ENDOCARDIAL FIBROELASTOSIS OF THE HEART

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The term endocardial fibroelastosis was first introduced by Weinberg and Himelfarb[1] in 1943. They described a cardiac condition of unknown aetiology in infancy in which there was diffuse endocardial sclerosis with enlargement of the left ventricle. Since then many cases have been reported and the subject has been well reviewed by Davis and Ball (1955)[2], Lynch and Watt (1957)[3] and Still (1961).[4]

Endocardial fibroelastosis is recognised clinically by two distinct patterns. In the first group the endocardial fibroelastosis is accompanied by other congenital anomalies common amongst which are mitral incompetence, aortic stenosis and aortic atresia, co-arctation of the aorta and patent ductus arteriosus. The second group constitutes the difficult clinical problems where the endocardial fibroelastosis is without other cardiac anomalies. The latter presents in infancy and childhood as obscure heart failure often with a marked gallop rhythm, a large left ventricle, cardiomegaly on chest X-ray and left ventricular hypertrophy and strain pattern in the electrocardiogram.

The pathological findings are the endocardial thickening most commonly involving the left ventricle and left atrium. The endocardium has an opaque, greyish white appearance. The valves may be thickened and sclerosed most commonly the mitral and aortic valves. The left ventricle is greatly hypertrophic, there may be mural thrombi on the affected endocardium which may give rise to emboli. Microscopically there is a marked increase of the elastic tissue of the endocardium. There is a sharp demarcation between the thickened endocardium and the underlying hypertrophied myocardium. There are no signs of inflammation or fibrosis in the myocardium. The purpose of this case report is to describe the clinical and pathological findings in a boy of 12 years presenting with the relatively uncommon combination of congenital aortic stenosis and endocardial fibroelastosis of the left ventricle. He had also hypospaedias and a horse-shoe kidney.

REPORT OF CASE

Clinical Features

Y.S.T., a 12 year old Chinese boy was admitted in General Hospital with fever, cough and dyspnoea for 3 days. He had pneumonia at the age of one year and a heart murmur was detected at the age of seven during a school health examination. At the age of ten he was admitted into the General Hospital for an operation for hypospaedias. Between that time and the last admission he was admitted into another hospital on three occasions with heart failure.

On examination the patient was found to be dyspnoeic and cyanosed. The temperature was 99·4° F. There was a praecordial bulge. The jugular venous pulse was raised to 5 cm. above the sternal angle. The pulse was 110/min., regular but small-volumed. The blood pressure was 80/70 mm. Hg. The heart was greatly enlarged and the apex beat was felt at the 6th left inter-space on the anterior axillary line and was forceful in character. There was also a marked left parasternal heave. A systolic thrill was felt over the aortic area where a grade 3 ejection systolic murmur was also heard. The murmur was conducted into the carotids in the neck. The aortic second sound was soft. There was no early diastolic murmur. Marked triple rhythm was heard over the apex and the 4th left sternal edge. There were signs of consolidation over the right lower lobe of the lung. The liver was palpable 2 fingers-breadth below the right costal margin. There was no peripheral oedema.

Investigations

Hb. 16·1 gm.%, W.B.C. 13,500/c. mm., Poly: 86%, Lymph: 12% Mono: 2%, Eos: 0%. The urine showed a trace of albumin. E.S.R. 9 mm./hour (Westergren). Blood Cultures—negative. A.S.O.T. 333 Todd units. Blood Urea 14 mg.%, Electrolytes—normal range. Plasma Protein: Albumin 4·7 Gm.%, Globulin 1·9 Gm.%, $alpha_1$ -Globulin 0·1 Gm.%, $alpha_2$-Globulin 0·5 Gm.%, beta-Globulin 0·5 Gm.%, gamma-Globulin 0·7 Gm.%. Chest X-ray (Fig. 1) showed a very large heart with bi-ventricular hypertrophy. Pulmonary Vasculature suggested marked pulmonary hypertension and some consolidation over the right lower zone. The electrocardiogram (Fig. 2) revealed marked left ventricular hypertrophy and was suggestive of some right ventricular hypertrophy as well. He was treated with digoxin, chlorothiazide, mersalyl, pot. chloride, penicillin and a low salt diet.

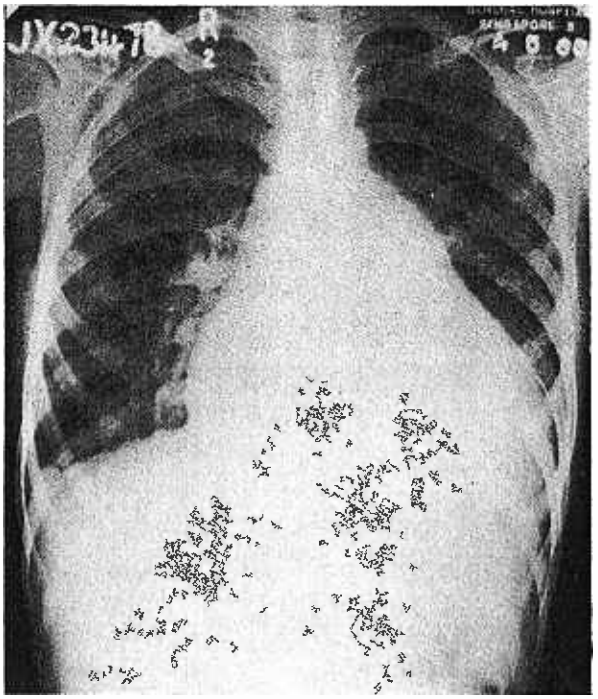

Fig. 1. Roentgenogram of thorax showing marked cardiac enlargement.

