The CENTRAL AFRICAN JOURNAL OF MEDICINE

Vol. 3. No. 9. SEPTEMBER, 1957.

CONTENTS

PUBLISHED MONTHLY, ANNUAL SUBSCRIPTION £2 2s. 0d.
Registered at the General Post Office as a Newspaper.

Symmetrical Gangrene Occurring in a Female African Affected with Endomyocardial Fibrosis

BY

MICHAEL GELFAND, O.B.E., M.D., F.R.C.P.
Physician, Salisbury African Hospital.

Endomyocardial fibrosis was first described in Africans in 1946 by Bedford and Konstam, who recognised it in West African soldiers, and two years later Davies gave a fuller account of it from Uganda. Since then several important and confirmatory papers from Uganda, the Union and elsewhere in Africa have appeared.

We now possess a fairly good conception of its pathology and clinical course. Briefly, it is characterised by an endomyocardial fibrosis of varying extent, especially marked at the apex of the left ventricle. Almost always a thrombus is seen on the damaged endocardium or a large one forms in the appendage of one or other of the auricular appendages. The fibrosis may be patchy or diffuse, involving the whole of the ventricle. The layer may be several millimetres thick, extending on to the epicardium, and in rare instances may be so pronounced that an obliterative effect from the fibrosis is produced, reducing considerably the capacity of the right ventricle (Ball, Williams and Davies, 1954). The site of election is in the apex of the left ventricle.

The papillary muscles are also commonly involved and the chordae tendiniae thickened and shortened. When the cusps become thickened and attached to adjacent structures, mitral and tricuspid incompetence are common accompaniments.

The clinical picture is usually that of congestive heart failure. Systolic murmurs due to mitral or tricuspid incompetence are common. The heart is generally enlarged, and on fluoroscopy movement may be markedly diminished, often due to an overlying pericardial effusion. Histologically there is a superficial endocardial layer of hyaline avascular fibrous tissue and a deeper layer with dilated blood vessels. The muscle fibres in many parts of the heart show varying degrees of degeneration.

In my experience embolic phenomena appear to be extremely rare in endomyocardial fibrosis, although from our knowledge of the pathological effects one would expect it to be seen more commonly. Williams *et al.* (1954) have not observed embolism in their series reported from Uganda. Although Becker, Chatgidakis and Van Lingen (1953) mention that embolic phenomena arise from cardiovascular collagenosis as a result of the mural thrombus, they give little indication of its frequency. Other writers (Smith and Furth, 1943; Gray, 1951), describing endocardial fibrosis with mural thrombosis, have commented on the frequency of embolic incidents in their patients in the absence of any infection.

The present case which is being reported is of further interest in that symmetrical gangrene of the lower extremities occurred. The causes of symmetrical gangrene in the African remain at present unknown (Gelfand, 1948). There are probably a number of different causes. Many of these cases follow a clinical pattern. It is seen in Africans of either sex, but mostly in the male between 20 and 60 years of age. The extent of the gangrene varies—it may be confined to the toes or the foot itself, but occasionally reaches above the ankle. Usually it is remarkably symmetrical in extent on both sides, but this is not always so, and I have seen cases where it was unequal. Usually too the gangrene starts in both feet at the same time. Symmetrical gangrene has been reported from different parts of Africa and causes such as syphilitic endarteritis, tropical angiitis or even malnutrition have been mentioned. It has been seen, for instance, among prisoners of war in Japan. Another suggestion made has been ergot poisoning, but there is no proof of this. I have seen a few cases follow meningococcal septicaemia and meningitis. The present case is of special interest in that symmetrical gangrene followed embolisation in a subject suffering from subendomyocardial fibrosis.

CASE ILLUSTRATION

Mary was an adult Native female aged about 32 years who was admitted on 12th May, 1954, to the Native Hospital, Salisbury. She complained of pain in both feet and had noticed six weeks before admission that at first the left foot and shortly after the right foot suddenly became numb and cold. There was no pain at the time of onset, but later she began to experience severe pain in both feet. Some three weeks later she started to cough and her sputum was blood-stained. At the same time she became increasingly short of breath.

On examination she looked ill, appeared to have lost a little weight, but her skin was blackish and of fairly healthy colour. She had

a small thyroid adenoma, but there was no evidence of thyrotoxicosis. On inspection she was breathless in bed and the heart enlarged. The apex beat was felt 4½ inches from the mid-sternum in the fifth intercostal space on the left side. A triple rhythm and a soft blowing systolic murmur was audible at the mitral area. The heart sounds could be heard fairly clearly. Numerous rales were present at the bases of the lungs and there was also dullness with impairment of the air entry in the left upper zone. The jugular veins were distended. The liver was enlarged, smooth, tender and reached four fingers below the right costal margin. A moderate ascites was present. The blood pressure was 120/90 (left arm). The left radial and brachial pulses could not be felt on either side. The left femoral pulse was absent, but the right one was just palpable. Pitting oedema of the sacrum was well marked, but oedema in the legs and ankles was slight. The pulse rate varied between 100 and 120 and the rhythm was regular. The fundi were clear. The decholin arm-tongue time was 17 seconds.

On X-ray the heart showed generalised enlargement, the C.T.R. being 58 per cent. A dense opacity about the size of a half-crown was present in the apex of the left lung towards its periphery. Mottling was seen in both middle and lower zones. The sputum did not contain *M. tuberculosis*. The total white cell count was 18,200 per cmm., the differential count being neutrophils, 50 per cent.; lymphocytes, 45 per cent.; monocytes, 4 per cent.; and eosinophils 1 per cent. The haemoglobin was 78 per cent. (Newcomer) and the blood Wassermann reaction was negative. The E.C.G. showed absent T waves in the standard and unipolar chest leads, a left axis shift and multiple ventricular extra-systoles.

