AORTIC SUBVALVULAR STENOSIS FROM MYOCARDIAL HYPERTROPHY

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Obstruction to the left ventricular outflow tract occurs in aortic valvular disease and, in rare instances, is the result of a developmental defect (congenital aortic subvalvular stenosis) which forms an irregular raised ridge just proximal to the aortic valves. In the absence of these two conditions, gross muscular hypertrophy per se has recently been found to be another cause for functional obstruction of the left ventricle. Brock (1957) described such an instance in a patient who was operated on for apparent aortic stenosis but at operation the valves were found to be normal. Catheter withdrawal pressure readings demonstrated a stenosis about 2.5 cms. below the aortic valves. This, at autopsy, was found to be due to a grossly hypertrophied left ventricular wall bulging inwards below the aortic vestibule. The myocardial hypertrophy was the result of long standing hypertension (20 years) which in the last two years needed ganglion blocking agents for its control.

In 1960, Goodwin et al. described a series of eight patients in whom left ventricular hypertrophy of uncertain origin caused signs and symptoms simulating aortic stenosis. They used the term "obstructive cardiomyopathy" to designate this condition and stressed that obstruction can occur to outflow or inflow of either ventricle.

In the cases published by Brock (1957) and by Bercu et al. (1958) concentric hypertrophy was the cause of the obstruction. On the other hand in the patients of Teare (1958), Hollman et al. (1960), and Brent et al. (1960) asymmetrical hypertrophy of the ventricle was the lesion which caused obstruction at the mitral or tricuspid valves. Another feature was the presence of familial incidence pointing to an inherited disorder.

That muscular hypertrophy of the heart from varied causes, can simulate valvular disease is well illustrated in the abovenamed published works. An awareness of this is the importance.

Reported below are the findings of a patient who was thought to be suffering from mitral incompetence but autopsy revealed severe subaortic stenosis from a gross left ventricular hypertrophy. The aortic valves were normal.

CASE REPORT

L.A.P., a Chinese female aged 61 years was admitted on 11.3.61 into Thomson Road Hospital for investigation because of the possibility of subacute bacterial endocarditis.

On 20.1.61 she had been admitted into the General Hospital, Singapore for gangrene of the left foot for which a mid-thigh amputation was done on 1.2.61. On further enquiry she reported that she had had a low grade fever for about a month's duration prior to the onset of her gangrene. She denied having dyspnoea on exertion but admitted having had palpitations. She had been attending T.T.S.H. as an out-patient and was being observed for possible pulmonary tuberculosis. A year ago she was treated for glaucoma of the left eye.

On examination she was found to be thin, in a poor general condition with a temperature of 100°F, a pulse rate of 120 per minute. The amputation wound was healed.

There was no digital clubbing or cyanosis. The B.P. was 125/55. The apex beat of the heart was located in the fifth intercostal space 3" from the M.S.L. There was a systolic thrill, felt best in the left fourth intercostal space parasternally. Both heart sounds were heard at all areas. A systolic murmur was heard all over the praecordium but loudest just internal to the apex beat. The murmur was conducted to the left axilla. After further examinations of the heart, it was thought that an ejection click was heard over the fourth space on the left side. The left femoral pulse was not felt. The liver was felt one finger's breadth below the costal margin but the spleen was impalpable. The bed-side diagnosis was that of mitral incompetence and subacute bacterial endocarditis.

Investigations showed that she was anaemic with a Hb. of 58%, a raised B.S.R. of 105 mm. (first hour), but her W.B.C. count was within normal limits. 5 specimens of blood on culture were sterile. Results of other investigations done were :— urine : albumin +, R.B.C. 2-3, W.B.C. 3-5; sputum : negative for tubercle bacilli repeatedly; blood K.T. : negative; L.E. cell phenomenon : negative; serum proteins : 7.4 G, albumin 2.4 G globulin 5 G mainly from a raised alfa-2-globulin (1.2 g.).

The X-Ray film of the chest (Fig. 1) showed an enlarged heart (mainly of the left ventricle) with a c.t.r. of 14/24. There were healed tuber-

culous scars over both apices. The E.C.G. (Fig. 2) revealed the presence of left ventricular preponderance with strain (depression of ST segments in leads II, III, V5, V6, V7).

