negative and other Gram-positive organisms have been incriminated in the development of the same clinical picture.[6]

Post-infection glomerulonephritis occurs most commonly in children between the ages of 3 and 7 years and with advancing age the number of cases decreases.[7,8] However, patients over 50 years comprise 5 - 10% of cases in some series, men being affected more frequently than women.[8,9] Its relative rarity in the adult may delay the diagnosis, as was the case in our patient whose clinical picture and history initially obscured the presence of underlying renal disease.

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Recurrent myocardial infarctions secondary to luetic coronary arteritis in hypertrophic cardiomyopathy

A case report

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Summary

A 43-year-old coloured man had no risk factors for atheromatous coronary artery disease but suffered two acute myocardial infarctions (MIs) in rapid succession. Serological reactions for previous syphilitic (luetic) infection were positive. Hypertrophic cardiomyopathy (HCM) without obstruction was verified, although right ventricular endomyocardial biopsy specimens did not demonstrate histological features of this disease. Extensive MI was verified on left ventricular cine angiography. Selective coronary arteriography showed that the coronary arterial tree was diffusely aneurysmal in the absence of any obstruction.

We postulate that syphilitic coronary arteritis, in the absence of the more pathognomonic coronary ostial stenotic lesions, was present and may have predisposed to coronary thrombus formation and repeated acute MI. Recurrent coronary vasospasm, associated with the HCM, cannot be excluded with certainty.

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Case report

A 43-year-old coloured man, married with 6 children, had smoked 3 cigarettes a day for several years but had no other specific risk factors for atheromatous ischaemic heart disease (IHD). On 27 December 1983 he experienced severe, crushing, retrosternal discomfort while pushing his father in a wheelchair, associated with marked sweating, dyspnoea and palpitations. The chest pain continued for some 2 hours before his arrival at the Intensive Coronary Care Unit (ICCU) at Tygerberg Hospital, where he was still in some pain and sweating.

The patient was very muscular and tattooed. He had a rapid and regular radial pulse, all peripheral pulses being easily palpable. The jugular venous pressure was not elevated and the blood pressure was 130/90 mmHg. There was no clinical cardiomegaly but the left ventricular (LV) apex was obviously very forceful, considering his large acute myocardial infarction (MI). A loud fourth heart sound was heard, and a grade 2/6 pansystolic murmur characteristic of mitral insufficiency and probably due to papillary muscle dysfunction was prominent. There were no diastolic murmurs and no pericardial friction rub or features of cardiac

failure. The fundi appeared normal and the remainder of the clinical examination revealed no further abnormalities.

The urine was normal; there was a leucocytosis of 15 400/l with a 94% neutrophilia, an erythrocyte sedimentation rate (ESR) of 100 mm/1st h and normal haemoglobin and platelet levels. Serum biochemical findings were unremarkable. The chest radiograph was normal. The ECG taken on admission displayed a sinus tachycardia of 108 beats/min, P-R interval of 0,15 second, mean QRS axis of 28° and features of a hyperacute transmural anteroseptal, apical and anterolateral MI. A striking feature on the ECG was the absence of any reciprocal changes in the inferior leads. Treatment was instituted with a heparin infusion, transdermal nitroglycerin and a small dose of β-blocker.

Approximately 24 hours after his admission to the ICCU the patient had a further episode of severe central chest pain, and an ECG now demonstrated the features of a hyperacute nontransmural inferior wall MI in addition to the evolution pattern of the previous extensive anterior MI. Nifedipine was now added to his therapeutic regimen, with no further recurrence of precordial pain. Daily ECGs signified the presence of an acute transmural anterior and nontransmural inferior MI. Serial cardiac enzyme estimations supported the ECG diagnosis; maximum values (in U/l with the normal range in parentheses) were as follows: creatine kinase 1 155 (0 - 50), aspartate transaminase 438 (0 - 40), alanine transaminase 76 (0 - 53), and lactate dehydrogenase 1 770 (100 - 350). On 30 December 1983, some 72 hours after his admission to the ICCU, a technetium-99m pyrophosphate scintiscan demonstrated a large circular region of increased isotope uptake in the anterior part of the myocardium with slight extension to the inferior segment. The patient's further course in the ICCU was uncomplicated. A resting technetium scintiscan (gated blood pool scan) on 4 January 1984 delineated a generalized hypokinesia of the LV with more significant abnormality of the inferior, apical and septal segments. The LV ejection fraction was 42%.

