IDIOPATHIC CARDIOMYOPATHY SIMULATING ORGANIC VALVE DISEASE

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Cardiomyopathy of unknown origin is extremely common in the African population of Johannesburg.[1-4] As in the cardiomyopathies in general,[5] the heart valves are normal and the principal auscultatory findings are gallop rhythm and functional systolic murmurs caused by dilatation of the mitral and tricuspid rings. Diastolic murmurs are generally absent.

The object of this paper is to describe 2 cases of idiopathic cardiomyopathy which presented with both diastolic and systolic murmurs of functional origin.

These cases simulated organic disease of the mitral and aortic valves, owing to the persistence of the murmurs and the peripheral circulatory findings.

CASE REPORTS

Case 1

E.S., an African male aged 13 years, was admitted to hospital in July 1959, complaining of swelling of the lower limbs and a dry cough of 2 weeks' duration. He had been having bouts of palpitations associated with dizziness for 'a long time' and, on questioning, admitted to breathlessness on exertion and episodes of paroxysmal nocturnal dyspnoea. There was no history of rheumatic fever.

On examination the patient was orthopnoeic and in severe congestive cardiac failure. He was apyrexial with a regular pulse rate of 100 per minute and a blood pressure of 110/60 mm.Hg. The pulse volume was poor and the extremities were cool. There was pronounced anterior bulging of the praecordium. The heart was markedly enlarged, with heaving impulses over both ventricles. A systolic thrill was palpable at the base and radiated into the neck. A systolic thrill was also felt at the apex. Auscultation at the apex revealed a protodiastolic gallop and a loud pansystolic murmur radiating into the axilla and around to the back. At the aortic and pulmonary areas there was a harsh mid-systolic murmur radiating into the neck. At the aortic area and down the left sternal border an early diastolic blowing murmur was audible. The pulmonary second sound was accentuated and closely split.

Radiography confirmed the gross cardiomegaly, with both left and right ventricular enlargement. The aortic knuckle was rather small and the pulmonary outflow tract was prominent. Posterior displacement of the barium-filled oesophagus and widening of the carina suggested left atrial enlargement. Electrocardiography showed left ventricular hypertrophy and strain (S in V_2 + R in V_6 = 50 mm.; T in V_6 inverted). The haemoglobin level was 13·9 G. per 100 ml., the ESR 5 mm. in the first hour (Wintrobe), and the leukocyte count and urine examination were normal.

The diagnosis made was that of congestive cardiac failure caused by aortic stenosis and incompetence and mitral incompetence, probably rheumatic in origin.

Following the administration of digitalis and chlorothiazide, the patient's condition improved slightly, but on the 4th day after admission he suddenly became markedly breathless. The pulse was rapid and irregular and an electrocardiogram showed numerous multifocal ventricular ectopic beats. Potassium chloride and procaine amide were given, but death occurred a few hours later. All the murmurs persisted throughout the illness.

At necropsy (Dr. A. Schmaman) the heart weighed 705 G. with gross hypertrophy and dilatation of all 4 chambers. The myocardium was 7 mm. thick at the right ventricular outflow tract and 13 mm. thick at the outflow tract of the left ventricle. The endocardium appeared normal, except for a small area of thickening measuring 1 × 2 cm. immediately below the aortic valve. The aortic and pulmonary valves were normal. The mitral and tricuspid valves easily admitted 3 and 4 fingers respectively, and the valve curtains showed no macroscopic lesion. The coronary arteries, aorta, venae cavae, and iliac vessels were normal. The pulmonary arteries were healthy and there was no evidence of pulmonary infarction.

Histological examination of the ventricular myocardium showed hypertrophy of the myocardial fibres with some increase in the interstitial fibrous tissue. In a few areas there was slight infiltration of the interstitial tissue by polymorphonuclear leukocytes and lymphocytes. The left atrial appendage showed antemortem thrombus adherent to the endocardium. Aschoff nodes were not found. All the valve cusps and the aorta were histologically normal.

The pathological features were those of idiopathic cardiomyopathy.