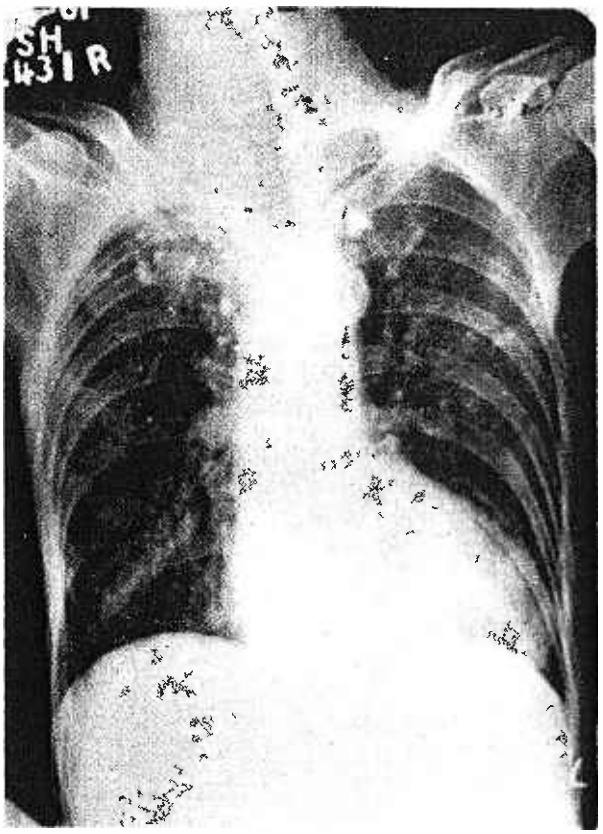

Fig. 1. The X-ray film of the chest showing the enlarged heart mainly of the left ventricle.

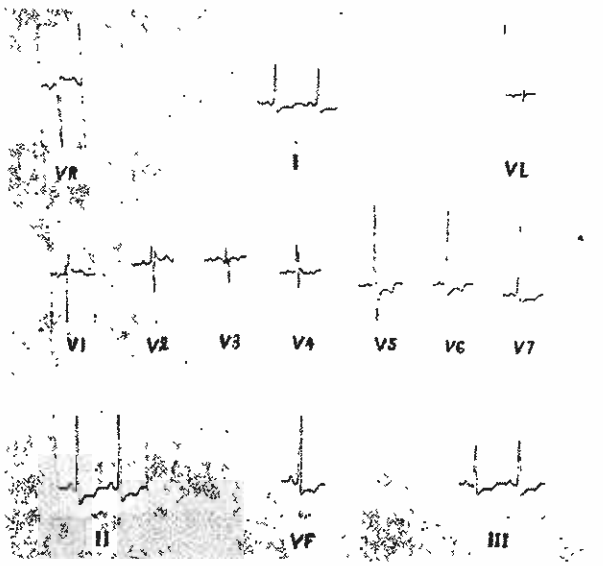

Fig. 2. The E.C.G. showing the presence of left ventricular preponderance and strain.

Throughout her stay in hospital she was having a low grade intermittent pyrexia of uncertain origin. Two weeks prior to her death on 28.4.61 she complained of bleeding per vaginum. Examination that day revealed bleeding through the os uteri which was bulky, fixed and retroverted. The cervix appeared normal. Carcinoma of the uterus was diagnosed but she was too ill to be submitted to surgery.

At POST-MORTEM the significant findings were mainly to be found in the cardiovascular system. The pericardial cavity contained a few mls. of clear serous fluid but the pericardium was within normal limits. The heart was enlarged, weighing 300 Gms. with a markedly hypertrophied left ventricle whose cavity was reduced to a mere slit (Fig. 3). The hypertrophy was most pronounced in a segment about 2.0 cms. long, beginning at a level about 1 cm. below the aortic orifice. This segment measured 2.5 cms. in thickness. Viewed through the aortic orifice, this muscle mass acting as the stenotic element was clearly demonstrable (Fig. 4). It almost completely obliterated the left ventricular cavity at this point except for a small slit-like opening. The right ventricle appeared normal, its wall measuring 5 mm. in thickness. The atria, the aortic, pulmonary and tricuspid valves were normal. The mitral valve cusps were normal