Further investigations were carried out in view of the improbability that the patient's two sequential MIs were due to atheromatous coronary artery disease (CAD). Tests for collagen vascular disease were all negative, but immunological investigation for luetic infection by the *Treponema pallidum* haemagglutination, rapid plasma reagin, VDRL (titre 1 : 8) and fluorescent treponemal antibody absorption tests was positive on repeated testing. The ESR also remained high (over 100 mm/1st h) but the white cell count returned to normal. The patient was given a course of intramuscular penicillin. A repeat resting technetium scintiscan on 13 January 1984 delineated marked dyskinesia of the apical and inferior segments, in keeping with an LV aneurysm. The interventricular septum (IVS) demonstrated better contractility than before (4 January 1984) and the LV ejection fraction rose to 50%.

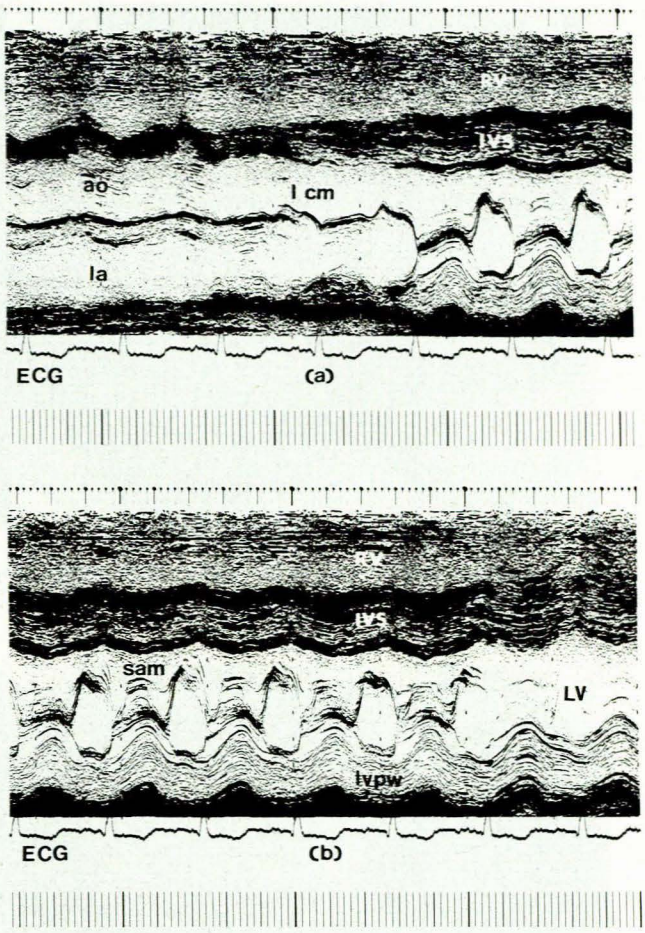

Fig. 1. M-mode echocardiograms: (a) asymmetrical septal hypertrophy is present, there is no left atrial enlargement; (b) marked systolic anterior motion (SAM) of the anterior leaflet of the mitral valve is demonstrated. Asymmetrical septal hypertrophy is again seen (ao = aorta; la = left atrium; RV = right ventricle; lvpw = left ventricular posterior wall).

