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Cardiac Disorders Simulating Endomyocardial Fibrosis

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Summary

Jaiyesimi F. Cardiac Disorders Simulating Endomyocardial Fibrosis. *Nigerian Journal of Paediatrics* **1984; 11:121.** Four patients, aged between 8 and 11 years, whose cardiac disorders closely mimicked endomyocardial fibrosis (EMF) are reported. They all had heart failure, tricuspid regurgitation and globular hearts with no murmurs. In three of the cases, the lung fields were oligaemic. Angio and echocardiographic studies subsequently revealed that the first patient had congenital pulmonary stenosis while the second had a coarctation syndrome. Gross cardiac dilation complicated these two malformations. In the third and fourth patients, the correct diagnoses, dilated cardiomyopathy and constrictive pericarditis respectively, were ascertained only at necropsy. These two lesions should therefore, be considered in the differential diagnosis of EMF. Furthermore, the four cases illustrate the unreliability of basing the diagnosis of this cardiomyopathy on clinical features alone.

Introduction

The earlier belief that endomyocardial fibrosis (EMF) was confined to tropical Africa,[1 2] has now been discarded following reports of the existence of the disease in other continents.[3 8] This recent awareness and the increasing evidence that eosinophilia is probably one of its major aetio-pathogenetic factors,[4 9 10] have evoked an upsurge of interest in EMF. It is therefore, anticipated that more reports will emanate from other places where the disease was previously unrecognized. For the evolving epidemiological data to be reliable, they must be based on accurate diagnosis. That demands, among other things, an awareness of other cardiac disorders that may simulate EMF. This communication concerns four cases which, in several ways, mimicked right ventricular EMF.

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Patients and Methods

The four cases from among patients who attended the paediatric cardiology unit, University College Hospital (UCH), Ibadan. In addition to physical examination and routine laboratory investigations, each patient had chest radiographs, standard 12-lead electrocardiogram (ECG), cardiac catheterization and angiocardiography.

Oxymetry was not performed because the requisite facilities were not available. Although two of the patients (Cases 3 and 4) were included in a previous publication on acquired tricuspid incompetence in childhood,[11] the salient features in all four patients are summarised below.

Case Report

Case 1

This was an 8-year old male patient who first attended the clinic in 1976 complaining of cough, exertional dyspnoea and generalized oedema for two months. Except for a febrile convulsion at the age of 2 years, he had enjoyed good health and developed satisfactorily. The initial examination revealed an acyanotic boy with slightly weak peripheral pulses, a blood pressure (BP) of 90/60 mmHg and a right ventricular heave. There was a loud pulmonary ejection systolic murmur and the second heart sound (S2) was single. No click or diastolic murmur was audible. There were signs of right-sided heart failure. Moderate cardiomegaly with a cardio-thoracic (CT) ratio of 62% and normal pulmonary vascular markings were evident on a chest radiograph, while the ECG showed biatrial hypertrophy, right bundle branch block and right ventricular hypertrophy (RVH).

Heart failure complicating pulmonary valve stenosis was diagnosed. He was therefore, treated with digoxin and diuretics. Cardiac catheterization was deferred since cardiac surgery was then, not feasible at our centre.

The patient attended hospital irregularly and by 1980, he had developed proptosis, central cyanosis and digital clubbing. The arterial pulses were still feeble (BP,80/65mmHg) and systolic pulsations were noted in the liver and jugular veins. The heart sounds had become muffled and the pulmonary stenosis murmur noted previously was no longer audible. Furthermore, he had developed massive ascites, but had very minimal pedal oedema. A repeat chest radiograph revealed gross cardiomegaly (CT ratio 83%)and pulmonary oligaemia (Fig 1A) but the ECG findings were unchanged. These new developments aroused a suspicion of right ventricular EMF.

Cardiac catheterizatiom became imperative in view of the uncertainty about the diagnosis. Right heart catheterization revealed an elevated right ventricular pressure (100/10mmHg) and a systolic pressure gradient of 80mmHg across the pulmonary valve. Angiogram of the right ventricle confirmed the presence of gross tricuspid regurgitation; both the atrium and ventricle were dilated, but the latter contained no filling defects (Fig 1B). The pulmonary valve appeared thickened and the main pulmonary artery was dilated. Left heart studies revealed no structural abnormalities. EMF was thus excluded, while the initial diagnosis of pulmonary valve stenosis was confirmed. Transarterial pulmonary valvotomy was performed in March 1982, with satisfactory results.

