INTRACARDIAC CALCIFICATION IN ENDOMYOCARDIAL FIBROSIS

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The finding of calcification on radiological examination of the heart is well recognized in mitral stenosis and aortic stenosis, constrictive pericarditis, syphilitic aortitis, and calcific coronary artery disease. Extensive calcification of the myocardium is rare apart from that occurring in a cardiac aneurysm. In each of these conditions, calcification is associated with a particular structural localization. Extensive intracardiac calcification has also been observed in endomyocardial fibroelastosis of infancy and adolescence (van Buchem et al., 1959). Recently a case has been reported of calcification occurring at the left ventricular apex, probably in a lesion primarily inflammatory in origin, in a young woman with no cardiac symptoms whatsoever (Barber and Murphy, 1961).

Calcification in endomyocardial fibrosis, as known in Uganda, has hitherto only been recognized at necropsy (Davies and Ball, 1955). In advanced cases of constrictive endocardial fibrosis, calcium deposition occurs in areas of extensive fibrosis particularly at the right ventricular apex. Calcification has not, as far as we know, been recognized in clinical radiography of the disease. The purpose of this report is to describe two patients with endomyocardial fibrosis in whom calcification was recognized at routine chest radiography.

Case 1. A Ruanda woman, aged 18 years, gave a history of cough and dyspnea on exertion, palpitation, precordial pain, abdominal swelling, and ankle edema for two weeks. There were no past illnesses of note.

Examination. Physical signs of congestive heart failure with considerable venous engorgement and hepatic distension were present. Scattered rhonchi were audible at both lung bases. The pulse rate was 100 a minute and regular. The blood pressure was 95/60. The apical impulse was quiet and was displaced to the left. A loud third sound was heard over the mid-precordium; there were no murmurs. The liver was enlarged four fingers' breadth and was tense. The spleen was palpated four fingers' breadth below the costal margin. Moderate ascites and ankle edema were present.

Electrocardiogram. T inversion was present in leads I, II, VL, and VF and was conspicuous in leads V2–V6. Clockwise rotation was present and there were prominent R waves in V2 and V3 (Fig. 1).

Radiography. Plain films of the chest showed generalized cardiac enlargement (Fig. 2 and 3). Both atria were prominent. Within the heart shadow and approximately an inch from the left heart border there were patches of amorphous calcification about 2.5 cm. in length. Screening and coronal and sagittal tomo-grams confirmed that the calcification was intracardiac and probably in the region of the apex of the right ventricle (Fig. 4 and 5). The pulmonary arteries looked normal.

Clinical Diagnosis. Endomyocardial fibrosis with predominant right ventricular constriction, based on the findings of gross jugular and hepatic engorgement and right ventricular gallop.

Progress. Response to in-patient treatment with digitalis and mercurial diuretics was fairly satisfactory. The patient was discharged but failed to return until three months later when she was readmitted with a recurrence of congestive failure. An additional physical sign in the cardiovascular system was a transient tricuspid systolic murmur, varying with respiration, which disappeared after a few days in hospital. The radiological and electrocardiographic signs were essentially unchanged. There was no finger clubbing.
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**Fig. 1.**—Case 1. Electrocardiogram showing T wave inversion in leads I, II, VL, and VF and leads V2-V6. R wave is prominent in V2 and V3.

**Fig. 2.**—Case 1. X-ray of chest, showing prominence of both atria. There are patches of calcification internal to the left heart border.

**Fig. 3.**—Case 1. Right oblique X-ray of heart showing enlargement of left atrium and patches of intracardiac calcification.
The patient was anaemic (Hb 8.2 g/100 ml.) and slightly jaundiced (serum bilirubin 2.3 mg./100 ml.). She had also a mild fever (about 100°F) and a reticulocytosis of 5 to 7 per cent on most occasions. The red cells showed a tendency to spherocytosis. No malarial parasites were seen. White cells numbered 4900/cu. mm. (polymorphs 53%, lymphocytes 43%, eosinophils 3%, basophils 1%). Direct and indirect Coombs' tests were negative. Erythrocyte sedimentation rate (Westergren method) was 25 mm./hour. Liver function tests and biopsy were normal. Stool examination showed no parasites or evidence of occult blood loss. The Mantoux test was negative. Blood urea was 19 mg. per 100 ml.