Echocardiography (M-mode and two-dimensional)

M-mode echocardiography was carried out with the patient supine and the transducer over the third intercostal space to the left of the sternum and directed so that the ultrasound beam passed across the LV from apex to base. The IVS and LV posterior wall (LVPW) were measured below the tips of the mitral valve leaflets before atrial systole and after rapid ventricular filling. The IVS/LVPW ratio was increased, at 1,4 (normal < 1,3), due to asymmetrical septal hypertrophy (Fig. 1(a)). The most striking feature observed was that of systolic anterior motion of the anterior leaflet of the mitral valve (Fig. 1(b)). The left atrium was not dilated and the LV internal dimensions were within the normal range. An ejection fraction of 52% was surprisingly high in view of the large MI. The IVS was slightly hypokinetic, while the LVPW was hyperkinetic.

Two-dimensional (cross-sectional) echocardiography with the beam directed so as to obtain a long-axis (saggital plane) view, as well as a short-axis two-chamber view, showed marked hypokinesia of the free anterior wall and apex of the LV with some slight hypokinesia of the IVS. Normal contractility of the inferior segment was noted. The echocardiographic features were in keeping with hypertrophic cardiomyopathy (HCM) complicated by abnormal LV segmental wall motion secondary to MI.

Cardiac catheterization

Cardiac catheterization was undertaken on 16 January 1984. Right-sided intracardiac pressures were all normal. Catheterization of the left side of the heart documented an LV pressure of 155/0 - 18 mmHg, central aortic pressure of 155/105 (mean 129) mmHg and a pulmonary capillary wedge pressure of 18 mmHg (equal 'a' and 'v' waves). No left-sided intraventricular (subaortic) pressure gradient could be detected at rest or after the Valsalva manoeuvre. The dp/dt_{max} was 2 625 mm/s, a remarkable finding in view of the MIs. Further haemodynamic parameters of LV function were: cardiac output 7,8 l/min; cardiac index 4,1 l/min/m^2; systemic vascular resistance 16 units; pulmonary vascular resistance 0,9 unit. Thus, again, LV function was exceptionally good despite the MIs. LV cine angiography in the right anterior oblique (RAO) (Fig. 2) and left anterior oblique (LAO) projections (Fig. 3) delineated marked dyskinesia of the free anterior wall, apical and IVS regions. The inferior segment of the LV appeared to contract normally. Systolic anterior motion of the anterior mitral leaflet was obvious, verifying the previous echocardiographic finding. Aortic cine angiography in the LAO view demonstrated a non-calcified valve with three leaflets and no aortic insufficiency. The thoracic aortic arch appeared normal. Selective coronary angiography showed that the right coronary artery (RCA) was dominant and diffusely ectatic (almost aneurysmal), but with no evidence of ostial stenosis or calcification (Fig. 4). Right-to-right collateralization was absent. Left coronary artery (LCA) cine angiography (Fig. 5) delineated a normal coronary ostium with aneurysmal