Case 2

P.G., an African male aged 28 years, was admitted in January 1959 with weakness of the right side of the body and inability to speak. This was of acute onset and he had previously been quite well.

On examination the patient had a right hemiplegia and aphasia. The temperature was 101°F. and the pulse rate was 96 per minute. The pulse was distinctly collapsing in character, with a blood pressure of 135/40 mm.Hg. The heart was markedly enlarged with heaving impulses over both left and right ventricles. At the apex a pansystolic blowing murmur and a mid-diastolic rumble were audible. At the aortic area there was a long blowing diastolic murmur, commencing immediately after the second heart sound. The murmur was also audible down the left sternal border. A short mid-systolic ejection murmur was heard at the aortic area.

Radiography confirmed the generalized gross cardiomegaly. The aortic knuckle appeared normal. The lung fields were congested. Electrocardiography showed left ventricular predominance (S in V_2 + R in V_6 = 43 mm.). The haemoglobin level was 15·7 G. per 100 ml. and the leukocyte count was 9,200 per c.mm. The ESR was 56 mm. in the first hour (Wintrobe), but C-reactive protein was absent. The antistreptolysin O titre was 1,000 units per ml. Blood cultures yielded no growth after prolonged incubation. Urinalysis was normal.

The diagnosis of aortic and mitral incompetence of rheumatic origin was made and, in view of the neurological signs suggesting a cerebral embolus and the pyrexia, the patient was treated for subacute bacterial endocarditis.

Improvement was rapid and within 1 week the power of speech and movement had largely returned. Seven weeks later, however, the patient suddenly developed congestive cardiac failure. The antistreptolysin O titre was now 1,600 units per ml., and the C-reactive protein was positive, ++. Response to digitalis and diuretics was satisfactory. Three weeks later, however, the congestive cardiac failure recurred and, after partial improvement, the patient's condition deteriorated rapidly and he died. The murmurs persisted throughout the 3-month course of the illness.

At necropsy (Dr. I. Spector) the heart weighed 507 G. There was marked hypertrophy of the left and right ventricular walls, which measured 18 mm. and 7 mm. in thickness respectively. The ventricles were also dilated. The mitral valve admitted the tips of 3 fingers and the tricuspid valve 4 fingers. The valve cusps were normal. The aortic and pulmonary valves, aorta, pulmonary artery, and coronary arteries were likewise normal. Numerous mural thrombi were present in both ventricles and in the right atrial appendage. A thrombus was present in the right femoral vein. The lungs were mildly oedematous and showed several areas of recent infarction. The brain showed an area of softening in the left internal capsule.

Histological sections of the myocardium showed hypertrophy of the muscle fibres. The endocardium was normal and no Aschoff nodes were found.

The pathological features were those of idiopathic cardiomyopathy with mural thrombi, right femoral-vein thrombosis, and cerebral and pulmonary infarction.

DISCUSSION

The 2 cases described were found at necropsy to be examples of the idiopathic cardiomyopathy commonly seen in the Johannesburg African population.[1-4] The heart valves and aortas were normal. Yet, clinically, they presented as cases of organic valvular disease. Case 1 had the murmurs and thrills of both aortic stenosis and mitral incompetence, the murmur of aortic incompetence, and a poor pulse volume, while case 2 had the murmurs of aortic and mitral incompetence and a collapsing pulse. The murmurs persisted throughout the clinical course, and in case 2 were clearly audible both in the presence and absence of heart failure. The diagnosis of idiopathic cardiomyopathy, a condition with which we are thoroughly familiar at Baragwanath Hospital, was not

even remotely suspected. In view of the normality of the valves at autopsy, all the signs of valvular disease must have been functional in origin and the question of their mechanism arises.

