The Contracted Form of Endocardial Fibroelastosis in an Adult: Hemodynamic and Angiographic Observations*


Hemodynamic and angiographic observations are described in an adult patient with the contracted form of endocardial fibroelastosis. Of note were the findings of elevated diastolic pressures in the left and right side of the heart, decreased left ventricular compliance, mitral regurgitation and distortion of the usual left and right ventricular angiographic anatomy, presenting a picture indistinguishable from that of a primary cardiac neoplasm. These findings are related to those described in children.

The term endocardial fibroelastosis (EFE) was introduced in 1943† to describe a disease characterized by diffuse fibroelastic endocardial thickening associated clinically with the occurrence of congestive heart failure and death early in life. The clinical entity has been further classified into two types: a “dilated” form, associated with marked left ventricular enlargement, low cardiac output and terminal heart failure; and a “contracted form associated with normal or reduced ventricular volumes, markedly altered ventricular compliance, and secondary pulmonary hypertension. The contracted type is unusual and accounts for probably no more than 5 percent of reported cases.

Although EFE is typically seen in infancy and early childhood, rare cases have been noted in adults. In the older age group the diagnosis is often not considered during the course of evaluation and is established rather at autopsy. This report describes clinical, hemodynamic, and angiographic observations in an adult with the contracted form of primary endocardial fibroelastosis who presented with an angiographic picture indistinguishable from that of a cardiac neoplasm. The patient appears to be the first adult in whom such data have been recorded.

CASE REPORT

The patient was a 26-year-old white woman who had been free of cardiac symptoms until four years prior to this evaluation when she noted the gradual onset of easy fatigability, decreased exercise tolerance, dyspnea on exertion, and poorly characterized, infrequent chest pain. However, two full-term pregnancies terminating two years and eight months previously were not associated with apparent cardiac complications. Over the eight months prior to admission she noted a significant decrease in exercise tolerance, marked dyspnea on exertion, and paroxysmal nocturnal dyspnea.

Physical examination revealed a blood pressure of 80/40 mm Hg and a pulse of 80 beats/minute. The carotid pulse was normal. The hepatojugular reflux was present. The lungs were clear. The cardiac apical impulse was not displaced and was of normal quality. There was a prominent parasternal lift. The first heart sound was normal and the pulmonic component of the second sound was accentuated. A third heart sound was audible at the apex. A blowing holosystolic murmur was heard at the apex, with radiation to the left axilla. No diastolic murmur was audible. There was no hepatomegaly, cyanosis, clubbing or edema.

Roentgenograms of the chest revealed enlargement of the right atrium, right ventricle and left atrial appendage, and prominence of the main pulmonary artery segment. Pulmonary venous congestion and Kurley B lines were evident (Fig 1). A standard 12-lead electrocardiogram showed normal sinus rhythm, with left axis deviation and generalized low voltage in the standard leads. There was delayed progression of the R waves, with prominent S waves in the left precordium. In several leads wide fluctuation of QRS amplitude was apparent. The initial component of the R wave in V1 was tall (4.5 mm) and peaked.

Cardiac catheterization demonstrated elevation of the left ventricular end diastolic pressure and pulmonary capillary wedge pressures, with concomitant abnormalities in right heart pressures (Table 1). Both left and right ventricular diastolic pressures were elevated, with a “dip and plateau” pressure contour suggestive of constrictive or restrictive disease. Prominent X and Y descents were also noted in the atrial pressure contours. The resting cardiac index was 2.5 liters/min/M².

On selective biplane cineangiography the left ventricle was noted to have a markedly abnormal spherical shape, with truncation of the apex (Fig 2). Significant mitral regurgitation was present. In the left anterior oblique view the regurgitant jet was asymmetric, with initial layering of contrast along the posterolateral wall of the left atrium. Left ventricular end diastolic volume was 76 ml/M², with an ejection fraction of 54 percent.

The left atrium was visualized during the levophase of a pulmonary artery angiogram and was enlarged and had no filling defects. The mitral valve was not thickened. Right ventricular angiography demonstrated a large persistent filling defect in the region of the right ventricular apex (Fig 3).
Coronary angiography yielded normal results. These data suggested the possibility of an infiltrating intracardiac tumor involving the intraventricular septum, with extension into both ventricles and involvement of the left ventricular anterolateral papillary muscle. Because of the findings cardiac surgery was undertaken.