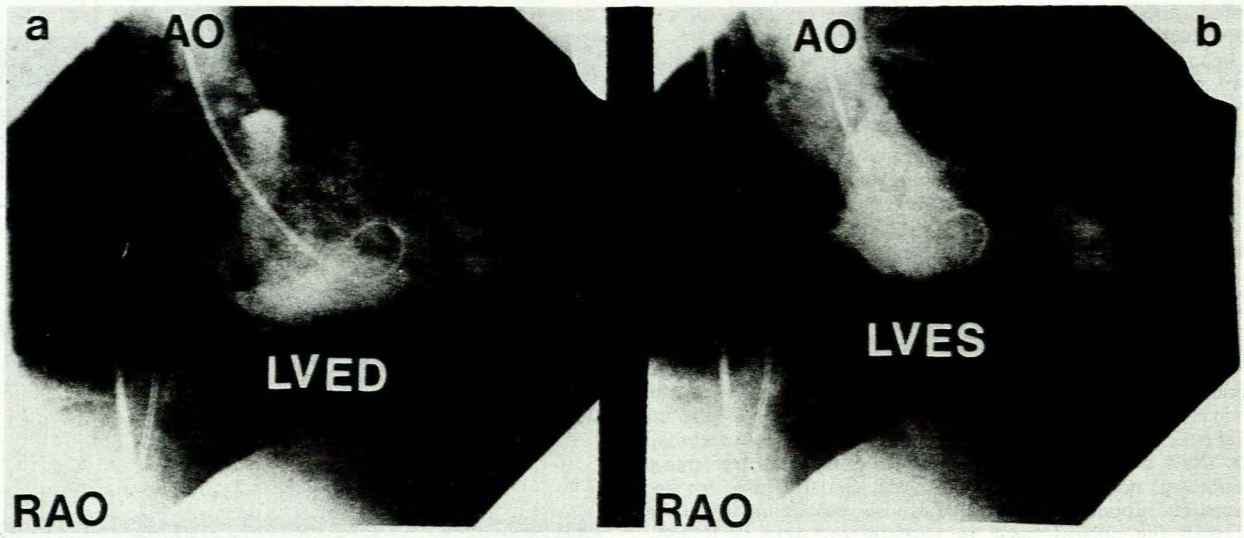

Fig. 2. LV cine angiograms in the right anterior oblique (RAO) projection. Dyskinesia of the antero-apical segment is seen. The inferior segment appears to contract quite normally and there is no mitral insufficiency. a — LVED = LV in end-diastole; b — LVES = LV in end-systole (AO = aorta).

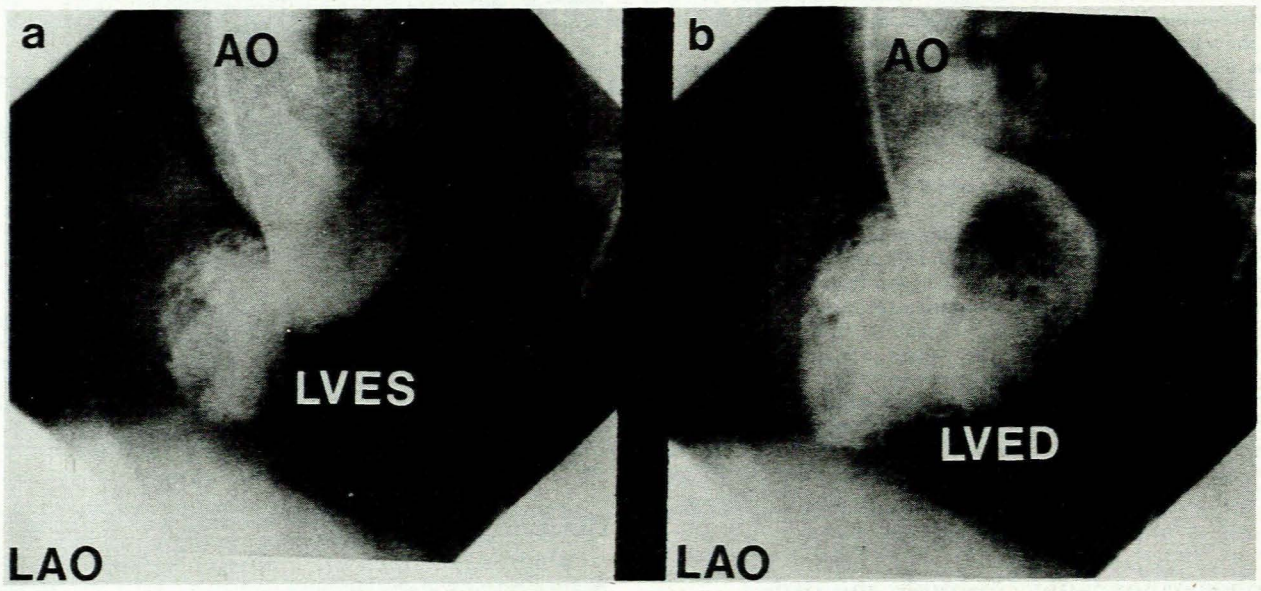

Fig. 3. LV cine angiograms in the LAO view. The IVS appears dyskinetic. a — LVES = LV in end-systole; b — LVED = LV in end-diastole (AO = aorta).