It was concluded that haemolytic anaemia was an additional feature. The splenomegaly and haemolytic anaemia were probably unrelated to her cardiac condition. The patient improved on oral iron and prednisone. She was discharged on maintenance treatment but regrettably was never seen again.

Case 2. A Ganda man, aged 40 years, gave a four-week history starting with pain and swelling under the right costal margin and later effort dyspnea and cough.

Examination. Physical examination showed an orthopnoeic patient with signs of congestive heart failure. The jugular veins were distended 8 cm. above the sternal angle and showed systolic pulsation. The liver was four fingers' breadth enlarged with a palpable systolic pulsation. Moderate ascites was present. The blood pressure was 120/80. The pulse rate was 80 a minute and it was regular in rate and volume.

The apical impulse was quiet and poorly palpable to the left of the mid-clavicular line. A high pitched pansystolic murmur, maximal in early systole and radiating to the axilla, was heard at the mitral area. A split second sound was heard just internal to the apex. At the tricuspid area an early systolic murmur was noted.

Electrocardiogram. Low voltage in the standard and limb leads with incomplete left bundle-branch block (Fig. 6).

Phonocardiogram. The mitral and tricuspid murmurs were recorded as auscultated and a split second sound was clearly demonstrated.

Radiography. A plain chest X-ray showed a generalized enlargement of the heart with an ovoid mass of calcium about 2.5 cm. internal to the left border of the heart (Fig. 7). Fluoroscopy demonstrated that the area of calcium was intracardiac and located in the region of the right ventricular apex. Coronal and left oblique tomograms confirmed the location of the calcification (Fig. 8).

Clinical Diagnosis. Endomyocardial fibrosis with mitral regurgitation, constriction of the right ventricle, and some tricuspid regurgitation.

Progress. The patient was treated with digitalis and chlorothiazide diuretics to which response was
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FIG. 6.—Case 2. Electrocardiogram showing incomplete left bundle-branch block.

FIG. 7.—Case 2. Plain film of chest showing an ovoid mass of calcium internal to the left heart border.

FIG. 8.—Case 2. Left oblique tomograph showing intracardiac calcification.
moderately satisfactory. The liver did not recede much in size and the jugular venous engorgement remained essentially unaltered, presumably owing to the structural lesion of right ventricular constriction. The orthopnoea and pulmonary congestion were much relieved by treatment.

**DISCUSSION**

Endomyocardial fibrosis is a common cardiopathy in Uganda. Clinical diagnosis is reached by exclusion of other causes of heart failure, and the recognition of certain syndromes (Ball et al., 1954), associated with distinctive auscultatory signs (Somers and Williams, 1960) and electrocardiographic findings (Williams and Somers, 1960). Recent haemodynamic studies (Shillingford and Somers, 1961) have correlated pathological and clinical observations. In the two patients described here, the diagnosis of endomyocardial fibrosis was based on the absence of any other cause, the syndrome of right ventricular constriction, suggestive electrocardiographic findings, and radiological calcification in the right ventricular cavity consistent with right ventricular constriction. In the second patient left ventricular endocardial fibrosis was also likely to be present in view of the presence of mitral regurgitation.

Radiologically, the irregular appearance of the calcified mass and its localization within the area of the right ventricular apex were distinguishing features in contrast with the linear shadow of pericardial calcification or the valvular localization of calcification in mitral or aortic stenosis.

These cases illustrate the importance of seeking calcification in establishing the clinical diagnosis of endomyocardial fibrosis. Although the chance of finding radiological calcification in plain films is small, tomography, when appropriate, may be expected to reveal it in a proportion of cases.

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**REFERENCES**