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Aortic Embolism in Endomyocardial Fibrosis (EMF)

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Summary. Aortic embolism is seen in ten out of 236 cases with endomyocardial fibrosis examined at autopsy and it was not seen in any other form of heart disease. In every case, there was EMF in the left side of the heart, and the emboli arose from the left ventricle in nine cases and from the mitral valve in one case. Most of those who presented clinically with acute severe abdominal pain had aortic embolism at the level of the coeliac and superior mesenteric arteries, whereas those who presented mainly with lower limb symptoms had embolism at the bifurcation of the aorta.

Résumé. L'embolisme aortique comme complication mortelle de la fibrose endomyocardique fut observé dans dix autopsies sur 236 cas analysés. Il est à noter que pendant la même période cette complication ne fut pas trouvée parmi d'autres cardiopathies. Dans chaque cas il y avait une fibrose endomyocardique du ventricule gauche compliquée par une thrombose murale donnant origine l'embolisme, exception faite d'un seul cas où il existait une thrombase valvulaire.

La symptomatologie clinique était celle d'ischémie se présentant comme abdomen aique lors de l'obstruction des artères mésentériques ou coeliaques, ou alors comme ischémie des membres inférieurs en cas d'obstruction au niveau de la bifurcation aortique.

Embolic occlusion of the aorta is a recognized complication of intracardiac thrombosis, especially in rheumatic heart disease and myocardial infarction (Rothstein, 1935). It was at first thought that systemic embolism in endomyocardial fibrosis (EMF) was rare, but Shaper, Hutt & Coles (1968) found evidence of embolic phenomena in 16% of subjects in a post-mortem review of Ugandan cases. They also referred to seven cases of aortic embolism in cases with EMF one of which had been described by Connor *et al.* (1967 & 1968). In Nigeria, Brockington & Edington (1972) mentioned four cases of endomyocardial fibrosis with massive systemic embolism, causing cerebral infarction in one and occluding the abdominal aorta in three. In this paper, ten cases of aortic embolism associated with EMF are reported.

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MATERIAL AND METHODS

The autospy records of Mulago Hospital, Kampala, Uganda between 1956 and 1971 were examined and all cases of aortic embolism were recorded. Four of these cases were examined at autopsy by the author. In each case, the clinical and autopsy notes were studied. The histological sections were obtained from the records and re-examined.

RESULTS

During the period under review, there were 236 autopsy cases of EMF and ten (4%) of these had aortic embolism. Aortic embolism was not seen in rheumatic heart disease, or in cases who had rheumatic heart disease associated with EMF. There were seven females and three males with an age range of 14–56 years. The main clinical and pathological features are shown in Table 1 and two illustrative case summaries are provided below:

Case 1

A female aged 56 years was admitted to hospital with swelling and early gangrene of the right foot for 2 weeks. There were no pulses in the right leg. A diagnosis of saddle embolus at the level of the bifurcation of the aorta was made and embolectomy was performed. A week after the operation, thrombosis occurred at the same site and another operation was done, but the patient died 2 days later.

At autopsy examination, there was gangrene of the right foot. The heart weighed 290 g and at the apex of the left ventricle, there was endomyocardial fibrosis, with superimposed recent thrombus. There was no other lesion in the heart. The aorta and common iliac arteries were free of atherosclerosis and there was no further thrombosis in the aorta at the site of the operation.

Case 2

A 25-year-old male was referred from a health centre with the history of acute abdominal pain and distension for 24 hr. He had also vomited three times. On physical examination, the heart was normal. The abdomen was distended, tympanitic and there were increased bowel sounds. A provisional diagnosis of volvulus was made, but the patient died before laparotomy was done.

Autopsy examination revealed 100 cm^3 of straw coloured pericardial fluid and bilateral apical EMF. The heart weighed 400 g. In the left ventricle, the apical EMF extended along the inflow tract for about 4 cm. The posterior cusp of the mitral valve was also involved by EMF, causing incompetence. The left ventricle was hypertrophied. At the apex of the left ventricle there was a friable greenish-yellow thrombus (Fig. 1).

A large thrombus 5 cm long and 3 cm in diameter was located at the level of the openings of the coeliac and superior mesenteric arteries. Small projections of the embolus entered into these vessels. There was no atherosclerosis of the aorta (Fig. 2). The liver showed chronic venous congestion and the lungs were oedematous and congested with evidence of haemosiderin in histocytes, and pulmonary atherosclerosis suggesting that there had been long-standing heart failure.

