ILLUSTRATIVE ECHOCARDIOGRAM

Echocardiographic Pseudo Idiopathic Hypertrophic Subaortic Stenosis*

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A patient with primary pulmonary hypertension is presented whose echocardiogram showed the features of idiopathic hypertrophic subaortic stenosis (IHSS), including asymmetric septal hypertrophy and a systolic anterior motion of the anterior mitral leaflet. Although IHSS and pulmonary hypertension usually are not confused clinically, the echocardiographic pattern may be similar in the two conditions. In this situation, asymmetric septal hypertrophy is not pathognomonic of IHSS.

Echocardiography has been demonstrated to be a very sensitive and specific noninvasive diagnostic method for the evaluation of patients with idiopathic hypertrophic subaortic stenosis (IHSS). It has been suggested that asymmetric hypertrophy of the left ventricle, with the interventricular septum at least 1.3 times as thick as the left ventricular free wall as identified by echocardiography, is the pathognomonic anatomic abnormality of IHSS. This finding, in conjunction with the characteristic systolic anterior motion of the anterior leaflet of the mitral valve which can also be identified by echocardiography, has been thought to be specific for the diagnosis of IHSS. The purpose of this report is to present a case of primary pulmonary hypertension simulating IHSS by the echocardiographic criteria mentioned above.

CASE REPORT

A 41-year-old white man was referred to Stanford University Hospital for evaluation of dyspnea. Throughout his life he had been exceedingly active and had participated in competitive sports. In the month prior to admission he had experienced progressive chest pain and dyspnea associated with exertion. Pertinent physical findings included normal lung fields, a normal cardiac apex impulse, a prominent left parasternal lift, normal first heart sound, increase in the intensity of the pulmonic component of the second heart sound, and a parasternal fourth heart sound. The remainder of the physical examination was within normal limits. The electrocardiogram revealed right axis deviation and right atrial and right ventricular hypertrophy. Enlargement of the right side of the heart and proximal pulmonary arteries with a decrease in the peripheral pulmonary vasculature was seen on chest x-ray film. There was evidence of mild obstructive lung disease by pulmonary function tests. Cardiac catheterization revealed pulmonary hypertension with a systolic pulmonary artery pressure of 100 mm Hg. There was no evidence for an intracardiac shunt. There was no pressure gradient within the left ventricle either at rest or during provocative maneuvers and no evidence of mitral stenosis. Large central pulmonary arteries with rapid tapering of their branches was demonstrated by angiography. No evidence for pulmonary emboli was seen. Forward angiomography showed no evidence for IHSS.

An echocardiogram obtained during hospitalization (Fig 1) showed asymmetric septal hypertrophy with gross thickening of the interventricular septum. In systole a distinct anterior motion of the anterior mitral leaflet began soon after the onset of systole and returned to meet the posterior leaflet before the end of systole. The right ventricular dimension (not shown well in Fig 1) was enlarged.

DISCUSSION

The echocardiogram in this patient revealed the typical findings of IHSS, i.e., asymmetric hypertrophy of the left ventricle with the septal thickness greater than 1.3 times the thickness of the left ventricular free wall, and a systolic anterior motion of the anterior leaflet of the mitral valve (Fig 2). Asymmetric hypertrophy of the septum is characteristically seen in patients with primary pulmonary hypertension, and can be found in most patients who have right ventricular systolic overload. This is probably related to hypertrophy of the interventricular septum in response to the right ventricular pressure overload.

It is not clear why the anterior leaflet of the mitral valve demonstrated the systolic anterior motion so characteristic of IHSS in this patient with primary pulmonary hypertension. In IHSS it is thought that abnormal traction of the papillary muscles secondary to their distortion by the hypertrophied septum may produce the abnormal motion of the anterior leaflet—the leaflet pulled into the outflow tract during systole. It is conceivable that sufficient septal hypertrophy from any cause may also produce ab-
normal geometry of contraction relative to the papillary muscles. This, then, could produce abnormal traction on the mitral leaflets causing the systolic anterior motion. No gradient need be present when the systolic anterior motion of the leaflet is present.6

While it has been thought that the echocardiographic appearance of asymmetric septal hypertrophy with systolic anterior motion of the anterior mitral valve leaflet is specific for IHSS, this case presents an interesting exception to this rule. The clinical situation rarely requires differentiation of IHSS from pulmonary hypertensive states.

ACKNOWLEDGMENTS: We wish to acknowledge the assistance and cooperation of Kitty Filly and Elizabeth London in performing these studies, and the secretarial and editorial assistance of Glenda Rhodes and Dorothy McCain in the preparation of this manuscript.

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