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Loasis as a possible trigger of African endomyocardial fibrosis: a case report from Nigeria

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Summary

A Nigerian boy, previously in good health, presented with a two-day history of fever with chills; followed on the third day by periorbital swelling, urticarial rash and itching; and on the sixth day by dyspneoa, abdominal swelling and leg swelling. There was clinical, radiological and electrocardiographic evidence of dominant right-sided heart failure. Loa-loa was isolated from the blood and eosinophilia was marked but both were cured by diethylcarbamazine therapy. Heart failure, however, persisted and ended fatally 25½ months later. Endomyocardial fibrosis, more severe on the right sided chambers, but affecting both ventricles was diagnosed. Evidence is presented from the literature to indicate loasis as the trigger of endomyocardial damage in this patient.

Key words: loasis; eosinophilia; African endomyocardial fibrosis.

Introduction

Endomyocardial fibrosis (E.m.f.) is common in the tropical rain forest belt of Africa where it accounts for 10–15% of heart disease (Davies, 1956, 1961). Its cardiac morphology is very similar to the morphology of the eosinophilic cardiomyopathies (Roberts et al., 1969; Brockington and Olsen, 1973). Evidence derived mainly from European residents in Africa who developed E.m.f. appears to incriminate parasite – induced eosinophilia in causation of E.m.f. (Brockington et al., 1967; Gerbaux et al., 1957). Among African patients, Ive et al. (1967) presented evidence associating E.m.f. with Loa-loa induced eosinophilia, however on extension of these initial observations the evidence for eosinophilia did not hold-up (Brockington, 1974). But most of the patients that they studied had well established E.m.f. disease.

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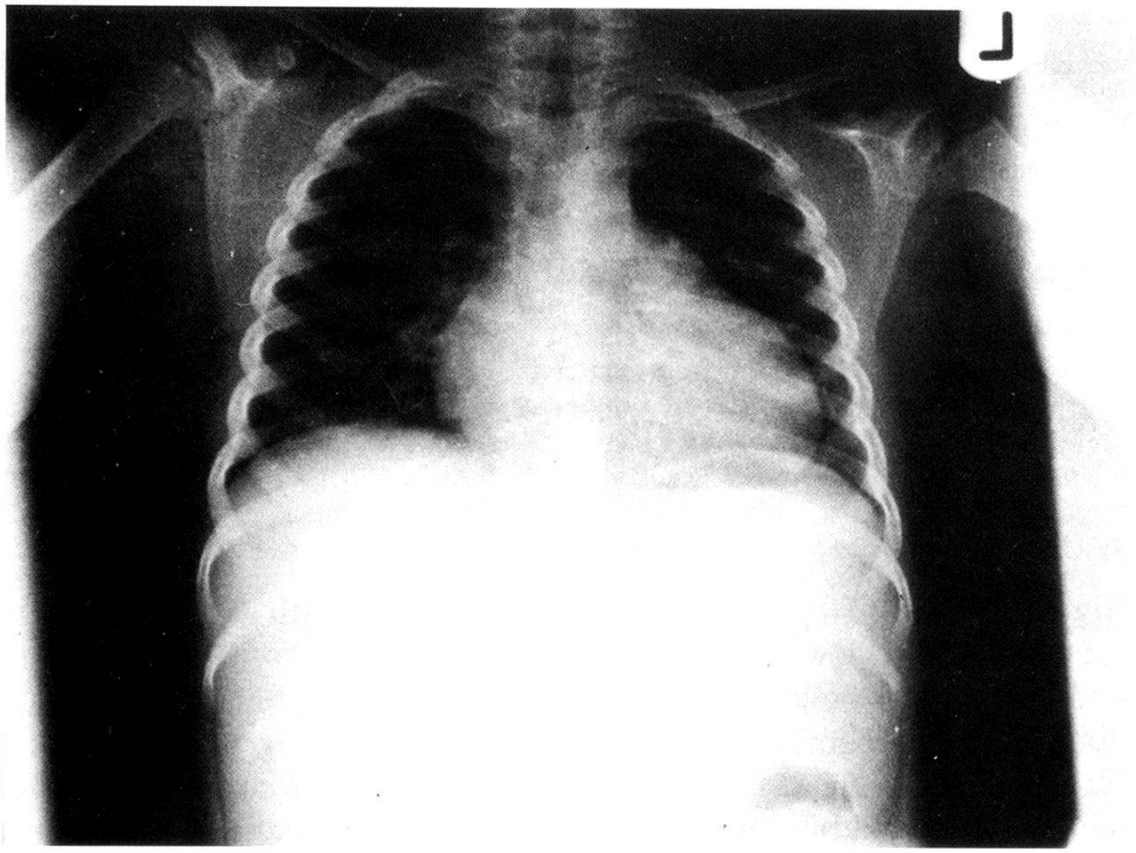