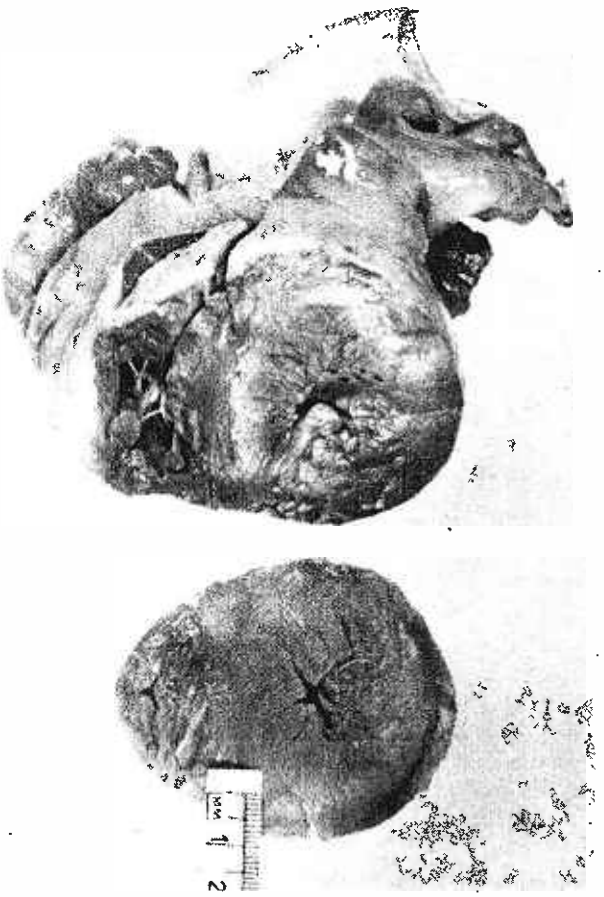

Fig. 3. The heart cut transversely showing the marked left ventricular hypertrophy. The ventricular cavity is reduced to a slit.

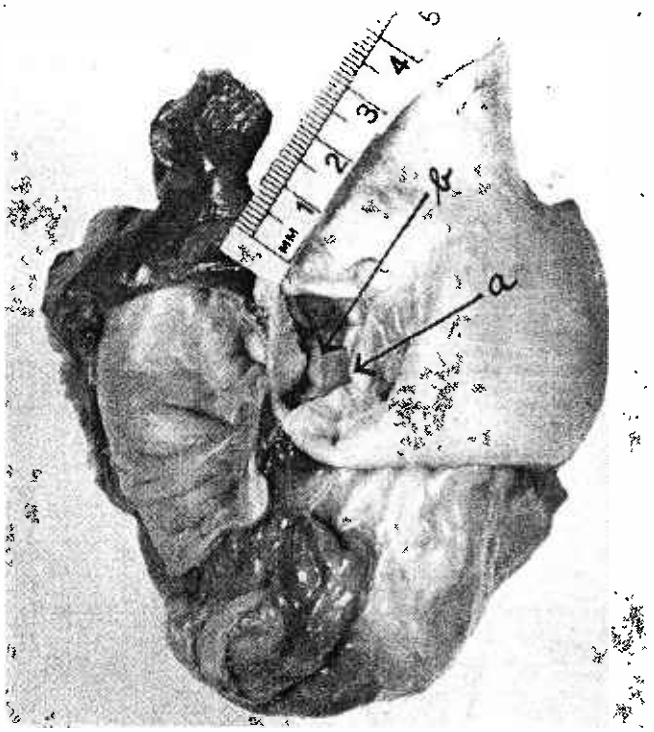

Fig. 4. The huge muscle mass seen through the aortic orifice from above (a) the valve cusps b) muscle mass.

also. The coronary vessels and their ostia were remarkably free from disease.

The aorta showed a severe degree of atherosclerosis. The left common iliac artery was occluded partly by atheroma.

The uterus was bulky weighing 210 Gms. It was fixed to the posterior pelvic wall. The surface was uneven and on sectioning yellowish-white tumour tissue almost replaced the whole uterine body. The cervix appeared normal.