involvement (similar to that of the RCA) of the left mainstem, proximal left anterior descending (LAD) and left circumflex branches. The first diagonal branch of the LAD contained several small filling defects which appeared to be resolving thrombi. No atherosclerotic lesions or coronary calcification could be visualized. In view of previous investigations suggesting HCM, multiple right ventricular (RV) endomyocardial biopsies from various sites were taken. Light microscopy demonstrated that the myocardium was somewhat hypertrophied but otherwise unremarkable. Electron microscopy revealed hypertrophied myocytes with mitochondriosis and lipofuscin pigmentation (Fig. 6). There was no significant myofibrillar disarray. Thus, biopsy could not confirm the diagnosis of HCM demonstrated clinically, echocardiographically and by cine angiography.

Discussion

Of several interesting aspects concerning this patient the most important are the coronary artery cine angiographic findings of ectasia or aneurysmal dilatation; the possible role of syphilitic involvement of the coronary arteries; the pathogenesis of the two acute MIs in rapid succession; and the influence, if any, of the HCM without obstruction.

The serological investigations established the presence of previous syphilitic infection. Cardiac involvement by syphilis is perhaps best known because of the aortitis which gives rise to aortic arch aneurysms, aortic valve incompetence, and coronary ostial stenosis.[1-4] Involvement of the coronary arteries distal to the ostia, 'coronary arteritis', is controversial, although there does not appear to be any patho-anatomical reason for the exemption of the more distal segments of the coronary arteries from the disease. Moritz[5] has documented syphilitic coronary arteritis up to 12 mm distal to the coronary ostia. Warthin[6] has stated that 'the larger branches of the coronaries only rarely show lesions that can be recognized as syphilitic',

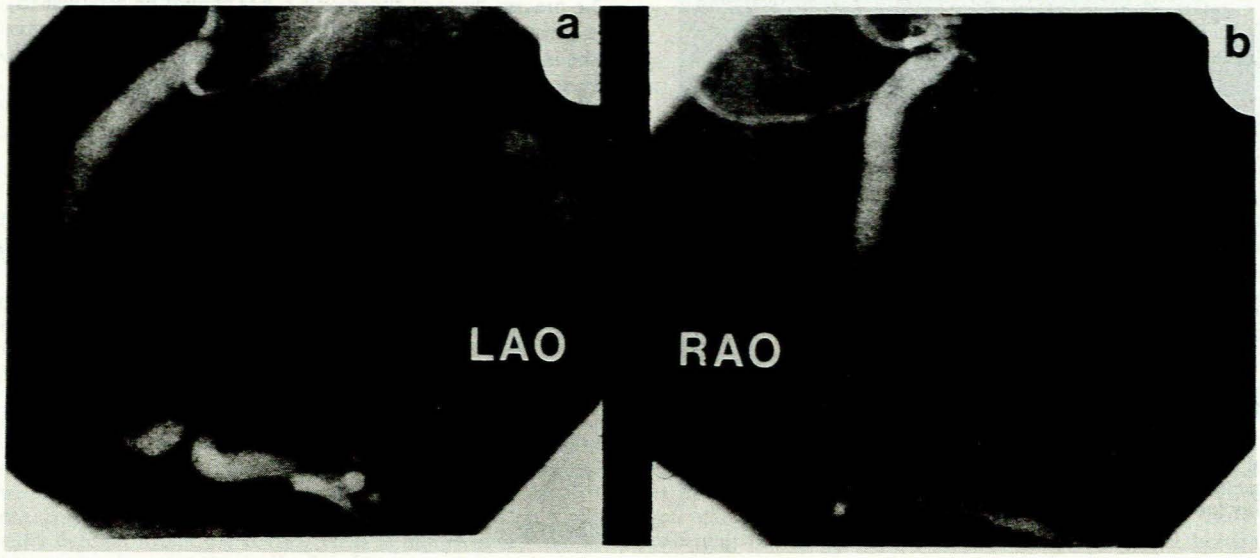

Fig. 4. Right coronary cine angiograms in the (a) LAO and (b) RAO projections. The vessel is dominant and diffusely ectatic but there is no ostial stenosis. No right-to-left collateral vessels can be seen.

