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SESSION II

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Endomyocardial disease—clinical features

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Summary

The clinical features of tropical and temperate zone endomyocardial fibrosis (EMF) are the same, allowing for certain regional, environmental and possibly genetic variations. For example, the seasonal incidence in rainy humid areas probably reflects the large and repeated parasitic infestations in tropical EMF, while the absence of tissue eosinophilia in organs other than the heart in tropical EMF may reflect racial and environmental differences between tropical and western geographical areas that have still to be elucidated. That EMF occurs in Europeans who have lived in the tropics is undoubted, but the absence of right ventricular involvement in Europeans in the tropics, but not in temperate climes, is unexplained; perhaps it is a chance finding. It is also apparent that the extreme degrees of right ventricular EMF that are commonly seen in the tropics, with almost complete obliteration of the ventricular cavity are not usually seen in eosinophilic EMF in temperate areas. Involvement of both ventricles and of both atrioventricular valves is, however, common both in the tropics and in temperate climate EMF.

KEY WORDS: endomyocardial disease, tropics, temperate zone.

The previous history and presenting symptoms

In the tropical variety of endomyocardial disease, endomyocardial fibrosis (EMF), there is often a history of repeated attacks of fever and these may be accompanied by palpitations and dyspnoea on effort. Palpitations may be the result of episodes of atrial fibrillation. In between attacks, the patient may be well, but a gradual deterioration in well-being may occur. The attacks of fever may be related to the rainy season, and it has been implied that outbreaks of EMF may occur at these times, suggesting an infective origin.

As the disease progresses, symptoms of congestive heart failure and left ventricular insufficiency appear, fatigue and dyspnoea become worse, peripheral oedema develops, and eventually there is severe limitation of function. When atrial fibrillation becomes established, the cardiac output falls further and symptoms increase. Naturally, when the right ventricle only is involved, the symptoms are principally those of fatigue, lassitude, abdominal discomfort and swelling due to hepatomegaly and ascites. When left ventricular disease is present, there is cough, with dyspnoea, paroxysmal and on effort, and haemoptysis due to the high left atrial pressure. Biventricular disease gives symptoms due to congestion in both systemic and pulmonary circulations.

In severe right ventricular disease, the very high central venous pressure may cause exophthalmos, or even oedema of the face, which becomes moon-shaped.

In the endomyocardial disease of temperate zones, usually associated with eosinophilia, the initial symptoms may be referable to the eosinophilic syndrome and include features outside the cardiovascular system; symptoms due to anaemia, splenomegaly, or even cerebral involvement. Cardiac symptoms are those of progressive heart failure involving usually both ventricles. However, the extremes of ascites, and of intense cardiac restriction are not seen so commonly as in tropical EMF.

Physical signs and haemodynamics

The physical signs of endomyocardial fibrosis depend upon the pathology and haemodynamics. When the right ventricle alone is involved, the signs are those of restriction of inflow to the ventricle and later obliteration of the cavity. The restriction of inflow acts in a way similar to pericardial constriction, ventricular filling is rapid at first, then slows

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down in the latter part of diastole as the endocardial fibrosis restricts further expansion of the ventricle and limits further increase in diastolic volume. This results in a characteristic pressure pulse in the right ventricle, with early diastolic dip, plateau, and high end-diastolic pressure. The early diastolic dip does not always reach, or go below, zero, while the systolic pressure is often elevated. The effect on right atrial and central venous pressures produces a characteristic 'M' shaped pulse with full 'a' and 'v' waves and deep 'x' and 'y' troughs: the central venous pressure is elevated, and may rise further on inspiration.

As the ventricle becomes progressively obliterated by the increasing fibrosis, the pressures rise higher, and eventually tend to be similar in right atrium, ventricle and pulmonary artery. Eventually, the right ventricle may become virtually reduced to a small outflow tract sinus and the pulmonary artery is filled from the right atrium, which becomes enormously enlarged. Since the tricuspid valve is involved in the pathological process, some degree of regurgitation is common. Eventually atrial fibrillation becomes established, and severe congestive heart failure occurs.

There is commonly a pericardial effusion. Even when atrial fibrillation is established, the giant right atrium can produce an adequate cardiac output, presumably because of the elevated preload due to the extremely high venous pressure. Ascites and hepatomegaly develop and hepatic fibrosis can occur eventually.

Investigations

The heart may be small or grossly enlarged. Enlargement is usually due to atrial dilatation, which is prominent when there is severe atrio-ventricular valvar insufficiency. The ventricles do not tend to be enlarged because the endomyocardial restriction tends to prevent dilatation. Hypertrophy is not a striking feature, and systolic function remains good until the later stages of the disease. When there is severe mitral regurgitation, the left atrial pressure is raised and the lung fields show the characteristic vascular pattern with increased flow to the apices and interstitial oedema.

A very characteristic sign is a linear strip of calcium in the region of the left ventricle as a result of calcification of thrombus in the left ventricle. When the right atrium is massive the heart has a globular shape and may fill the thorax; the presence of a pericardial effusion accentuates this appearance.

The electrocardiogram is not specific; flat T waves, sinus tachycardia, and low voltage QRS waves are all common features. Bundle branch block may occur, but atrio-ventricular block is uncommon. M-mode echocardiography is of limited value, but two-dimensional studies show filling defects in the apices of the ventricles (George *et al.*, 1982; Acquatella, 1983). The 4 chamber view is the best view for showing the lesions.

Angiocardiography is an important diagnostic method. The combination of effective systolic contraction of the ventricles with normal or only slightly reduced ejection fraction with obliteration of the apex of the ventricles is characteristic, but angiocardiography is of little value in the early stages when there is merely a thin layer of endocardial thickening. Gated blood pool angiograms (MUGA) give good estimates of ejection fraction, but are of doubtful value when atrial fibrillation is present.

As technology develops and experience accumulates it is likely that two-dimensional echocardiology will provide the best means for suspecting the diagnosis early in the disease, so that endomyocardial biopsy can confirm it. Biopsy of the right ventricle will usually yield material from which a diagnosis can be made from the characteristic features (Olsen, 1983). The diagnosis should not be regarded as disproved unless no evidence of EMF can be found on multiple specimens obtained from different parts of the septum. Left ventricular biopsy is not recommended because of the high incidence of thrombus in the cavity and the risk of systemic embolism.

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