Fig. 1. Chest radiogram of the patient, showing cardiomegaly. Lung fields are not congested despite the fact that the patient had moderate degree of ascites and venous distention. Lateral radiogram (not shown) revealed that cardiomegaly was predominantly right-sided.

Early disease of E.m.f. among Africans is not clinically recognizable, and since eosinophilia induced by parasitic diseases is not usually permanent, the presence or absence of eosinophilia in chronic E.m.f. (if eosinophilia plays a role) could depend on how long it takes for the chronic clinical features to evolve, following the initial insult.

Recently, we presented evidence of a high incidence of recent onset myocardial disease in patients with severe parasite (Loa-loa) induced eosinophilia. We also showed that 73% of this associated heart disease evolved typical clinical features of E.m.f., long after the eosinophilia had been returned to normal by diethylcarbamazine therapy (Andy et al., 1980). In this communication we present clinical details of one of these patients who came to autopsy, to highlight the possible role of loasis in aetiology of E.m.f., and to call attention to some peculiar clinical features of early African E.m.f.

Case report

F. O. (W. G. H. No. 5962) was a normal child when he was born in the University Hospital of Ife, Nigeria, on January 28, 1972. He attended the infant and child welfare clinic of this hospital regularly and was in good health except for occasional episodes of malaria and mild diarrhoea treated as outpatient.

He developed a low-grade fever with chills on July 28th, 1977. Periorbital and facial swelling with itching which were worse in the mornings followed three days later. About the same time he