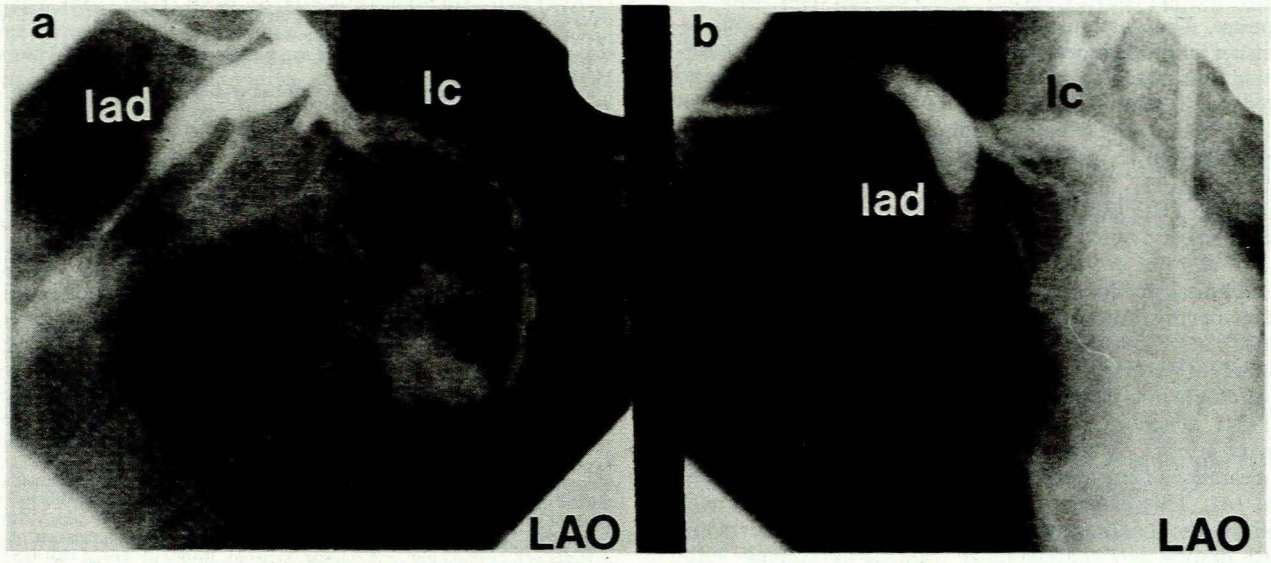

Fig. 5. Left coronary cine angiograms in the (a) LAO view and (b) shallow LAO projection with cranial angulation. The left mainstem, LAD and left circumflex (lc) coronary arteries are aneurysmal in the proximal portions. Some left-to-left collateral flow is present. There is no ostial stenosis.

while Maher[7] reported pathological changes in the distal parts of the epicardial coronary arteries similar to those found in the ascending aorta. Bruenn,[1] in an autopsy series of 148 cases, claimed that 'syphilis of the coronary arteries distal to their orifices is an uncommon lesion and is rarely of clinical importance'. Denham[8] reported on a thrombus in a syphilitic aneurysm of the LAD coronary artery in an 81-year-old Negro man which caused a massive acute MI. Autopsy revealed markedly tortuous coronary arteries but no atherosclerosis. The cine angiographic findings in our patient were grossly abnormal in that the RCA and proximal portions of the LCA were diffusely ectatic or aneurysmal. There was no evidence of atherosclerotic coronary obstruction and resolving thrombi were residua of the cause of the acute MIs. We believe that these angiographic features were due to a diffuse syphilitic coronary arteritis, which is essentially an obliterative endarteritis of the vasa vasorum supplying the medial layer of the coronary arteries. Ectatic or aneurysmal coronary arteries are not known to occur in HCM and are probably most common in coronary atherosclerosis.[9]

