Echocardiographic Findings in Endomyocardial Fibrosis*

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Surgical treatment has recently been shown to improve the outcome of endomyocardial fibrosis; therefore, any finding helping to distinguish this condition from other cardiac diseases will be of unquestionable interest. Few reports of echocardiographic findings in endomyocardial fibrosis are available. We report a patient with autopsy-proved endomyocardial fibrosis whose echocardiogram showed striking features allowing suggestion of the correct diagnosis.

CASE REPORT

A 41-year-old white man was admitted to the Hospital for the first time in 1974 because of severe right-sided heart failure. From 1959 to 1967 he had lived in Equatorial Guinea, where he had suffered filariasis, malaria and dengue. Exertional dyspnea had begun in 1970; in 1971, malleolar edema appeared, developing in two years into a full-blown clinical picture of right-sided failure with jugular vein distension, hepatomegaly, ascites and edema. Proptosis was apparent. Cardiac examination did not disclose mitral or tricuspid regurgitation murmurs, and showed intermittent right ventricular gallop. ECG showed A-V junctional rhythm alternating with atrial tachyarrhythmia. X-ray examination of the chest disclosed huge cardiac enlargement with clear lungs. Eosinophilia was not detected.

From 1974 onward, the patient had at least four major episodes of heart failure, and in none did intensive medical treatment produce complete compensation. The patient repeatedly refused catheterization.

In 1980, an echocardiogram showed a massively thickened right ventricular free wall (27 mm) with no reduction of right ventricular diameter. Toward the apex the right ventricular lumen suddenly disappeared. The left ventricle was normal and pericardial effusion was present (Fig 1).

The patient died in July, 1981 with a low cardiac output syndrome. Autopsy showed enormous dilatation of the right atrium and biventricular endomyocardial fibrosis with a striking preponderance on the right ventricle, which was

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FIGURE 1. Echocardiographic sweep from the outflow of the right ventricle (left) toward the apex (right). The striking thickness of the anterior right ventricular wall (ARVW) (vertical arrows) and the sudden obliteration of the right ventricular lumen (horizontal arrows) can be seen. IVS: interventricular septum, PLVW: posterior left ventricular wall, PE: pericardial effusion.
In our patient, the dramatic involvement of the right ventricle by endomyocardial fibrosis correlated in the echocardiogram with an enormous thickening (27 mm) of right ventricular wall echoes and a sudden, complete, both systolic and diastolic obliteration of its cavity shown when the transducer was directed toward the apex. The fact that these striking right ventricular echocardiographic findings were not seen in the patients of Diénot et al.10 can be accounted for by their young age (2-19 years, mean 10.8 years). It seems reasonable to assume that in these patients the condition might have been in a less advanced degree of evolution than in our patient, whose illness had presumably developed during an unusually long period of at least 14 years.

Right ventricular anterior wall dimensions are difficult to establish in the echocardiogram.9 However, when the tracing is satisfactory, measured echocardiographic thickness has a good correlation with anatomic findings.8 Although right ventricular anterior wall thickness can be increased in many conditions (Table 1), it is not greater than 15 mm in right ventricular hypertrophy due to pulmonary hypertension or right ventricular outflow obstruction, or in amyloidosis with infiltrative heart disease. Echocardiographic differentiation of non-pedunculated right ventricular tumors could be more hazardous; however, the sudden obliteration of right ventricular wall thickness seems to us to be an important clue to the diagnosis of endomyocardial fibrosis. Bidimensional echocardiography might possibly be helpful as well in differentiating among these conditions.

In summary, we believe that a thorough echocardiographic examination of the right ventricular anterior wall and right ventricular cavity is warranted whenever a restrictive or obliterative syndrome is suspected, insofar as the echocardiogram may clearly point to the diagnosis of endomyocardial fibrosis.
REFERENCES


8 Child JS, Krivokapich J, Abbasi AS. Increased right ventricular wall thickness on echocardiography in amyloid infiltrative cardiomyopathy. Am J Cardiol 1979; 44:1391-95


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