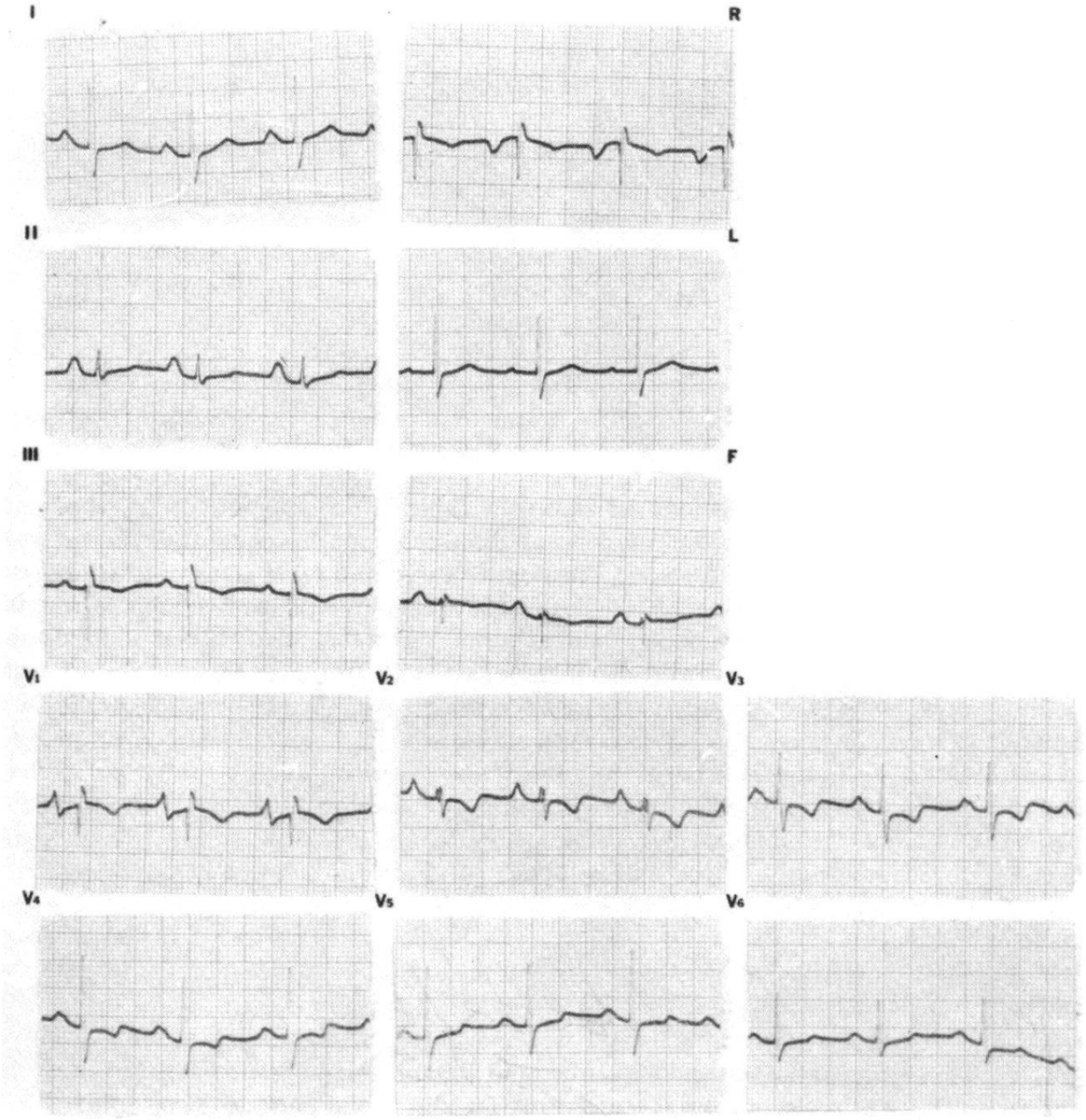


Fig. 2. Electrocardiogram: right atrial hypertrophy and incomplete right bundle branch block. The ECG findings did not change throughout the duration of illness.

had transient itchy urticarial-type rash which lasted about 2 days. About August 2nd 1977, he developed dyspnea on exertion, followed soon after by abdominal and leg swelling. There was no orthopnea, no nocturnal dyspnea, no cough and no chest pain, no prior history of sorethroat or arthritis. He was admitted on August 4th. 1977 and physical examination revealed a well-nourished and well-developed child in slight respiratory distress. The face and periorbital areas were puffy but without proptosis. Neck veins were distended and the «Y» descent was brisk. An abnormal systolic retraction was visible in the second left intercostal space. Cardiac apex was located in the 5th intercostal space but was displaced to the anterior axillary line. A left para-sternal heave, a quadruple rhythm and a grade III to IV tricuspid regurgitant murmur with an accentuated pulmonic closure sound were present. Moderate degree of ascites was present, and the liver and spleen were enlarged. Pedal oedema was slight. Low grade pyrexia with temperatures of 99°–100° F was present during the first two hospital days. No skin rash was observed in the hospital. The chest was clinically clear and the chest radiogram (Fig. 1) showed a moderate cardiomegaly, with relatively clear lung fields. ECG showed incomplete right bundle branch block (r.b.b.b.) and right atrial hypertrophy (Fig. 2).

Table 1. Results of laboratory investigations

Date	4/8/1977	29/8/1979
Haematocrit	37%	39%
White blood count	13,000/mm^3	15,000/mm^3
Total eosinophil count	3,040/mm^3	240/mm^3
Microfilariae:		
Thick film	negative	negative
Concentration technique	Loa-loa	negative
Blood culture	sterile	sterile
Malarial parasite	negative	negative
Stool ova and parasites	negative	negative
Urine microscopy	normal	normal

Results of laboratory investigations are shown in Table 1. Eosinophilia was marked and microfilaria Loa-loa was demonstrated by the concentration technique. Clinical improvement with frusemide and digoxin therapy was dramatic, the ascites cleared and other signs of heart disease improved. The child was discharged on the 14th hospital day. Therapy with diethylcarbamazine 100 mg/daily for 21 days returned the eosinophil count to normal and it remained normal till his death.

