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REVIEW ARTICLE

The Diagnosis of Endomyocardial Fibrosis*

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Summary. A review of the clinical findings of about 102 cases of endomyocardial fibrosis (EMF) is presented. It is concluded that right ventricular EMF can be recognized in most instances on clinical grounds. The recognition of left ventricular EMF would still require sophisticated investigation. Features which may be used for the rapid recognition of EMF in the field are presented.

Résumé. Les constatations cliniques faites chez 102 cas d'endomyocardite fibreuse sont présentés. Dans la pluplart des cas, il est possible de diagnostique l'endomyocardite fibreuse du ventricule droit sur la base de signes cliniques. Le diagnostic d'endomyocardite fibreuse du ventricule gauche exige des investigations plus poussies. Les caracteristiques qui permetent le diagnostic rapide de l'endomyocardite fibreuse sur le certain sont présentées.

The haemodynamic complexities of endomyocardial fibrosis have been the subject of numerous publications from East and West Africa: Shillingford & Somers (1961), Abrahams (1962), Parry & Abrahams (1963). The structural abnormalities which form the basis of the haemodynamic picture have also been well described: Davies & Ball (1952), Edington & Jackson (1963). World Health Organization study groups have defined EMF as a progressive disorder characterized in the established condition by fibrosis in the ventricular cavities and affecting in particular the apex and subvalvular regions; a definition based chiefly on our knowledge of the pathology of EMF (Hutt *et al.*, 1965; WHO Chronicle, Vol. 21, No. 10, pp. 407–412). Despite all these publications there has been no comprehensive study of the clinical manifestations of endomyocardial fibrosis.

A number of factors prompted this study. The first was an attempt to obtain for the first time a comprehensive and statistically meaningful picture of the clinical characterization of endomyocardial fibrosis. The second was in the expectation that such a study would yield criteria, with which a definitive diagnosis of endomyocardial fibrosis could be made in life without resort to sophisticated and often dangerous techniques of investigation. The third

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was the need to provide simple and sensitive criteria for field work in the epidemiological study of EMF.

PATIENTS AND METHODS

Much of the data presented here is incomplete because a good deal of the material collected has been lost. Part of the purpose of this paper however is to draw your attention to the unsolved problems of EMF. About 102 cases of endomyocardial fibrosis had been collected in the Cardiac Registry at the UCH Ibadan up to July 1966. Cases of EMF were selected for inclusion in the Registry as follows:

(i) Autopsy proven cases of EMF (thirty-four cases).

(ii) Patients with angiocardiographic evidence of obliteration of the apex of either ventricle.

(iii) Patients with haemodynamic evidence of ventricular constriction and atrio-ventricular incompetience.

The grouping of these cases into the three clinical syndromes of EMF has not been fully studied. In over one-half of the cases, the status of the left heart had not been determined. The right ventricle was involved alone or together with the left in the majority of cases in which both ventricles were studied (less than one-third of the series). In a group of twenty-four cases with autopsy confirmation, there were six cases of right venticular EMF, six of left ventricular disease and twelve of biventricular disease.

The following procedure was adopted in all cases seen in the clinics and admitted for haemodynamic studies: A social questionnaire designed by the Department of Preventive and Social Medicine was completed. A detailed history was obtained and clinical examination performed. The following investigations were undertaken: a plain chest radiograph, an electrocardiogram, a phono- and impulse-cardiogram where possible, a right heart catheterization and angiocardiogram and 2 weeks later, a left heart catheterization and angiocardiogram. A final diagnosis was made during a group discussion of the data between the unit and the Department of Radiology. These data were then transferred onto a WHO *pro forma*.

RESULT

(1) *Social questionnaire*

No detailed information on the result of this study is available, but it was obvious early on that most cases of EMF came from the poorer suburban or rural classes.

(2) *Age, place or origin*

Figure 1 shows the age distribution of fifty-one patients with EMF attending at out-patients in early 1966 and of thirty-four cases with autopsy confirmation. Ninety percent of the out-patients cases and 88·5% of the autopsy material were under the age of 35 years. The peak incidence was in the second and third decades.