Case 2

The second patient, also a boy, presented at the age of 10 years. The parents recalled that he was rather tachypnoeic as an infant, but since he had developed normally, medical advice was not sought. Five weeks before referral to the UCH, he presented at another hospital complaining of dyspnoea, lassitude and generalized swelling of the body. Examination there, revealed signs of severe heart failure, with generalized oedema, engorged jugular veins and an enlarged liver, 10cm below the right costal margin. No cardiac murmur was heard. A chest radiograph showed a globular heart (CT ratio 75%) with slightly increased pulmonary vascular markings. A provisional diagnosis of biventricular EMF was made and he was then referred to the UCH where the previously noted signs were confirmed. Other signs elicited included central cyanosis, digital clubbing and peri-orbital hyperpigmentation, all of which are characteristic findings in EMF.[12] There was a right ventricular heave and the arterial pulses were feeble (BP 100/80 mmHg), but synchronous. A pulmonary ejection click was present; the pulmonary closure sound was

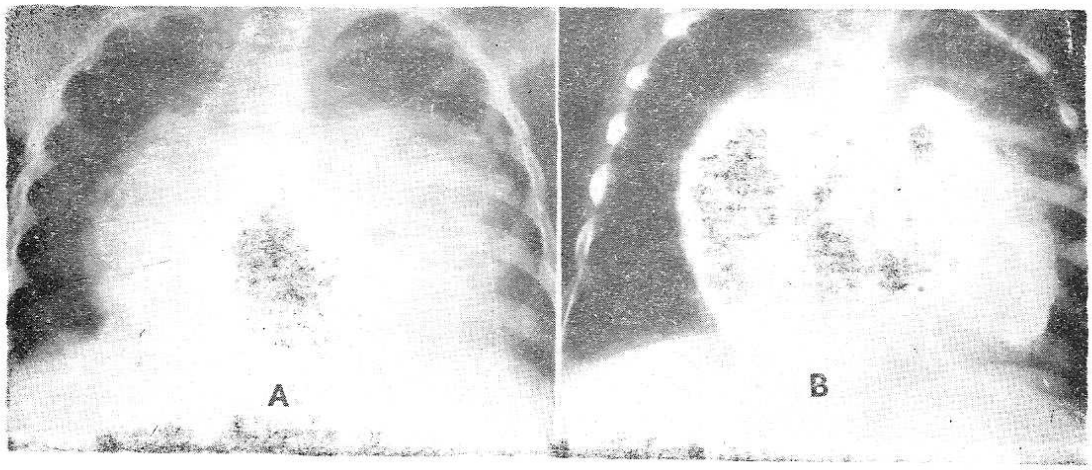

Fig 1. A chest radiograph of Case 1 with severe pulmonary stenosis showing gross cardiomegaly and pulmonary oligaemia (Fig 1A). Right ventricular angiogram (Fig 1B), showing normal ventricular apex and dilated main pulmonary artery.

accentuated, but no murmurs were heard. The ECG revealed right ventricular hypertrophy.

The globular heart, absence of cardiac murmurs and the extracardiac signs were suggestive of right ventricular EMF, but the presence of pulmonary hypertension and RVH militated against this diagnosis. Left ventricular EMF with severe pulmonary hypertension and right-sided heart failure seemed more probable.

At right heart catheterization, the right ventricular (RV) pressure (120/20 mmHg) was higher than the systemic pressure (100/80 mmHg), but the main pulmonary artery could not be entered because bradycardia and hypotension developed each time the catheter was placed in the infundibulum. Angiocardiography revealed a hypertrophied, dilated and poorly contractile RV. There was tricuspid regurgitation, but no ventricular filling defect. The left ventricle (LV) was dilated, hypertrophied and hypocontractile. Based on these findings, a diagnosis of pulmonary valve stenosis with gross myocardial disease was made. However, subsequent two-dimensional echocardiography carried out in a London hospital revealed the presence of persistent ductus arteriosus (PBA) and preductal coarctation of the aorta. The pulmonary valve appeared normal. Surgery was not attempted because of the very poor myocardial function. He died of heart failure shortly afterwards.

Necropsy confirmed the echocardiographic diagnosis. The aortic isthmus was extremely hypoplastic and the descending aorta was aligned with a large PDA (Fig. 2) and an atheromatous dilated main pulmonary artery. All the cardiac chambers were grossly hypertrophied and dilated. There was haemorrhagic necrosis of the LV endocardium. Both old and fresh infarcts were present in the ventricles as well as in the interventricular septum. There were neither septal defects nor features of EMF.