Clinically, aortic embolism was diagnosed in only one case who had an embolectomy performed, but the rest were incidental autopsy findings. The clinical symptoms suggesting

TABLE 1.

Sex	Age (years)	Symptoms due to aortic embolus	Interval between onset of symptoms and death	Site of embolus
M	14	Abdominal pain, paraplagia	12 hr	From coeliac axis to bifurcation of the aorta
F	25	Gangrene of left foot	6 days	At bifurcation
M	32	Nil	—	At bifurcation, embolus, 3 cm long
F	30	Abdominal pain and vomiting, muscular pain in legs	24 hr	From coeliac axis to just below renal arteries
F	56	Swelling and gangrene of right foot	2 weeks	At bifuraction
F	14	Pain and weakness of legs, incontinence of urine	8 days	At bifurcation
F	40	Abdominal pain and distension	24 hr	At coeliac axis
F	30	Abdominal pain	6 hr	From coeliac axis to just above renal arteries
F	16	Abdominal pain	31 days	At bifurcation
M	25	Abdominal pain and distension	24 hr	From coeliac axis to just above the renal arteries. Embolus 5 cm long

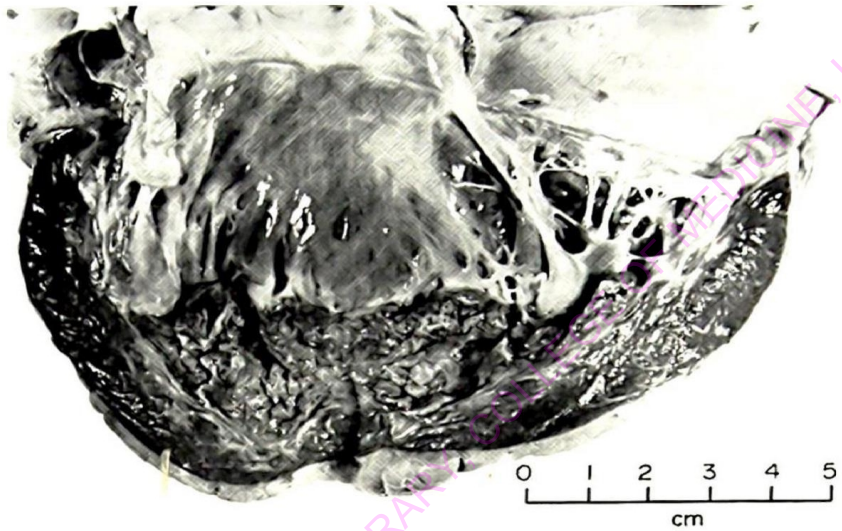

FIG. 1. Left ventricular EMF with superimposed thrombosis.

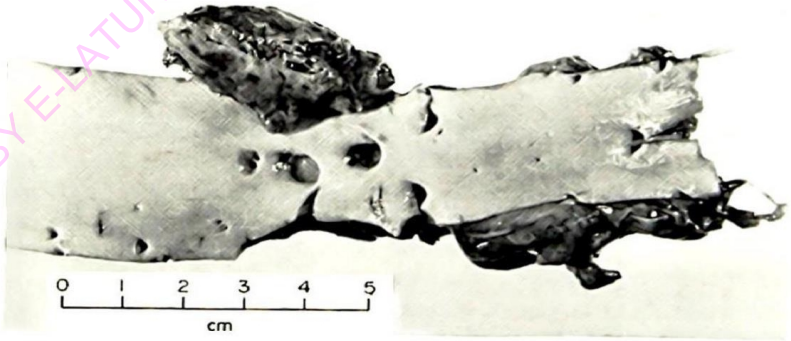

FIG. 2. Aortic embolism at the level of coeliac and superior mesenteric arteries.

