Echocardiography in the diagnosis of right ventricular endomyocardial fibrosis

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Summary

Twenty-one patients with endomyocardial fibrosis (EMF) and right ventricular involvement were studied by M-mode echocardiography. All 21 patients showed echocardiographic findings consisting of (i) increased right ventricular dimension, (ii) paradoxical septal motion, (iii) increased right ventricular outflow dimension (iv) thickening of the right ventricular anterior wall with increased right ventricular anterior wall motion, and (v) easily recordable tricuspid valve. In addition, some of the patients had posterior pericardial effusion, and fine fluttering of the tricuspid valve. EMF was diagnosed clinically in all the patients, haemodynamically and angiographically in 15 and confirmed at autopsy in one.

None of these echo findings was present in two patients with constrictive pericarditis, and two patients with massive ascites due to portal hypertension.

Introduction

The clinical features of right-sided endomyocardial fibrosis are well recognized (Shillingford and Somers, 1961; Parry, 1976). These features however become obvious only with advanced right ventricular involvement. Problems in diagnosis often arise, when it may be difficult to distinguish at the bedside between endomyocardial fibrosis (EMF), constrictive pericarditis, rheumatic valvular disease, and congestive cardiomyopathy (Falase, Kolawole and Lagundoye, 1976). Congestive cardiomyopathy and rheumatic valvular disease can be distinguished by their echocardiographic patterns (Millward *et al.*, 1973; Segal, Likoff and Kingsley, 1966), and by cardiac catheterization and angiography. Constrictive pericarditis and right-sided EMF on the other hand have similar haemodynamic patterns (Somers *et al.*, 1968).

If characteristic echocardiographic patterns of

right ventricular EMF could be defined, it would enable clinicians to make a bed-side diagnosis, obviating the need for cardiac catheterization and angiography, with their attendant risks. Since the role of surgery in this disease is not yet clearly defined, a method of diagnosis without attendant risks would be most desirable. Furthermore if, it could prove possible with echocardiography to diagnose EMF much earlier, before the classical clinical features present, surgery could perhaps be more beneficial.

We here report our observations on M-mode echocardiographic studies on 21 Nigerian patients with right-sided EMF.

Subjects and methods

The 21 patients (11 females, and 10 males) were referred to the cardiac clinic of the Lagos University Teaching Hospital with diagnoses varying from unexplained cardiomegaly, and EMF, to pericardial effusion. Each patient was examined clinically, followed by electrocardiographic (ECG) and radiological examination. Ultrasound examination was carried out, using a standard technique, with an SKI Ekoline 20 ultrasonograph. A 2.25 mHz transducer, with a repetition rate of 1000 impulses/sec was placed in the 2nd, 3rd or 4th left parasternal space, with the patient supine. Echocardiograms were obtained on Kodak direct print or photographic paper on a Cambridge strip-chart recorder.

Fifteen patients had cardiac catheterization and angiography. In addition, 2 patients who had phonocardiograms underwent aortic root angiography. Autopsy was obtained in one patient. For comparison, two patients with constrictive pericarditis, and 2 patients with massive ascites due to portal hypertension were also studied echocardiographically.

Results

Nineteen patients presented with the characteristic clinical picture of right ventricular EMF, namely elevated jugular venous pressure, disproportionately massive ascites, and minimal pedal oedema. In 2 cases, the patients presented with only mild to moderate ascites. Fifteen patients had clinical evidence of tricuspid regurgitation, 12 had a pansystolic murmur of mitral regurgitation, and an early diastolic murmur was heard in 2 patients. Eighteen were in sinus rhythm, and 3 in atrial fibrillation. There was no ECG evidence of left bundle branch block. Of the fifteen who were studied haemodynamically and angiographically, 9 had isolated right ventricular EMF. The remaining 6 had left as well as right ventricular involvement. The 2 patients with an early diastolic murmur had normal aortic root angiograms.

Certain echocardiographic features were observed in all 21 patients. Those features, thought to be associated with right ventricular EMF, were,

- (i) increased right ventricular dimension,
- (ii) paradoxical septal motion (Fig. 1),
- (iii) increased right ventricular outflow tract dimension (Fig. 2),
- (iv) thickening of the right ventricular anterior wall, with increased right ventricular anterior wall motion (Fig. 3), and
- (v) easily recordable tricuspid valve (Fig. 4).

In addition, 3 patients had posterior pericardial effusions (Fig. 1), and 2 patients (who had early diastolic murmurs) had fine fluttering of the tricuspid valve (Fig. 4).