Case 3

The third patient, a 10-year old boy, first attended the clinic in 1975 with heart failure, gross tricuspid incompetence, bilateral proptosis, central cyanosis, muffled heart sounds and massive ascites with minimal pedal oedema, all

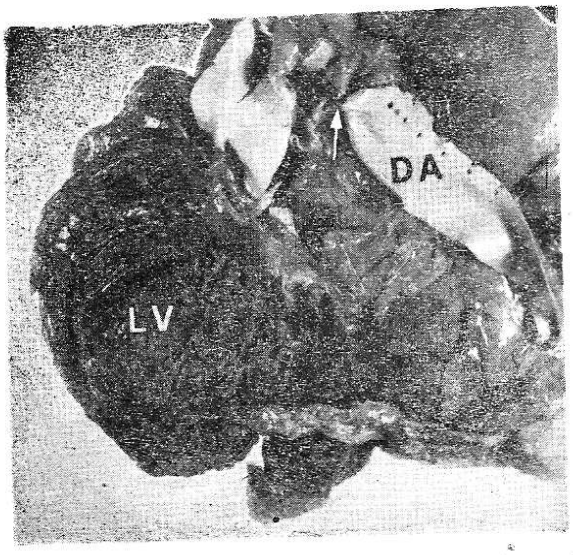

Fig 2. A necropsy specimen of the left ventricle of Case 2 who had preductal coarctation, patent ductus arteriosus, pulmonary hypertension and myocardial infarction. Note the hypertrophied left ventricle (LV) and the descending aorta (DA) that is aligned to the ductus arteriosus (arrow).

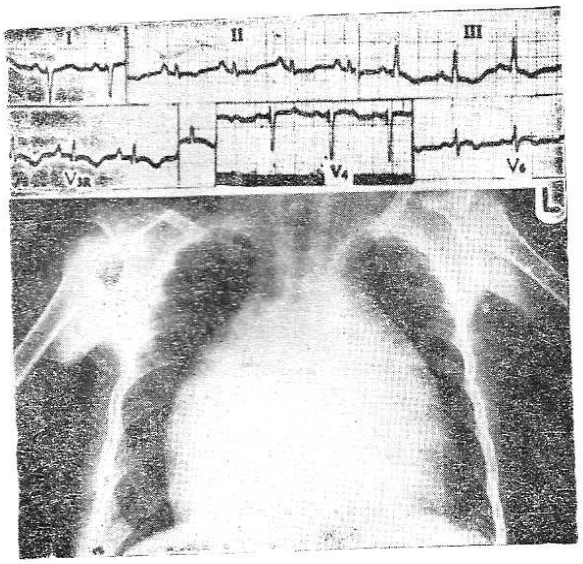

Fig 3. ECG (top) and chest radiograph (bottom) from a boy (Case 3) with dilated cardiomyopathy. Note that dwarfed QRS complexes, qr pattern in V3R and T-wave inversion in leads II, III, V_4 and V_6. The heart is globular and the lung fields are oligaemic.

of which are common findings in right ventricular EMF.[12] Chest radiograph revealed a globular heart (CT ratio, 72%) and oligaemic lung fields, while the ECG showed a qr pattern in V3R, dwarfed QRS complexes in precordial leads and widespread inversion of T-waves (Fig. 3). These ECG features were also characteristic of EMF.[13] But the findings at right heart catheterization were atypical of this condition. The pressures in the right atrium and ventricle were identical (RV pressure, 15/5mmHg) and there was marked regurgitation of contrast material into a dilated right atrium, findings which are usually seen in EMF. The RV was dilated, but it contained no filling defects (Fig. 4). A left ventriculogram was normal. It was therefore, concluded that the patient had either a dilated cardiomyopathy or patchy EMF with distortion of the tricuspid valve. Treatment with digoxin and diuretics was commenced.

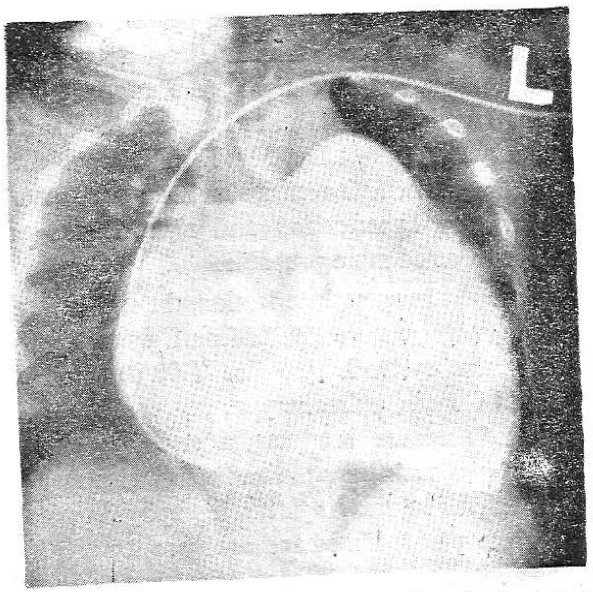

Fig 4. Right ventricular angiogram in Case 3, showing a dilated ventricle. Note the absence of filling defects.

