ECG of the Month

Cardiac Failure and Stroke in a 43-Year-Old Woman

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A 43-year-old woman with a long history of heavy cigarette smoking was in good health until she developed fatigue, dyspnea on exertion, and paroxysmal nocturnal dyspnea approximately three months before admission to our hospital. Four weeks before admission, she was admitted to another hospital for the sudden onset of a right hemiparesis. She was noted to be in atrial fibrillation, and cardiac catheterization and angiocardiography revealed triple-vessel coronary arterial disease and moderately severe mitral regurgitation. Because of repeated episodes of paroxysmal nocturnal dyspnea, she was referred to our hospital for cardiac surgery. On admission, an electrocardiogram was recorded (Figure).

Figure: Electrocardiogram recorded on admission.

What is your diagnosis?

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DIAGNOSIS: Coarse atrial fibrillation indicating left atrial enlargement and left ventricular hypertrophy with repolarization abnormality.

On physical exam, neck veins were normal. Crackles at both lung bases posteriorly partially cleared with cough. The carotid pulses were brisk. With the patient on her left side, the left ventricular impulse was large and forceful. A grade 3/6 systolic ejection murmur was loudest at the lower left sternal border, radiated to the cardiac apex and left axilla, and was accentuated by the Valsava maneuver.

The posteroanterior chest radiograph showed pulmonary plethora with prominent upper lobe vessels. The left ventricle was large, and the right atrium and right and left pulmonary arteries were prominent. The left atrium was huge with elevation of the left main bronchus, a double density over the right atrium, and a large appendage. On the lateral film, the retrosternal space was opacified by the cardiac silhouette.

The physical exam, electrocardiogram, and chest radiograph were compatible with rheumatic mitral regurgitation or hypertrophic obstructive cardiomyopathy, but the quality of the murmur, ejection rather than pansystolic or late systolic, and its accentuation by the Valsava maneuver favored a diagnosis of hypertrophic obstructive cardiomyopathy. The echo-Doppler study definitively resolved the issue by showing marked left ventricular hypertrophy with disproportionate thickening of the ventricular septum, cavity obliteration during systole, and marked systolic anterior motion of the anterior mitral leaflet against the hypertrophied septum that not only caused a left ventricular outflow tract velocity >6 m/s, but also permitted severe mitral regurgitation into a large left atrium. All cardiac valves appeared to be structurally normal.

Although reports of dying suddenly and having autopsies findings consistent with hypertrophic cardiomyopathy date back at least to 1868,1,2 the reports from London of Brock3 and Teare4 in 1957 and 1958, respectively, ignited the interest in hypertrophic cardiomyopathy that continues to burn brightly today. As would be expected, a new disease entity is often defined by its most overt manifestations. For hypertrophic cardiomyopathy, these were left ventricular hypertrophy, often striking and asymmetrical and without ostensible cause, as observed at necropsy by Teare,4 and obstruction to left ventricular outflow without definable fixed anatomic obstruction at operation, as described initially by Brock5 and, soon thereafter, by Morrow and Braunwald.6

Over the next decade, Braunwald and his co-workers determined that the dynamic subaortic obstruction increased with any situation, maneuver, or drug that decreased left ventricular volume and decreased with any situation, maneuver, or drug that increased ventricular volume.1 Morrow and colleagues successfully relieved the obstruction first by ventricular septal myotomy7 and later, by septal myectomy.8

Angiocardigraphers9,10 and then echocardiographers11 described the location of obstruction in hypertrophic cardiomyopathy as the site of apposition of the edge of the anterior mitral leaflet to the hypertrophied ventricular septum during systole, the result of systolic anterior motion of the anterior leaflet. Soon thereafter, simultaneously recorded echocardiograms and intracardiac pressure tracings showed that an echocardiographic obstructive index (duration of narrowing ÷ average septal-mitral distance) closely matched the pressure gradients on the same beats with a correlation coefficient of 0.95.12

Initially, atrial fibrillation was thought to be a rare occurrence in patients with hypertrophic obstructive cardiomyopathy.13-15 As more patients were followed for longer periods of time, however, the development of atrial fibrillation was noted commonly. This was not surprising considering the frequently increased left atrial pressure and volume resulting from diminished left ventricular compliance, even in the absence of mitral regurgitation. Of the 167 patients studied up to 1970 at the National Institutes of Health in Bethesda, Maryland, 16 developed atrial fibrillation.16 Each of them experienced a decrease in exercise capacity with the onset of atrial fibrillation, and 13 of them had more dramatic manifestations of cardiac dysfunction: cardiac failure in 11, syncope or presyncope in seven, angina pectoris in six, systemic arterial hypotension in five, and a generalized seizure in one.

Just as the mechanisms and characteristics of left ventricular outflow tract obstruction dominated early investigations of hypertrophic cardiomyopathy, more recently, the genetics of the disorder and sudden cardiac death, usually due to ventricular fibrillation and largely prevented by a prophylactic implantable cardioverter-defibrillator, have occupied researchers’ center stage. Clinically determinable risk factors for sudden cardiac death in patients with hypertrophic cardiomyopathy are a family history of sudden death related to the disease, nonsustained ventricular tachycardia on an ambulatory electrocardiogram, a history of unexplained syncope, a fall in blood pressure with exercise, and massive hypertrophy of the left ventricle (wall thickness ≥30 mm).17 Some patients with sudden cardiac death, however, have had none of these risk factors.17

Soon after hypertrophic cardiomyopathy was recognized as a distinct clinical entity, an autosomal dominant mode of inheritance was identified. To date, more than 1,400 mutations have been recognized in 11 genes encoding for either sarcomere or sarcomere-related proteins.17 Initial hopes that genotyping would accurately identify those patients at greatest risk for sudden cardiac death have for the most part faded. The presence of two or three sarcomere mutations in a single patient, however, seem to foretell early onset of the phenotype, i.e., hypertrophic cardiomyopathy, marked left ventricular hypertrophy, and the eventual need for cardiac transplantation because of severe left ventricular systolic dysfunction.17 Furthermore, a recent study has found double or compound sarcomere mutations in three patients with hypertrophic cardiomyopathy, sudden cardiac death, and...
none of the conventional clinical risk factors for sudden cardiac death.\textsuperscript{17} Further study obviously is needed.

Another recent approach to prognosis in patients with minimal or no cardiovascular symptoms has been metabolic exercise testing.\textsuperscript{18,19} Such patients with a peak myocardial oxygen consumption <60\% of predicted had a four-year survival rate free of death or severe symptoms of only 59\%.\textsuperscript{18} The severity of the resting left ventricular outflow tract gradient was the other predictor of a poor prognosis in these patients.\textsuperscript{18}

Our patient underwent ventricular septal myectomy (the Morrow procedure), a biatrial radiofrequency maze procedure, and a 3-vessel coronary arterial bypass procedure using the left internal mammary artery to the left anterior descending coronary artery and saphenous veins to the posterior descending branch of the right coronary artery and the first obtuse marginal branch of the left circumflex coronary artery. Four and one-half months postoperatively, she was in sinus rhythm with the left bundle branch block pattern expected after the Morrow procedure. She had no left ventricular outflow tract pressure gradient and no mitral regurgitation. She continued to have problems with fluid accumulation, however, which was probably the result of the restrictive component of her cardiomyopathy.

REFERENCES