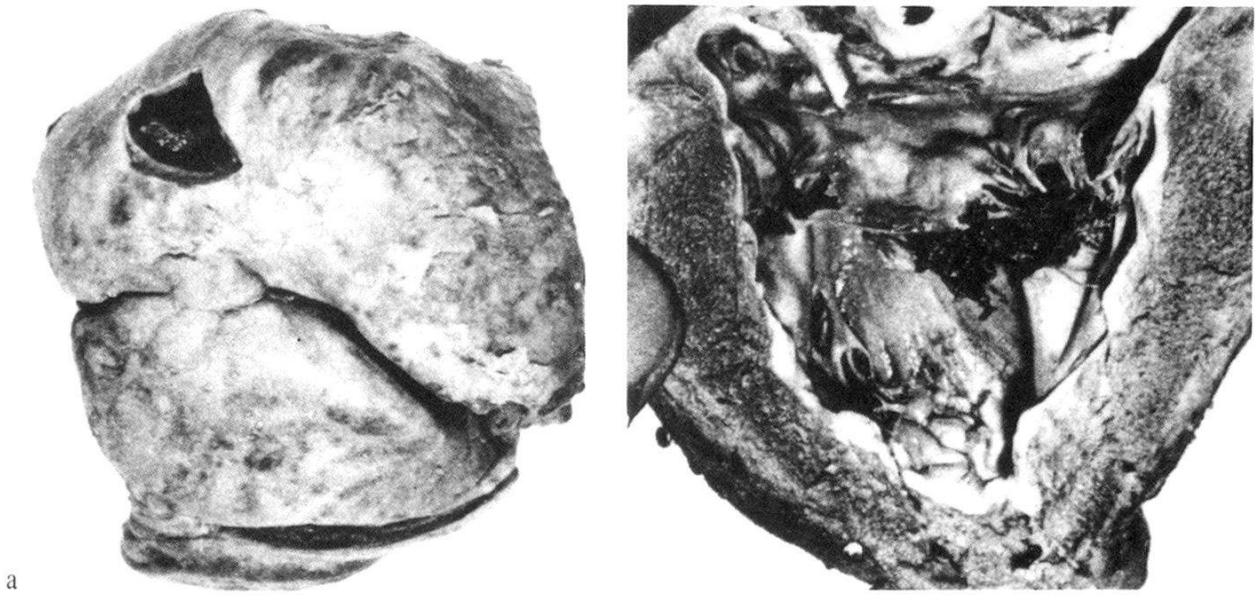
The child did well as outpatient on digoxin therapy alone and started school in 1978. Although the quadruple rhythm persisted, and the liver gradually enlarged from 8 cm to 12 cm below the right costal margin with systolic pulsations, and the spleen enlarged to 4 cm below the left costal margin, he had no ascites and his exercise tolerance was limited only when exertion was more than moderate. On February 4th, 1979 a limited right heart catheterization was attempted via the right antecubital vein, using a number 7 cournand catheter. The catheter got coiled-up in a grossly dilated right atrium and it proved impossible to advance the catheter to the right ventricle or beyond. Right-atrial «a» wave was 10 mm Hg, and «V» wave 12.5 mm Hg; the «y» trough was deep but the «x» descent was shallow.

On 27/8/1979 he developed fever and facial puffiness. Abdominal swelling and vague abdominal pain occurred the following day and he was admitted again on August 29, 1979. His face was markedly puffy especially periorbitally and moderate degree of ascites was present. The liver and spleen had not enlarged further; and the cardiac and pulmonic findings did not differ clinically or radiologically from previous findings. Temperature was 100° F, and pulse 108/min. Other laboratory findings were as shown in Table 1.

Facial puffiness and ascites disappeared on frusemide and digoxin therapy. But the abdominal pain became worse and colicky and did not respond well to analgesics. The patient died suddenly on September 14th, 1979 while on admission.

Results of autopsy

A limited autopsy was permitted. The heart and a small portion of the liver were removed. The heart weighed 197 g, it was moderately enlarged. There was a moderately severe grade of fibrinous pericarditis. The epicardial coronary arteries were normal. The right atrium and its appendage was dilated (Fig. 3a) and almost completely filled by an antemortem thrombus (Fig. 3b). The tricuspid valve ring was dilated but the leaflets appeared normal, the chordae tendinae were slightly thickened and shortened. There was fibrosis of the endocardium of the inflow tract and the apex which involved the papillary muscles and extended to the outflow track of the right ventricle (Fig. 3b). The pulmonary



a b

c

Fig. 3. Autopsy results: a) the whole heart with fibrinous pericarditis and a grossly dilated right atrium; b) the ante mortem thrombus in the right atrium, the fibrosis of the right ventricle which extended to the out flow tract; c) fibrosis of the inflow tract and the apex of the left ventricle.

valves were normal. The myocardium of the right ventricle which was about 4 mm thick, had small patches of fibrosis.

The left atrium and its appendage appeared relatively normal. The mitral valve leaflets and the chordae tendinae were normal. There was fibrosis of the endocardium of the inflow tract and the apex of the left ventricle, but its myocardium which was about 12 mm thick appeared relatively normal (Fig. 3c).