During a 6-year follow-up, more characteristic features of right ventricular EMF evolved; these features included intermittent atrial fibrillation, recurrent pericardial effusion and massive refractory ascites which necessitated repeated abdominal paracenteses. These developments strengthened the suspicion that typical morphological features of right ventricular EMF might have evolved since the first cardiac catheterization. The procedure was therefore, repeated in February 1982, but the findings were identical with those obtained at the first study. He died later that year, of severe hyponatraemia.

At necropsy, the right atrium and ventricle were hypertrophied and dilated; the tricuspid annulus measured 15cm, the valve leaflets were normal, but the papillary muscles were hypertrophied. There were no signs of EMF or pulmonary vascular disease. Left heart structures were grossly normal, except for a moderate dilatation of the LV cavity and mitral valve. Histology revealed features of dilated cardiomyopathy[14] mainly in the RV.

Case 4

The fourth patient was an 11-year old boy who first presented with hepatomegaly of obscure aetiology. Three months later, clinical signs of tricuspid regurgitation and heart failure became evident. There was, however, on cardiac murmur. Chest radiographs revealed no pericardial calcification, but the heart was globular (CT ratio, 64%) and there was a right pleural effusion. The ECG findings consisted of biatrial hypertrophy, low-amplitude QRS complexes and widespread inversion of T-waves. Right ventricular EMF was considered as the diagnosis and confirmation of this was sought at cardiac catheterization.

Intracardiac studies confirmed the presence of tricuspid regurgitation. This valve defect prevented a clear definition of RV morphology during ventriculography, because most of the contrast material regurgitated into a dilated right atrium. However, the infundibulum was dilated whereas the ventricular body and apex appeared obliterated. These findings were interpreted as confirmatory evidence of right ventricular EMF. He was managed conservatively and kept symptom-free for about $3\frac{1}{2}$ years, after which he defaulted from the clinic. He reappeared later, in severe heart failure and died shortly after re-admission.

Necropsy revealed constrictive pericarditis. The parietal pericardium, 8mm thick, was separated from a fibrotic, calcified but smooth-surfaced visceral layer by 225ml of serous fluid. Both atria were dilated, so also were the tricuspid and mitral valve rings (circumference, 12.5 and 10cm respectively). The RV was severely constricted (Fig. 5); the LV was also constricted, but not as severely as the RV. There was no evidence of EMF.

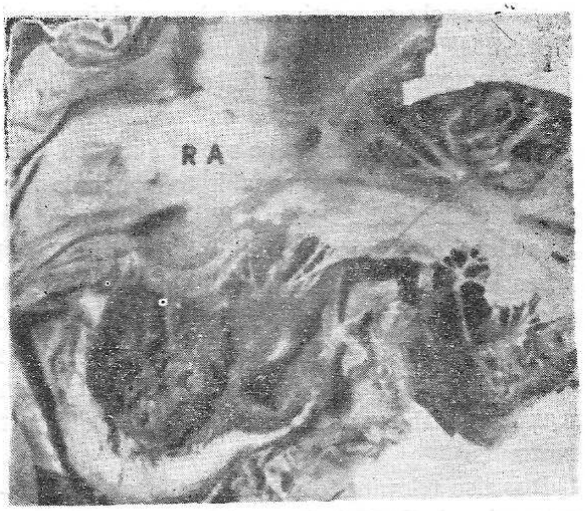

Fig 5. Necropsy specimen of the right heart in Case 4. Note the dilated right atrium (RA) and shrunken ventricular cavity.