aortic embolism are shown in the table. It should be appreciated that diagnosis of EMF itself may be difficult to make clinically; and many cases are admitted in terminal states or are discovered at routine autopsy examination. In this study, a clinical diagnosis of EMF had been made in two cases. Acute severe abdominal pain, present in six cases, was the commonest major cause of hospital admission. Two of these cases had abdominal pain and distension and one of these two was clinically diagnosed as a volvulus. Two other cases were diagnosed as ectopic pregnancy and bacillary dysentery. The cause of abdominal pain in the remaining two cases could not be determined clinically. In five of the six cases, the patients died within 24 hr and the thrombus was found to be lodged at the level of the coeliac and superior mesenteric arteries. Five cases had symptoms in the lower limbs consisting of muscular pain, weakness, paraplegia or gangrene. Those patients who had lower limb symptoms associated with abdominal pain died within 24 h and were found to have an embolus lodged at the level of coeliac artery; those who only had symptoms in the lower limbs, such as gangrene, died after several days and were found to have the embolus at the bifurcation of the aorta. In seven of the ten cases, the onset of the symptoms suggesting aortic embolism was sudden and the patients were said to be previously well. However, on microscopic examination of lungs and livers, two of these cases had evidence of long standing heart failure. The remaining three of the ten cases were in congestive heart failure when the acute symptoms occurred; clinically, two had been diagnosed as EMF and one as hypertensive heart failure.

The site of EMF and sources of emboli

In nine cases, the source of emboli was the left ventricle and in one case, who had EMF with superimposed infective endocarditis of the mitral valve, the embolus came from the mitral valve. None had an aneurysm or atherosclerosis of the aorta which could be the source of emboli. In all cases, EMF involved the left side of the heart; involvement of the left ventricle alone was seen in four cases; left and right ventricle two cases; left ventricle and the posterior cusp of the mitral valve, two cases; left and right ventricles and posterior cusp of mitral valve, one case and this case had superimposed infective endocarditis on the mitral valve. None had thrombosis in the left atrium or appendage even the case who had atrial fibrillation. In seven of the ten cases, the thrombus in the left ventricle was described as massive and recent; and in three cases the colour was greenish-yellow.

Emboli elsewhere

Systemic embolism was observed in seven cases. The spleen was affected in five cases, the kidney in three cases and the brain in one case. One of the two cases with thrombosis in the right ventricle had small pulmonary emboli. In six of the seven cases of systemic embolism, the resulting infarcts were older than would be expected if the systemic small embolism occurred at the same time as the massive aortic embolism. The small systemic emboli therefore preceeded the massive aortic emboli.

DISCUSSION

Intracardiac thrombosis in endomyocardial fibrosis is seen in about 43% of cases at autopsy and in most of these the thrombi occur in the left ventricle and right atrium and very

rarely in the left atrium. This contrasts with the findings in rheumatic heart disease where intracardiac thrombosis occurs in about 16% of cases and the thrombi are usually found in the right and left atria. The incidence of emboli however, remains about the same in the two diseases (Shaper *et al.*, 1968). Systemic emboli in endomyocardial fibrosis therefore arises mainly from the left ventricle and in rheumatic heart disease they arise mainly from the left atrium. This finding correlates closely with the results of the present study in which nine cases had thrombosis in the left ventricle and not left atrium as is usually the case in rheumatic heart disease. The higher frequency of aortic embolism in EMF is probably related to the site of origin of the thrombus which partly determines its size and shape. In EMF, very large thrombi form in the left ventricle and it is these that are likely to be arrested in the aorta. The site of the aortic embolism depends on the size of the embolus and in the rheumatic heart disease, the size is probably determined by the degree of mitral stenosis, since most come from the left atrium. However, in endomyocardial fibrosis, thrombi come from the left ventricle and they would be expected to be large and to block the aorta high up, but that is not always so in this study, because in five cases the embolism was about the level of the coeliac and superior mesenteric arteries.

The formation of embolus may also depend on the type and size of thrombus in the left ventricle. In EMF, the thrombi in the left ventricle which give rise to the aortic emboli are usually massive and recent. It is not clear whether the thrombus in the left ventricle forms itself into a 'ball' and then the embolization occurs; or it detaches itself as a flat sheet and then it rolls up as it passes through the aorta. If the former is true, then a round embolus would be expected, but this is not the case and in most cases the embolus was described as sausage-shaped or long. This suggests that the embolus is formed from a detached flat thrombus. It is suggested that when thrombosis occurs in the left ventricle, some organization may take place, but also small emboli may be formed, giving rise to small systemic embolism and infarction. At a later stage, a massive thrombus is formed and is detached as a large embolus which blocks the aorta and produces the acute severe symptoms. In none of these cases did the aorta show atherosclerosis and so it is extremely unlikely that the thrombi seen in the aorta were the result of local thrombosis. Superimposed thrombosis in the aorta at the site of the embolus was observed in one case who lived for about 1 month after the onset of the symptoms.