HISTOLOGICAL EXAMINATION of the left ventricular myocardium showed a definite enlargement of the individual muscle fibres, with an average thickness of 30 u (Fig. 5). The nuclei

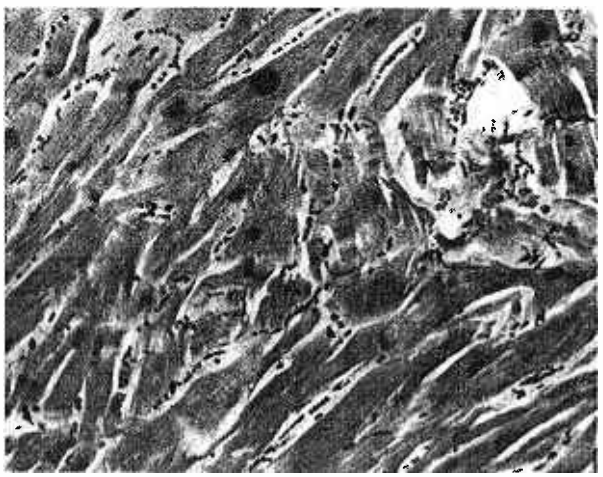

Fig. 5. Photomicrograph showing enlargement of the individual muscle fibres (H. & E. x 150).

of many fibres were increased in size, some assuming bizzare shapes. Throughout both ventricles there was the presence of a diffuse, interstitial fibrosis, especially dense in parts (Fig. 6). Special staining methods revealed the presence of both increased reticulin and collagen formation.

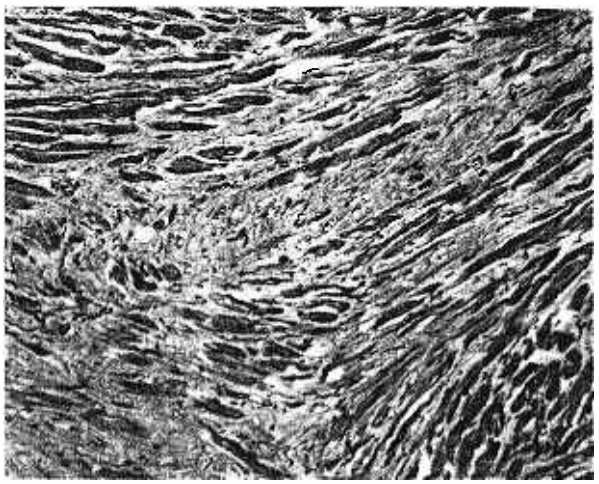

Fig. 6. Photomicrograph showing a patch of dense interstitial fibrosis (H. & E. x 75).

DISCUSSION

That obstruction to the left ventricular outflow tract can occur in the presence of normal aortic valves, from myocardial hypertrophy per se is well exemplified in this instance.

The cause for the myocardial hypertrophy was, however, obscure. There was no evidence of hypertensive disease both in her history and in the physical findings. Her cardiomegaly was not that associated with, or forming a part of, a generalised disorder like amyloidosis, haemochromatosis, glycogen storage disease (Goodwin et al. 1961) The histological findings here approximated closely to those published by Brent et al., Goodwin et al., and Hollman et al., all in 1960. In them too, no single significant cause for the hypertrophy could be found, beyond the familial incidence.

Diagnosis of this condition from aortic valvular stenosis may be difficult but is important. In as much as the latter is amenable to surgical correction, operation on patients in the former can be difficult as well as fatal, (Brock, 1957).

Clinically, an awareness of this condition may be aroused when a patient labelled as suffering from aortic stenosis has a normal or even quick-rising pulse. Further, the site of maximum intensity of the murmur may be internal to the apex beat or left parasternally. This murmur may

simulate closely that of a ventricular septal defect or even mitral incompetence as indeed was in the case herein reported.

Helpful investigations are a closer enquiry into other members of the family, angiocardiography and pressure gradient records from percutaneous ventricular puncture. Even then final diagnosis may not be possible until an operation is being done to obtain relief of the outflow tract obstruction.

A further importance of this condition becomes obvious when this concept of muscular obstruction to the left ventricle is accepted. There arises the possibility of this occurring in patients with enlarged hearts with hypertrophy from the commoner causes as in systemic hypertension of whatever origin.

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