The pathophysiology of this patient's two acute MIs is intriguing. Swaye *et al.*,[9] in discussing aneurysmal CAD, state that 'the presence of dilated coronary segments, even in the absence of obstructive disease, is believed to result in alterations in blood flow and stasis, which predispose these patients to myocardial ischemia and infarction'. Our patient may have suffered a similar fate. Another explanation is recurrent coronary artery vasospasm. Mautner *et al.*[10] documented coronary artery spasm in 3 patients with hypertrophic obstructive cardiomyopathy (HOCM), one of whom had underlying coronary atherosclerosis. These authors[10] state that 'if the mechanism of hypertrophic cardiomyopathy is increased myo-

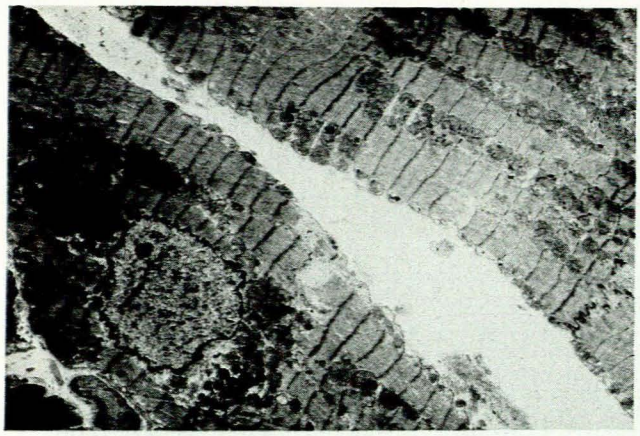

Fig. 6. Section of myocardium showing mitochondriosis and lipofuscin pigment. There is no evidence of any myofibrillar disarray (uranyl acetate x 7 500).

cardial responsiveness to catecholamines, it is possible that the coronary arteries of these patients may behave in a similar fashion and be prone to vasospasm'.

The coexistence of atherosclerotic CAD and HOCM may be impossible to appreciate clinically since both can cause identical symptoms. This combination was first reported by Gulotta *et al.*[11] using selective coronary angiography. Walston and Behar[12] reported an incidence of 15% of significant obstructive coronary atherosclerosis in HOCM in a series of 42 patients and emphasized that the ECG can be misleading since pseudo-infarction patterns are well known in HOCM.[13] Maron *et al.*[14] reported 'silent' acute MI in patients with HOCM and associated insignificant IHD and raised the likelihood of coronary vasospasm. They also noted a marked haemodynamic deterioration with the onset of acute MI in that cardiac dilatation ensued with a clinical profile almost identical to dilated (congestive) cardiomyopathy. Maron *et al.*[14] suggested that 'small-vessel disease'[15] may have contributed to the acute MI. It is not commonly appreciated that the clinical, haemodynamic and echocardiographic features of HCOM can disappear with an acute MI which may occur with angiographically normal coronary arteries.[6] Thus, Come and Riley[17] documented the disappearance of intraventricular gradients with acute transmural MI. This experience may well be applicable to our patient in whom no intraventricular gradient could be demonstrated despite echocardiographic features in keeping with HOCM.

The precise cause of this man's CAD cannot be established, but we strongly contend that diffuse syphilitic coronary arteritis is to be incriminated and that superimposed recurrent coronary artery spasm associated with HCM cannot be entirely ignored in explaining the pathophysiology of the MIs.

We wish sincerely to thank Professor J. J. van der Walt of the Department of Anatomical Pathology, Tygerberg Hospital and the University of Stellenbosch, for his assistance in the interpretation of the RV endomyocardial biopsy material. Sincere thanks are also extended to Mr Christopher Wilberforce, formerly Head of the Photographic Unit, Bureau for Medical and Dental Education, University of Stellenbosch, for preparing the photographs. We also thank Miss H. W. Weymar of the Cardiac Unit for preparing the ECG illustration. Finally, due appreciation is shown towards Dr J. P. van der Westhuyzen, Chief Medical Superintendent of Tygerberg Hospital, for permission to publish.

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