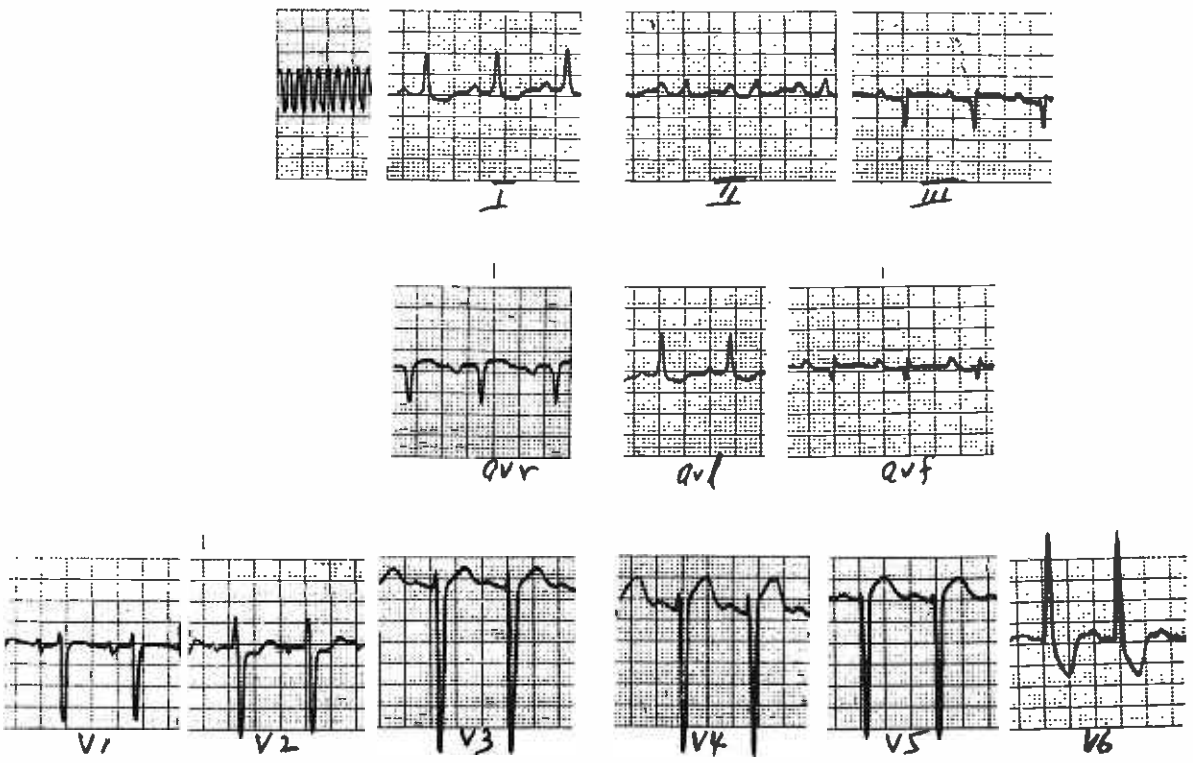

Fig. 2. Electrocardiogram showing marked left ventricular hypertrophy.

The patient was showing some response when he suddenly developed cardiac arrest and died.

Pathological Features

The body was that of a thin Chinese boy with hypospaedias. The heart was greatly enlarged, weighing 471 grams. The thickness of the right ventricle was 1·5 cm. The external positions of the great vessels were normal, the pulmonary artery was larger than the aorta having a diameter of 2 cm., whilst that of the aorta was 1·5 cm. (Fig. 3). The three cusps of the aortic valve were fused together forming a dome-shaped stenosis of the congenital type leaving a lumen

Fig. 3. Heart showing dome-shaped stenosis of the aortic valves.