Histopathology

There was ante mortem thrombus superimposed on a thin layer of dense hyalinised avascular tissue in the right atrium. The myocardium of the right atrium showed changes similar to those of the ventricles but the left atrium appeared relatively normal.

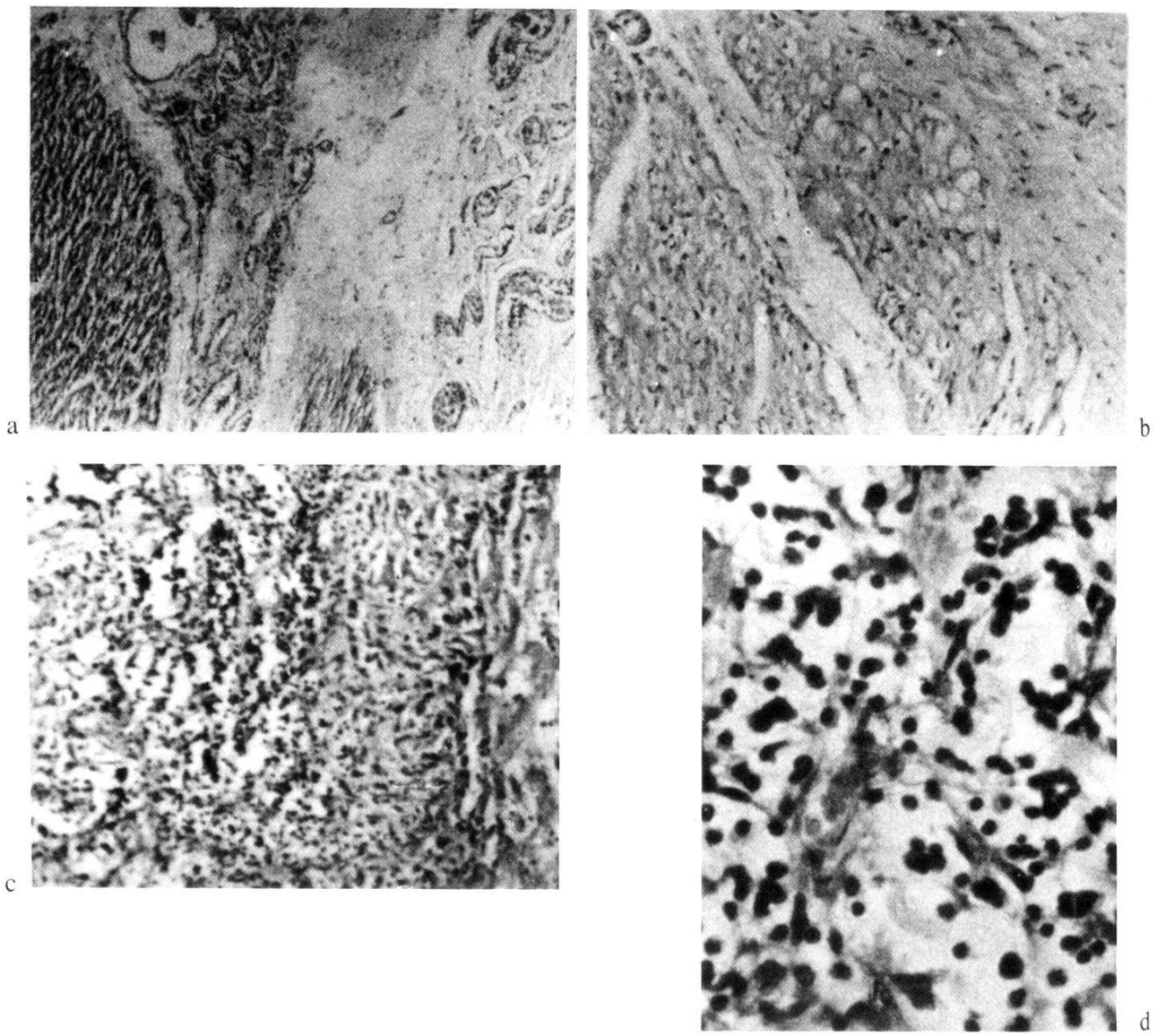


Fig. 4. Histopathology showing pericardial inflammation and endomyocardial fibrosis: a) the layer of endocardial and endomyocardial granulation tissue with dilated blood vessels and chronic inflammatory cells, × 100; b) myocardial fibrosis, vacuolation and myocytolysis, × 100; c) there was a moderately heavy degree of chronic inflammatory reaction in the pericardium, × 100; and d) most of the inflammatory cells were mononuclear and eosinophils, × 400.