Dry gangrene was present in both extremities, the left foot being affected up to the level of the ankle joint, where there was a sharp line of demarcation; in the right limb it was confined to all the toes.

The urine did not contain sugar or albumin and the stool on microscopical examination was clear of ova.

The patient was put on a salt-free diet, limited fluid intake, digoxin, 0.25 mg. twice daily by mouth, and mersalyl 2 c.c. twice a week by intramuscular injection. The diet was essentially a high protein one and included casilan. Daily injections of 1 c.c. of vitamin B complex were also given.

There was failure to improve, and as the pain in the feet increased we were obliged to administer injections of morphine and pethidine.

On the 28th May it became obvious that the patient would not survive. We had come to the conclusion that she was probably a case of endomyocardial fibrosis with occlusion of the arteries in the legs by emboli.

AUTOPSY

The deceased, aged about 30 years, was fairly well covered, with no signs of malnutrition. The left foot was black and shrivelled to the level just below the ankle. All the toes of the right foot were blackened, dry and shrivelled. The lower limbs above the line of gangrene were slightly oedematous. There was no clubbing of the fingers and no icterus. On opening the chest wall a small amount of straw-coloured fluid was present in each pleural space as well as in the pericardial sac (about half a pint). In the left upper lobe towards the periphery was a large whitish hard infarct about 2 inches square. The rest of the lungs were congested.

The liver was enlarged, its surface being smooth, and on cross-section the organ presented a typical nutmeg appearance. The spleen was moderately enlarged.

The Heart.—The heart was not enlarged, the left ventricle thickness being 1 cm., the right ventricle 0.25 cm. The valves were unaffected. No abnormality was detected in the aorta and the coronary arteries.

On inspecting the endocardium of the left ventricle four small whitish patches about a quarter of an inch square were seen at different sites. In the middle of one of the papillary muscles a white opaque patch a quarter of an inch long was noted. At the apex of the left ventricle, lying between the chordae tendiniae, was a hard whitish yellow thrombus about 2 cm. in diameter. It was irregular, coarse and could not be readily separated from the endocardium.

On cutting into the myocardium a more or less uniform layer of whitish tissue, varying at different levels from ¼ to ½ cm. in depth and extending from the endocardial surface into the myocardium, was found. The remainder of the myocardium appeared to be normal. The right ventricle and both auricles appeared healthy. The large thrombus as well as about 1½ inches square of the left ventricle were submitted to the South African Institute for Medical Research for histological examination. In the external

iliac artery was a long firm adherent thrombus occluding the lumen of the vessel. It was firmly adherent to the artery and could not be separated.

Histology (report from South African Institute for Medical Research).—A section was taken which included the thrombus, the underlying endocardium, myocardium and epicardium. The epicardium was congested and oedematous and in some areas it was infiltrated by a few lymphocytes. A branch of the coronary artery in the section showed no abnormality. In the myocardium were areas of interstitial fibrosis most marked in the inner and middle thirds of the wall. In these areas many of the fibres were degenerated and a few were hypertrophied. Scattered throughout the myocardium were small foci of lymphocytic infiltration, and in the inner third numerous dilated vascular and lymphatic channels.

The endocardium was normal in appearance in some areas. The normal endocardium changed abruptly to the grossly abnormal, where it was markedly thickened and took the form of polypoid projections into the lumen of the heart. In these areas the connective tissue showed the presence of mucinous oedema and stained metachromatically with toluidine blue. There were foci of fibrous necrosis, evidence of recent and past haemorrhage and small foci of lymphocytic infiltration. In one such area there was a definite increase in elastic tissue. The endothelial cells covering these affected areas were swollen, stained basophilically and many had pyknotic nuclei. Attached to one of the affected areas of the endocardium was a thrombus which was already partly organised.

The tissue shows the characteristic histological features of "cardiac collagenosis," previously known as Loeffler's parietal endocarditis.

CONCLUSION AND SUMMARY

The case was of interest, as the patient's main complaint was that of symmetrical gangrene, and it was only on physical examination that it was found that the patient was suffering from congestive cardiac failure. The next step was to discover the cause of the heart failure, and by a process of elimination, and after having observed the patient for some time, we arrived at the conclusion that she probably had subendocardial fibrosis with embolisation. This was confirmed at autopsy.

Embolisation may occasionally occur in endomyocardial fibrosis in Africa; and further, this form of heart disease may occasionally account for the symmetrical gangrene seen in the extremities of the African.

REFERENCES

BECKER, B. J. P., CHATZIDAKIS, C. B. & VAN LINGEN, G. (1953). *Circulation*, 7, 345.
BEDFORD, D. & KONSTAM, G. L. (1946). *Brit. Heart J.*, 8, 236.
GELFAND, M. (1948). *Brit. med. J.*, 2, 847.
GRAY, I. R. (1951). *Brit. Heart J.*, 13, 387.
SMITH, J. J. & FURTH, J. (1943). *Arch. int. Med.*, 71, 602.
WILLIAMS, A. W., BALL, J. D. & DAVIES, J. N. P. (1954). *Trans. Roy. Soc. trop. Med. & Hyg.*, 48, 290.

Acknowledgment

I wish to thank Dr. D. M. Blair, O.B.E., for permission to publish this paper.