Discussion

Liver cirrhosis, nephrotic syndrome and abdominal tuberculosis sometimes produce massive ascites like the type seen in right ventricular EMF but, unlike the latter, these other conditions are rarely associated with gross cardiac dilatation. Ebstein's anomaly, a rare malformation, is

morphologically similar to right ventricular EMF, but only once has it been confused with EMF.[6] Therefore, for practical purposes, the differential diagnosis of EMF is limited to other relatively common causes of a globular heart without cardiac murmurs. These include severe dilated cardiomyopathy, effusive tuberculous pericarditis and those cases of constrictive pericarditis in which there is impressive cardiac dilatation. These cardiac diseases are, however, usually associated with pulmonary venous congestion,[2 15] in contrast to the pulmonary oligaemia which is invariable in right ventricular EMF.[2 16] That was the rationale behind the popular dictum that a combination of pulmonary oligaemia and a globular heart without cardiac murmurs should, in the older African child or adolescent, signify right ventricular EMF until proved otherwise.[11] Such a combination was present in Cases 1, 3 and 4, while the cardiovascular findings in Case 2 were strongly suggestive of advance left ventricular EMF.

It is noteworthy that central cyanosis developed in Cases 1, 2 and 3. Since Cases 1 and 2 apparently became symptomatic at the age of 8 and 10 years respectively, it was reasonable to conclude that these patients had acquired heart disorders. The absence of murmurs in Case 2 and in Case 1 when he was re-assessed in 1980, further strengthened that view and seemingly reinforced the diagnosis of EMF, this being the only notable acquired cardiopathy in childhood that often manifests with cyanosis and an absence of heart murmurs.

It is also relevant to note that the arterial desaturation in EMF is due mainly to a ventilation/perfusion imbalance which results from wasted perfusion of lung segments that are compressed by the huge heart.[17] This physiological shunt is often further compounded by the reduction in pulmonary blood flow caused by impaired right ventricular contractility. These factors were replicated in Case 1 who became cyanosed when the CT ratio increased from 62 to 83% and the myocardial function had declined to such an extent that the murmur of pulmonary valve stenosis was no longer audible.

Case 2 was remarkable in many other respects. At necropsy, the aortic isthmus was barely probe-patent. Therefore, for all practical purposes, he had an interrupted aortic arch and the RV had to generate systemic pressure in order to pump desaturated blood through the PDA into the descending aorta, thus causing the RVH, pulmonary hypertension and clubbing of the toes. The cyanosed tongue and clubbed fingers in this patient indicated that the left ventricular blood was also desaturated. This, in the absence of a septal defect or transposed great arteries, meant that the pulmonary venous blood was undersaturated. In this instance, such an event could only have resulted from impaired diffusion caused by pulmonary congestion, or a ventilation perfusion imbalance caused by the enlarged heart. The latter, as stated earlier, is the cause of cyanosis in EMF.

The clinical and autopsy findings in the patient suggest that the course of events was as follows: during infancy and early childhood, the systolic pressure load imposed by coarctation led to left ventricular hypertrophy and upper limb hypertension. Over the years, these two complications became more severe, while the myocardial oxygen supply became progressively reduced, because of the relatively inadequate coronary flow and the arterial unsaturation, thus causing myocardial ischaemia and infarction. With the onset of gross myocardial disease and heart failure, the systolic pressure in the LV, ascending aorta and the brachiocephalic arteries declined (the so-called decapitated hypertension) and the murmurs of the associated PDA and coarctation became inaudible.

The uncertainties in the clinical diagnosis in Cases 1 and 2 arose from the late presentation and the disappearance of cardiac murmurs, because of advanced disease, in what were really congenital cardiac malformations which usually manifest with characteristic murmurs. The value of the ECG in both cases was however, remark-

able. In the two cases, the ECG revealed RVH, an extremely uncommon event in children with right ventricular EMF[13] and thus cast serious doubts on the accuracy of the clinical diagnosis (EMF) in both patients. Therefore, if confronted with a similar case, the clinician should evaluate the ECG very carefully.

Cases 3 and 4 simulated EMF more closely than the other two cases because the physical signs, radiographic and ECG findings were indistinguishable from those of right ventricular EMF. Angiocardiography was somewhat helpful in Case 3, but not in Case 4, whose RV was severely constricted, resulting in partial obliteration of its cavity and dilatation of the right atrium and tricuspid valve ring. The combination of a small RV cavity, tricuspid regurgitation and dilated right atrium, completely simulated the previously published angiocardiographic features of right ventricular EMF.[2 13] A clear demonstration of ventricular filling defects should therefore, be a pre-requisite for the angiocardiographic diagnosis of right ventricular EMF. In the presence of poor ventricular opacification and indistinct filling defects, diagnosis can be made with certainty only if other characteristic features are present. These features include radiographic evidence of calcified endocardium in the right ventricular infundibulum,[2 16 19] a qr pattern in leads V3R or V1[13] and echocardiographic evidence of sudden obliteration of the ventricular cavity or apex.[5 20-22]

Acknowledgements

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