In contrast, none of these echocardiographic features were seen in the patients with constrictive pericarditis or with massive ascites due to portal hypertension.

FIG. 1. Echocardiogram of right-sided endomyocardial fibrosis, showing increased right ventricular ("RV") dimension, paradoxical septal motion (IVS) and posterior pericardial effusion (pe). LV = left ventricle; plvw = posterior left ventricular wall.

FIG. 2. Echocardiogram of right ventricular EMF showing dilated right ventricular outflow tract (R.V.O.T.). LA = left atrium; Ao. R. = aortic root; IVS = intraventricular septum.

Discussion

The main feature of right-sided EMF is deep scarring of the endocardium and subendocardial layers of the right ventricle, usually starting at the apex and spreading to the inflow tract. The fibrosis eventually affects the papillary muscles, the chordae tendinae, and the posterior cusps of the tricuspid valve, leading to tricuspid regurgitation. Although there is apical obliteration and distortion of the cavity, the right ventricular outflow tract is dilated. Angiographic studies (Cockshott, 1965; Hess *et al.*, 1978) have demonstrated vigorous pulsation of the infundibulum of the right ventricle, tricuspid regurgitation, and grossly dilated right atrium.

The structures of the right heart are easily recorded in diseases with right ventricular volume overload such as atrial septal defect, Ebstein's anomaly, and tricuspid regurgitation. Echocardiograms in such instances show increased right ventricular dimension (McCann, Harbold and Giuliani, 1972) and paradoxical septal motion (Meyer *et al.*, 1972). Paradoxical septal motion has also been associated with left

bundle branch block (Abbasi *et al.*, 1974) anteroapical myocardial infarction (Feigenbaum, 1972a), and in postoperative patients after median sternotomy (Feigenbaum, 1972b). The increased right ventricular dimension, and paradoxical septal motion in our patients with right-sided EMF may in part be due to the associated tricuspid regurgitation. This does not however explain the finding of the dilated right ventricle in those patients without tricuspid regurgitation. A possible explanation may be that, as a result of distortion of the heart and its axis, the ultrasound beam may be partly traversing the dilated right atrium and not only the right ventricle (Fig. 5). Apical obliteration of the cavity in right ventricular EMF has been reported by Hess *et al.* (1978). The dilated right ventricular outflow tract, the vigorous anterior right ventricular wall contraction, and the pericardial effusion demonstrated by echo, are in keeping with the pathological and angiographic findings in this disease. Strong echoes from the right ventricular endocardium has been observed by Hernandez-Pieretti (1977). This was not demonstrated in

FIG. 3. Echocardiogram of right ventricular EMF showing thickening of the right ventricular anterior wall (RVAW), and strong right ventricular anterior wall contraction.

FIG. 4. Tricuspid valve (TV) echocardiogram in right ventricular EMF showing fine fluttering of the anterior leaflet.

any of our patients. It may be that this finding is only associated with calcification of the endocardium.

The aortic and pulmonary valves are not affected in EMF. An early diastolic murmur has, however, been reported in a few cases of haemodynamically and angiographically proven EMF (Shillingford and Somers, 1961; Hess *et al.*, 1978). In the 2 patients in our series with biventricular EMF in whom an early diastolic murmur was heard, aortic root angiogram was normal. Fine fluttering of the anterior tricuspid and aortic valve leaflets was observed in the echocardiogram of the 2 patients. Tricuspid valve fluttering in diastole has been reported in pulmonary regurgitation, atrial septal defect with large shunts, atrial fibrillation, and atrial flutter. The tricuspid valve

fluttering in our patients may be due to the influence of the jet stream from the strongly contracting right atrium against the anterior tricuspid leaflet, the posterior leaflet having been tethered down by the fibrosed chordae.

Although M-mode echocardiography shows the degree of increased right ventricular dimension to a certain extent, calculation of the right ventricular volume is difficult because of the unusual geometry of the chamber. Two-dimensional echocardiography provides more exact data for the evaluation of right ventricular size and volume. Newer techniques such as compound-B ultrasonography, which provides a two-dimensional cross-sectional image, and multi-scan echocardiography may provide a more accurate means of diagnosis.

FIG. 5. Schematic representation of possible basis of increased 'right ventricular' dimension in right ventricular EMF (see text). CW=chest wall; T=transducer; RV=right ventricle; S=sternum; ARAW=anterior right atrial wall; RA=right atrium; RV=right ventricle; IVS=interventricular septum; LV=left ventricle; LA=left atrium; PLVW=posterior left ventricular wall; USB=ultrasonic beam.

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