The actual figures relating to the places of orgin are not available. This part of the study was undertaken for cases originating from Western Nigeria. A high concentration of cases came from Ujebu-Ode and the surrounding villages. Very few came from the Ibadan area. To ensure that this distribution was not fortuitous a similar study was undertaken for

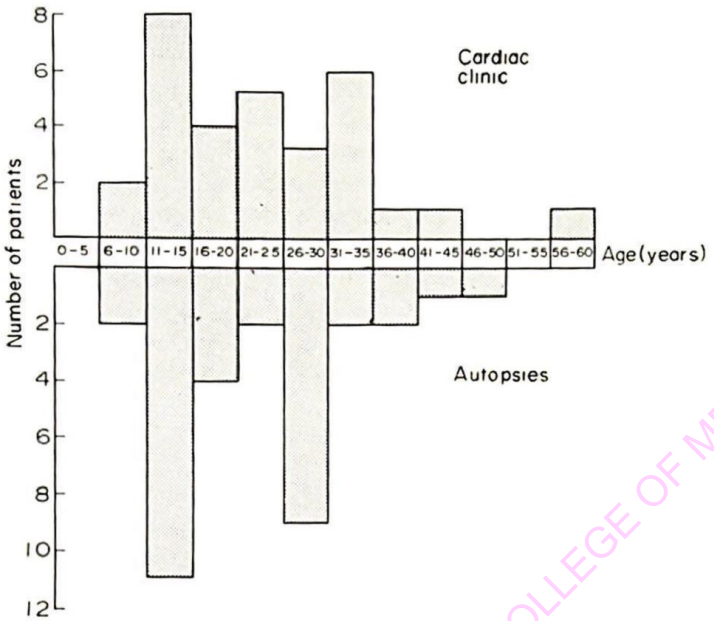

FIG. 1. Age incidence of fifty-one cases of EMF attending the cardiac clinic and of thirty-four autopsy proven cases (see text).

rheumatic heart disease, selecting only cases with mitral stenosis, and those with mitral incompetence and aortic incompetence. A low concentration of cases with rheumatic heart disease was observed in the Ijebu-Ode area while very many cases originated from the Ibadan area.

(3) *History*

Symptoms were remarkably unimpressive except in a few cases in which the left ventricle appeared to be severely involved. The following symptoms were noted with decreasing frequency: tiredness; swelling of the face, abdomen and legs; breathlessness; cough; chest pain; abdominal discomfort; haemoptysis; fever.

Tiredness and swelling were usually noted in cases in which the right heart was involved, but such cases sometimes also complained of breathlessness, chest pain and abdominal pain. In left heart disease, breathlesness and cough were the main features.

Most cases presenting with lone or dominant right ventricular disease had a history lasting over 1 year and often gave a history of recurrent swelling in the past. The few cases studied with lone left ventricular disease had a short history, usually of less than 6 months.

We observed no case we could refer to as early EMF. We were able to obtain from about twelve patients a previous history (often in childhood or adolescence), of fever, swelling of the face and body, progressive easy fatigability and breathlessness lasting weeks or months, often involving admission to hospital; and to all intents and purposes regressing completely until the onset of symptoms for which the patient was currently attending hospital. A few patients gave a history of recurrent episodes. The weaknesses of this form of investigation are obvious.

(4) *Clinical signs*

(a) *Isolated or dominant right ventricular EMF.* This group formed the majority of cases studied and included those cases in whom the status of the left ventricle was unknown. Most patients showed a marked puffiness around the eyes. A few had proptosis. Many patients exhibited retardation of growth. Cyanosis occurred in about 10% of the cases and was associated with clubbing of the fingers. Ascites and oedema were invariably present. Terminally two patients observed in this phase showed jaundice.

Most patients (90%) showed a dominant systolic pulsation of the jugular vein; a feature pathognomonic of tricuspid regurgitation.

A few patients in sinus rhythm showed a dominant atrial wave. This wave form was confirmed at right heart catheterization. The diagnosis of EMF in this group could only be made by angiocardiography.

A left parasternal pulsation maximal over the pulmonary area was frequently noted. This was shown to consist of a systolic retraction, when recorded on impulse cardiography. A precordial systolic recession may rarely be observed in constrictive pericarditis (Spodick, 1964), but it is usually not as localized as in EMF. This physical sign may be considered as important as the recognition of tricuspid regurgitation, in the diagnosis of right ventricular EMF.