Functional mitral incompetence with an apical pansystolic murmur is common in the cardiomyopathies, and is generally attributed to dilatation of the mitral ring. Mitral diastolic rumbles have also been described,[6] although in our experience they are rare. They may be caused by the association of left ventricular dilatation with a normal mitral ring, producing, as it were, a relative mitral stenosis.[7] Alternatively they may arise when the cardiomyopathy is associated with functional incompetence of the mitral or aortic valves. In case 2, in which a mid-diastolic rumble was heard, both valves were functionally incompetent. In organic mitral incompetence a mid-diastolic rumble is common, and is thought to be caused mainly by the increased blood flow through the mitral valve during the phase of rapid ventricular filling.[8] The factor of turbulence created by the diseased valve cusps *per se* is regarded as being of secondary importance. Functional mitral incompetence must also be associated with an increased atrio-ventricular flow in mid-diastole, and it is conceivable that on occasion this flow may be so large that it gives rise to a mid-diastolic rumble despite the absence of diseased mitral cusps. In organic aortic incompetence, mid-diastolic or presystolic rumbles of functional origin may occur,[9] the presystolic murmurs being classically associated with the name of Austin Flint.[10] Since these murmurs are thought to be caused by interference with mitral-valve function by regurgitating blood,[9] there is no reason why they should not also occur in cases of functional aortic incompetence.

The aortic stenotic murmur of case 1 may be explained by the observations of Brock.[11,12] He was the first to describe the occurrence of functional obstruction at the outflow tract of the left ventricle secondary to extensive myocardial hypertrophy. He pointed out that the walls of the lower part of the outflow tract of the left ventricle are chiefly muscular and that, when sufficiently hypertrophied, they may come together in early systole and obstruct the further emptying of the ventricle. Brock[12] reported 4 proved cases of this condition. All presented with the clinical features of aortic stenosis and had been referred to him for aortic valvotomy. Left ventricular pressure tracings, obtained either at operation or by percutaneous catheterization, demonstrated an obstruction at the level of the outflow tract. At operation or necropsy no evidence of organic stenosis, valvular or subvalvular, was found. In all, the principal finding was marked myocardial hypertrophy. In all but one, the aetiology of the myocardial hypertrophy was entirely obscure, the exception being a case in which systemic hypertension was thought to be the cause. Following Brock's description, there have been a number of further reports of left ventricular outflow obstruction secondary to idiopathic myocardial hypertrophy.[13-15] Our first case was characterized by massive biventricular hypertrophy of unknown cause, and it is therefore possible that the aortic stenotic murmur heard in this patient was also caused by functional obstruction of the left ventricular outflow tract. The associated murmur of aortic incompetence might possibly be explained on the basis of post-stenotic dilatation involving the aortic ring, as is known to occur with subvalvular organic stenosis.[16]

The free, functional aortic incompetence, which was present in case 2, has also been previously described in patients with cardiomyopathy.[17,18] Anders,[17] in particular, reported a number of cases and reviewed the literature critically. He attributed the incompetence to left ventricular dilatation. The mechanism he suggested was based on the anatomical studies of MacCallum[19] who demonstrated a sphincter-like band of muscle surrounding the left ventricular outflow tract. Anders postulated that in left ventricular dilatation this sphincter relaxed and so gave rise to dilatation of the outflow tract and the aortic ring, with resultant aortic incompetence. In support of this contention, Anders quoted experiments in dogs in which section of the sphincter resulted in immediate aortic incompetence. It would, however, be generally agreed that functional aortic incompetence is rarely found in left ventricular dilatation of cardiomyopathic or other origin; this may be related to the fact that the upper part of the left ventricular outflow tract (the aortic vestibule) and its continuation, the aortic ring, are relatively rigid structures consisting of tough fibrous tissue. MacCallum's sphincter surrounds only the lower portion of the outflow tract and this, as stated above, is chiefly muscular, and therefore much less rigid in structure. Dilatation following relaxation of the sphincter would therefore be expected to involve mainly the lower part of the tract, with considerably less effect on the upper part.

SUMMARY

Two unusual cases of idiopathic cardiomyopathy are described. They were characterized by persistent murmurs, diastolic as well as systolic, which suggested organic disease of the aortic and mitral valves. All the valves, however, were found to be normal at necropsy. The mechanism of production of these murmurs is discussed.

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