features suggestive of dynamic obstruction were present before infarction but disappeared following infarction. Only one patient underwent catheterization before and after infarction. Intraventricular pressure gradients, present at rest and during provocation before infarction, were absent following septal damage. In our three patients, disappearance of auscultatory or echocardiographic signs or both of dynamic obstruction was noted following myocardial infarction occurring in locations...
other than the interventricular septum. It is unlikely that lability of obstruction alone was responsible for the changes in our patients, since repeated echocardiographic studies and physical examinations after infarction in each failed to demonstrate the previous findings. Unfortunately, none of the three patients underwent catheterization before infarction. While a definitive diagnosis of obstruction based on pressure gradients cannot, therefore, be made, the physical findings and echocardiographic features in our patients were typical of those seen with dynamic obstruction. The duration of the auscultatory abnormalities in our patients is not known. It is possible that ventricular hypertrophy, including ASH, did not develop on a genetic basis in the first two patients, but occurred sporadically, in response to systemic arterial hypertension. The latter has been postulated to be etiologically related to the development of hypertrophic cardiomyopathy in some patients manifesting typical clinical, echocardiographic, and hemodynamic findings of dynamic obstruction.15,16

Dynamic obstruction is likely produced by abnormal anterior mitral leaflet movement into the left ventricular outflow tract during systole. The anatomic narrowing of the outflow tract, due to septal hypertrophy or to an unusual anterior position of the mitral valve, in conjunction with a hyperdynamic left ventricle, is thought to produce a Bernoulli effect, drawing the anterior mitral leaflet into an obstructing position within the outflow tract. It is possible that myocardial damage may relieve dynamic obstruction by one of several mechanisms. Septal damage, resulting in thinning of the interventricular septum, may widen the outflow tract. This mechanism may explain the disappearance of signs of obstruction in those previously reported patients with septal infarction. In one of the reported patients, thinning of the septum was associated with increases in left ventricular systolic and diastolic dimensions and an increase in outflow tract dimension. Our three patients, with infarction of the inferior or anterolateral walls, suggest that other mechanisms might also be operative. Myocardial infarction may cause sufficient left ventricular dilatation to widen a previously narrowed outflow tract. Alternatively, reduction of myocardial contractility may diminish ejection velocity through the outflow tract, and thereby eliminate the postulated Bernoulli effect. Any one or a combination of the three postulated mechanisms may be responsible for a decrease in dynamic obstruction following myocardial infarction.

References

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