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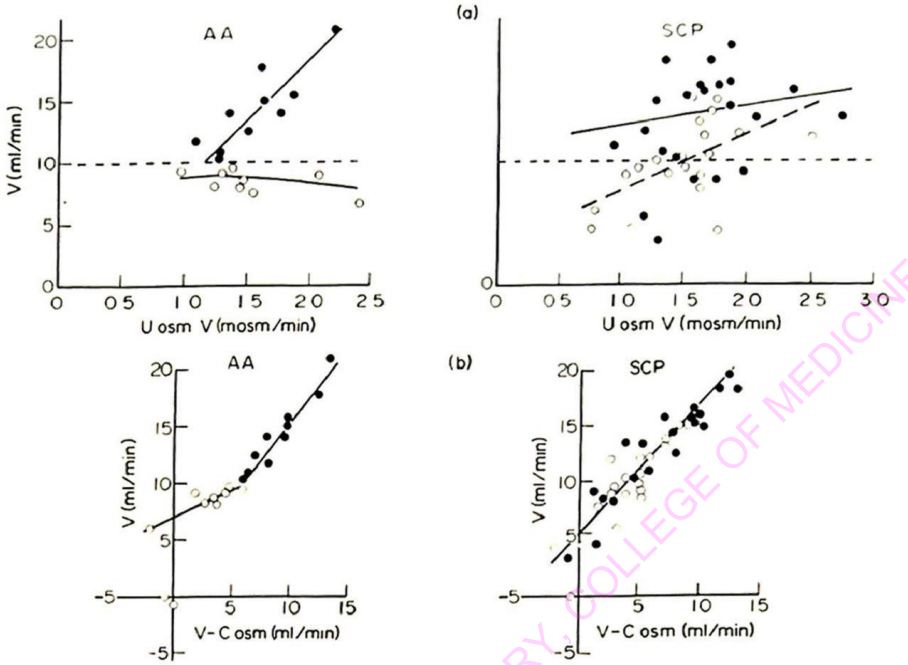

FIG. 2. (a) Composite relationship between U osm. V, and V in the AA at 20° and 40°. Absence of such relationship in the SCP (SS+SC). 20° and 40° values could not be segregated in the SCP (See also Fig. 1). Open circles, 40°C; closed circles, 20°C. Regression equations: AA at 20°C; $y = 0{\cdot}068x + 3{\cdot}0$. $r = +0{\cdot}70$. SCP at 20°C+40°C; $y = 0{\cdot}04x + 4{\cdot}94$. $r = +0{\cdot}34$. SCP at 20° only; $y = 2{\cdot}42x + 11{\cdot}37$. $r = +0{\cdot}24$. SCP at 40° only; $y = 4{\cdot}4x + 3{\cdot}02$. $r = +0{\cdot}55$. SCP. Solid line, regression line for all SCP at 20°C; broken line, regression line for 40°C.

(b) Relationship between V and V-C osm. Open circles, values at 40°C; closed circles, values at 20°C. Regression equations as follows: AA at 20°C; $y = 1{\cdot}29x + 2{\cdot}9$. r = +0·96. AA at 40°C; $y = 0{\cdot}39x + 7{\cdot}3$. $r = +0{\cdot}86$.

Note: No differentiation between 20° and 40° values possible (see also Fig. 1) in the SCP Equation for pooled 20° and 40° data: $y = 1{\cdot}01x + 5{\cdot}8$. $r = +0{\cdot}91$.

DISCUSSION

Because a brisk combined water and osmotic diuresis was produced and maintained throughout the study, V is used here as a rough estimation of the rate of delivery of filtrate to the distal nephron and V–C osm to approximate sodium reabsorption in the ascending limb of the loop of Henle. In Fig. 2b, the lower slope at 40° compared with that at 20° in the AA subjects suggests a greater rate of sodium reabsorption per unit volume fluid delivery at this site at 40° than at 20°C. This, it is suggested, is probably secondary to the higher titre of circulating vasopression. The enhanced vasopression secretion in the heat has been clearly demonstrated by Segar & Moore (1968). Whether indeed vasopressin enhances sodium transport in the nephron is debatable (Berliner & Bennett, 1967).

It is suggested that the curve for the AA at 40°C represents largely juxtamedullary nephron activity. This interpretation is based on the following considerations: (1) Radigan & Robinson (1949) and also Kenney (1952, 1963) have shown that the intrarenal sympathetic activity increases in the heat. Since increased sympathetic tonus has been held to be as-