The commonest finding on auscultation was a third heart sound. This occurred early, usually not later than 0·10 sec and was high pitched. Audible systolic murmurs were rare and were just as infrequently recorded.

(b) *Isolated left ventricular EMF or dominant left ventricular disease.* Cases of lone left ventricular disease were rare and we were able to study about four such cases.

The apical impulse was normal in position. In about two cases it showed a small displacement to the fourth intercostal space in the anterior axillary line. No significant pulsations were obtainable on impulse cardiography. However, in one case, a mistaken diagnosis of rheumatic heart disease was made on the basis of a displaced apex beat, a large and heaving left ventricule and mitral regurgitation; and was apparently confirmed at left ventricular angiocardiography.

About 1 year later a repeat left ventricular angiocardiogram showed early obliteration of the apex. It is obvious that at the moment when the ventricular lesion involves only the posterior mitral valve leaflet, the apical impulse would be indistinguishable from that of rheumatic mitral incompetence and a diagnosis of EMF would be impossible.

A parasternal heave was uncommon. The syndrome of mitral incompetence and pulmonary hypertension (Abrahams & Brigden, 1961) seems rare. However, not enough figures of the pulmonary artery pressure were available from cases of left ventricular EMF for any meaningful conclusion to be drawn.

The most important auscultatory finding was an apical pan-systolic murmur. The first sound was normal. An opening snap was frequently recorded. A third heart sound was the next commonly noted abnormality.

(c) *Biventricular EMF.* This is the largest group and shows no specific distinctive features from the two groups above. During this study it was the common experience to find that cases previously classified as lone right ventricular EMF on the basis of clinical and catheter data, had evidence of advanced left ventricular disease when such patients were subjected to left ventricular angiocardigraphy. The murmur of mitral regurgitation was absent although

regurgitation could be demonstrated at angiocardiography. The pulmonary artery pressure was not raised. The presence of associated right heart disease always tended to mask the features of left ventricular disease, no matter its severity.

(5) *Electrocardiography*

Most reports have suggested that no constant abnormalities are found in the electrocardiograms of EMF (Ball, Davies & Williams, 1954; Williams & Somers, 1960; Abrahams, 1962). We were able to analyse in a preliminary study the electrocardiograms of twenty-four autopsy proven cases of EMF (Ikeme & Uzodike, 1971). We found low voltage QRS complexes even in chest leads, usually 5 mm or less; a dominant R wave or 'qr' pattern in lead V_1; a mean frontal QRS vector directed to the right in association with a normal frontal T vector and up right T waves in leads V5 and V6. This last feature was distinctive enough to allow a differentiation from constrictive pericarditis. Electrocardiographic differentiation of the clinical syndromes of EMF is much less certain. One-third of eighteen patients in whom the right ventricle was involved had atrial fibrillation, and an additional patient had atrial flutter. Atrial fibrillation however occured in one out of six patients with a lesion confined to the left ventricle.

(6) *Radiology*

(a) *Lone right or dominant right ventricular EMF.* We owe a great deal to the Department of Radiology at the UCH Ibadan under Professor P. Cockshott for our understanding of the radiology of EMF (Cockshott, 1965; Cockshott, Saric & Ikeme, 1967). Most cases admitted to this study had a chest radiograph. The heart was invaribly enlarged and showed a stencilled outline due either to the often associated pericardial effusion or to a poorly moving heart. The lung fields were oligaemic. Less commonly (in about one-third to one-half of the cases), a bulge was noted on the left border of the cardiac silhouette due to the hypertrophied out-flow tract of the right ventricle. In a small proportion of cases the right atrium was aneurysmal and occupied the whole of the right border of the heart shadow.

(b) *Left ventricular EMF.* The heart was slightly enlarged in left ventricular EMF and showed a mitral configuration. In no case was the left atrium aneurysmal. There was no narrowing of the lower lobe arteries, a feature often found in rheumatic mitral valve disease associated with pulmonary hypertension. Septal lines were not seen.

In about half a dozen cases there was semi-lunar calcification at the apex probably in the left ventricle. Autopsy confirmation of the pathology has been made on occasion and some of us feel that this form of calcification is pathognomonic of EMF.