The luminal surface of the right ventricle had a thick layer of dense hyalinised vascular fibrous tissue. A layer of granulation tissue containing loose collagen fibres, dilated blood vessels and chronic inflammatory cells was situated between the fibrous endocardium and the myocardium (Fig. 4a). The papillary muscles and the trabeculae carnae of the right ventricle were embedded in a vascular fibrous tissue which extended in tongues into the inner third of the myocardium. Similar but less pronounced changes were seen in the left ventricle but its outflow tract only showed moderate fibro-elastosis. The muscle fibres varied in size and many showed vacuolation (Fig. 4b). The nuclei also varied in size and some appeared bizzare while some others were vacuolated. There was interstitial oedema and patchy areas of fibrosis. Areas of myocytolysis were also

seen. These myocardial changes were more prominent in the subendocardial and subpericardial zones of both ventricles. The pericardium was replaced by granulation tissue with loose collagen fibres, dilated blood vessels and a moderately heavy infiltrate of mononuclear cells and eosinophils (Fig. 4c and d).

Histopathology of the liver showed severe central congestion leading to central fibrosis and cardiac cirrhosis.

Discussion

This patient is unique among African patients with E.m.f. in that his birth and childhood illnesses were well documented as well as the onset of his cardiac symptoms, and the progression of this cardiac disease. The itchy periorbital swelling and urticarial type rash suggested microfilariasis. These together with the markedly elevated eosinophil count led to a thorough search for microfilariae which were identified as microfilariae Loa-loa using the Knott's concentration technique of blood examination. Diethylcarbamazine therapy restored the eosinophilia to normal and cured the loasis.

But despite cure of loasis and normalization of eosinophilia, the heart disease had persisted and gradually progressed to a fatal termination. Twenty-five Europeans have been reported with E.m.f. while resident in Africa but white count differentials were available in 22, and eosinophilia was marked in 19 of these (Brockington et al., 1967; Gardner-Thorpe et al., 1971). Also microfilariae were found in the blood of 15 of these 25 patients. Experience from these European patients suggests that once heart disease had occurred, reversal of eosinophilia to normal spontaneously (Brockington et al., 1967; Baltzenschlager et al., 1961) with cortisone (Gerbaux et al., 1957; Giraud et al., 1959) or with antiparasitic agents (Gerbaux et al., 1957; Gardner-Thorpe et al., 1971) did not appear to significantly affect the course of heart disease.

A major difficulty with the filarial-eosinophilic theory of causation for E.m.f. has been the fact that eosinophilia and microfilariasis are not encountered in the majority of African patients with E.m.f. (Patel et al., 1977). However, these latter patients are usually seen with well established E.m.f. disease. The clinical features of established E.m.f. which consist of a) features of cardiac constriction (gross ascites with minimal leg oedema, distended neck veins with proptosis and globular cardiomegaly with relatively oligaemic lungs on X-ray), and b) evidence of atrio-ventricular valvular incompetence (Abrahams, 1962; WHO, 1965) are easily recognizable. To our knowledge African patients seen early in the course of E.m.f. have not been previously documented.

Apart from recognizable features of heart failure that our patient had presented, there were other features not usually found in acute myocarditis. The absence of symptoms of orthopnea or nocturnal dyspnea suggested a pure right-sided failure and/or a restrictive heart disease. Findings on clinical examination, electrocardiography and chest radiography were also suggestive of a

predominantly right-sided failure. A systolic retraction in the second left intercostal space which he presented is not usually present in myocarditis, but is described in some cases of E.m.f. (Shaper, 1974). Facial and periorbital puffiness are usually not encountered in myocarditis.

The fact that clinical features of microfilariasis preceded onset of cardiac symptoms by a few days suggests a possible causal relationship. However, the mechanism by which microfilariasis and eosinophilia are associated with E.m.f. is not known. The short history of filarial infection which preceded the cardiac symptoms make an immunologic damage unlikely. But allergic clinical features like periorbital swelling, skin rash and itching in this patient appear to lend support to the conclusions reached by Connors et al. (1968) after detailed histopathologic studies, that E.m.f. is an allergic heart disease.

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