At operation, the external appearance of the heart revealed distortion of the apex, with superficial contracture of this portion of the heart. The mitral valve leaflets appeared normal, but the anterior papillary muscle was totally encased in a white, thick, fibrous tissue which arose from the endocardial surface of the left ventricle. The entire apex of the left ventricular cavity was covered by a layer of light tan, smooth, glistening tissue, which multiple biopsy specimens showed to be layered thrombus overlaying a thick fibrous endocardium. The apical portion of the right ventricular cavity and right ventricular septum also presented a white glistening surface from which multiple biopsy specimens were obtained. Biopsy material from the septum, left ventricle and right ventricle all showed markedly thickened endocardium with proliferation of elastic tissue. Myocardium seen beneath the endocardial layer was normal as were the mitral valve leaflets. Histologic examination of the left anterior papillary muscle also demonstrated endocardial thickening and normal underlying myocardium.

Because of the significant mitral regurgitation, the mitral valve was replaced with a Starr-Edwards prosthesis. The

Table 1—Cardiac Catheterisation Data

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure, mm Hg</th>
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<tbody>
<tr>
<td>Right atrium s/v(m)</td>
<td>15/9(8)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>42/13</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>42/18(30)</td>
</tr>
<tr>
<td>Pulmonary capillary wedge s/v(m)</td>
<td>25/30(20)</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>87/20</td>
</tr>
<tr>
<td>Aorta</td>
<td>87/67(76)</td>
</tr>
</tbody>
</table>
Our patient with the contracted form of EFE presents findings which are similar to those few cases recorded in the pediatric age group.\textsuperscript{4,12} To our knowledge, she is the first adult with this form of the disease in whom a complete hemodynamic and angiographic evaluation has been made. The findings in this case call attention to the need for early detailed laboratory evaluation, and if necessary, surgical intervention, in the patients with a clinically obscure type of cardiac disease.

There are two prominent pathophysiologic conditions associated with the contracted form of EFE; both are illustrated in our patient. Altered left ventricular compliance leads to significant elevation of diastolic pressures in the presence of normal or reduced ventricular volume. This altered left ventricular pressure-volume relationship results in elevated pressures in the left atrium and pulmonary vascular bed, and the clinical pattern of pulmonary vascular congestion and pulmonary hypertension. Secondly, fibroelastic proliferation in the region of the papillary muscles leads to papillary muscle dysfunction with hemodynamically significant mitral regurgitation and its clinically recognizable findings. In addition, as noted in our patient, marked proliferation of the endocardial and subendocardial regions of both ventricles, associated with a considerable degree of mural thrombosis, can produce an angiographic picture indistinguishable from that of a primary cardiac neoplasm.

These findings stand in direct contrast to the more familiar observations made in children with the dilated form of EFE.\textsuperscript{5,4,13} In that instance, primary importance is a marked generalized diminution in contractility of the left ventricle. A resultant marked increase in ventricular volumes, diminished ejection fraction, left and right heart hypertension, and markedly reduced cardiac output are apparent at the time of hemodynamic and angiographic evaluation. The clinical picture is dominated by the low output state and terminal heart failure.

The etiology of EFE in both children and adults is unclear. Various hypotheses regarding the cause of this disease have been offered, including intra- or extracutaneous infection,\textsuperscript{14,16} hereditary factors,\textsuperscript{12} lymphatic obstruction\textsuperscript{17} and an atypical response to ventricular pressure overload. Likewise, the relationship between the adult and childhood forms is unknown. The condition in adults must be distinguished from East African endomyocardial fibrosis which is not associated with abundant elastic tissue proliferation and is characterized by myocardial replacement with fibrous tissue.\textsuperscript{19}

The prognosis of EFE is generally poor, and is characterized by a progressive downhill course. Treatment has been primarily medical in nature. The replacement of a grossly insufficient mitral valve in our patient has at least temporarily stabilized her condition and may improve her longterm survival.

REFERENCES


SOSTRE, HURLEY, ZARET
Nairobi National Park, Kenya

This park is one of the most remarkable in Africa and indeed in the world, in that it lies only five miles from the General Post Office of Kenya's most important city; when the wind is in the right direction, the lions for which the park is famous can be heard roaring at night even in Nairobi's plussiest hotels. Although near to the equator, it is never too hot or too cold owing to its elevation, which is between 5,000 and 6,000 feet above sea-level. On the lower plains of the park the dominant trees are various species of acacias. Numerous flowering shrubs and plants add greatly to the beauty of the park and at sunrise and sunset the plains take on a warm and golden glamor enhanced by distant prospects of great mountains, such as Mount Kenya, Mount Kilimanjaro and the Aberdare Range. Over sixty species of mammals and reptiles and over four hundred species of birds can be seen in the park. Wildebeest, hartebeest, and impala are the most common ungulates, and form the main prey of lions which can be easily seen in the park. These lions are so used to visitors that at dusk it is common to see up to a dozen of them lying in unconcerned ease in a semicircle of cars, all with their headlights blazing full upon them.