Although I have not formally discussed angiocardiographic appearances, I wish to mention two features which were found on left ventricular angiocardiography in the presence of right heart disease (Fig. 2). The first was a marked rotation of the ascending aorta to the left so that in the antero-posterior view, it comes to overlie its descending thoracic part. The second was a bowing upwards of the right coronary artery best seen in the lateral view, as it traverses the right atrioventricular groove. Both features are produced by right atriomegaly and indicate the presence of right heart disease. Recognition of these appearances could provide a time-saving technique for determining the status of both ventricles in EMF.

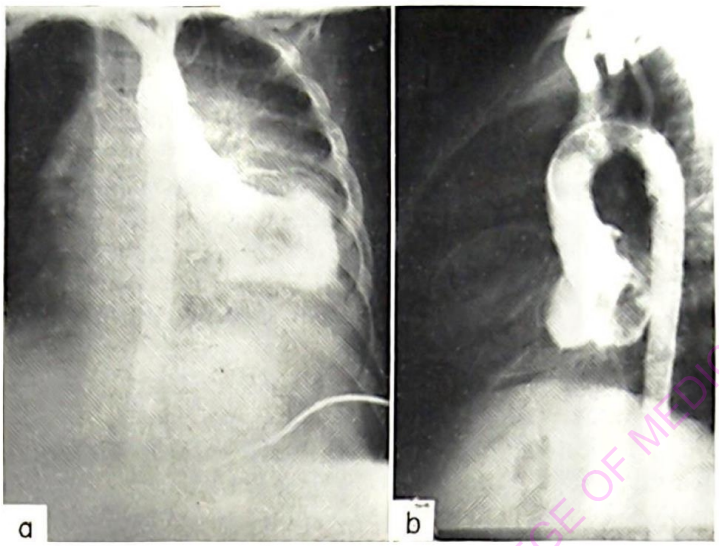

FIG. 2. Right ventricular EMF. Normal left ventricular angiocardiogram. (a) Antero-posterior vein; (b) left lateral. Note rotation of the ascending aorta in the antero-posterior view and in the lateral view, the arch formed by the right coronary artery.

CONCLUSION

Exhaustive studies of hospital cases of EMF still have a place in our search for the nature of EMF. This preliminary report should help to draw attention to the gaps in our knowledge of the clinical manifestations of EMF. Far too few cases of lone left ventricular EMF have been adequately studied and more work needs to be done on this score. We do not know yet with any certainty the relative frequency with which the ventricles are involved in endomyocardial fibrosis.

If the right ventricle is more commonly affected than the left in EMF, it is obvious that before a rheumatic aetiology can become fully acceptable, not only such questions as why EMF appears to be a disease of the mural and not valvular endocardium need to be answered, but also why it is more common in the right ventricle than in the left.

The needs of the epidemiologist are different from those of the physician. The physician will continue to require well documented cases; either because of the need for accuracy in each specific instance or because of the need to study the natural history of well proven cases of the disease. While accepting that a clinical diagnosis can be reached in many cases, he would still need to apply special methods of investigation frequently. The epidemiologist on the other hand does not require a full definition of the clinical syndrome. In the study of EMF, his principal weapons would be a number of indices which are easy to measure and which occur in sufficiently high frequency to enable a high proportion of cases of EMF to be recognized.

Field studies have become an essential tool in the search for the aetiology of EMF. Although it is not possible here to indicate the statistical significance of each index; we believe that EMF can be recognized in any population group under the age of 35 years by the following features:

(1) A plain chest radiograph showing: cardiomegaly; a stencilled outline and oligaemic lung fields; a large right atrium and a bulge over the out flow tract of the right ventricle.

(2) A plain chest radiograph showing apical semilunar calcification.

(3) A person with a raised jugular venous pressure showing a dominant systolic pulsation; and an early high pitched third sound on auscultation.

(4) Systolic retraction in the second intercostal space to the left of the sternum together with an early high pitched third sound.

(5) An electrocardiogram showing: low voltage QRS complexes even in chest leads; a frontal QRS vector directed to the right and a normal T wave vector.

Except for the second index, these are all features of dominant right ventricular EMF and until more data is available on dominant or lone left ventricular EMF, case recognition in the field would depend principally on recognizing signs of right ventricular abnormality.

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