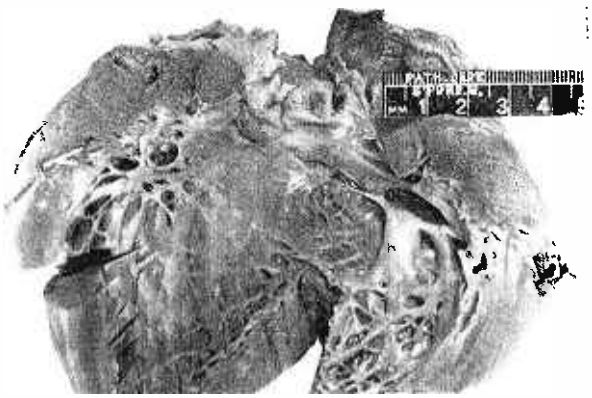

Fig. 4. Left ventricle showing the greyish white patch of endocardial fibroelastosis.

of 0·5 cm. (Fig. 3). The pulmonary cusps leaflets were normal looking but there was incompetence. The endocardium in the left ventricle showed a glistening greyish-white appearance almost involving the whole of the ventricular wall. It was in continuation with the aortic valves (Fig. 4). There were also isolated whitish patches in the right ventricle. However, the intimae of the aorta and pulmonary artery were normal. There was no thrombus present. The mitral and tricuspid valves were normal. Microscopically the endocardium of the left ventricle showed marked thickening of the elastic tissue (Fig. 5). There was no evidence of inflammation in the myocardium or endocardium. There was hypertrophy of the myocardial fibres and small areas of myocardial infarcts were seen.

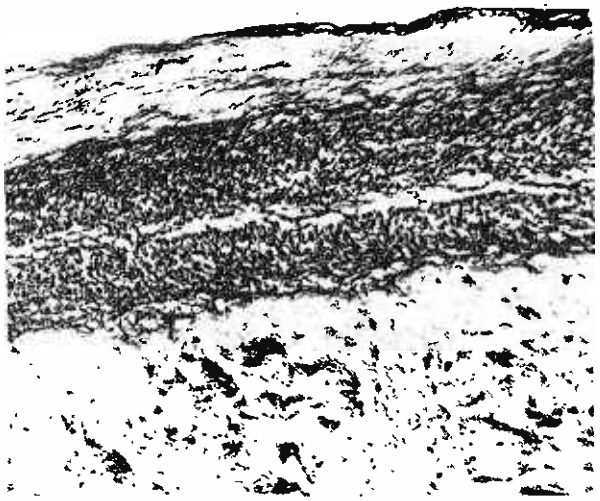

Fig. 5. Verhoeff's elastic tissue stain showing the elastosis of the endocardium.

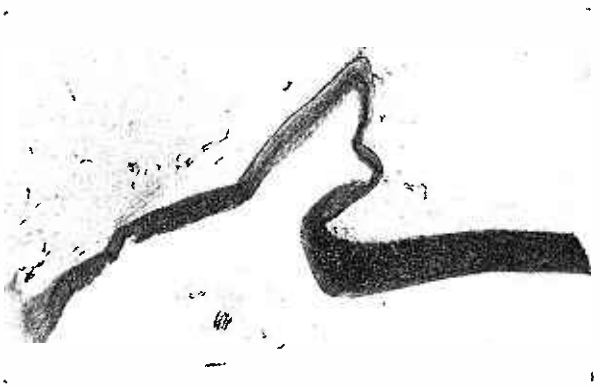

Fig. 6. Verhoeff's elastic tissue stain showing the elastosis in the endocardium is continuous with that of the aortic valve.

Examination of the lungs revealed congestion and oedema especially of the right lung. There were several cysts largest 1 cm. in diameter, at the periphery of both lungs. At microscopic examination of the lungs congestion and oedema were confirmed. There were many pigment-containing "heart failure" cells. The arterioles showed evidence of pulmonary hypertension of media in arterioles less than 100, medial hypertrophy of the larger arterioles and dilated vein-like arteries were also prominent features. Subintimal fibrosis was uniformly seen. The cysts were lined by a single layer of low cuboidal epithelium.

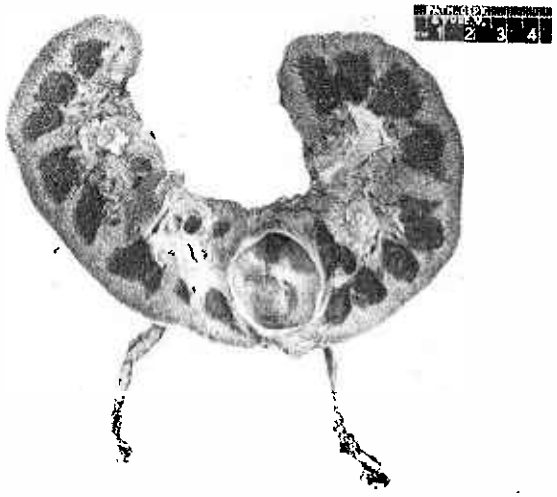

Fig. 7. Shows the horse - shoe kidney with a thick walled cyst at the site of fusion. The ureters are seen coming down from the anterior surface.

The kidneys weighed 295 gm. Both kidneys were fused at the lower pole forming a horse-shoe kidney. At the site of fusion there was a thick walled cyst (Fig. 7). Both pelves were situated anteriorly and both ureters crossed in front of the fused lower pole (Fig. 7). Microscopically the cyst was lined by cuboidal epithelium with a thick fibrous wall. The tubules were widely separated by oedematous stroma and they appeared like glomerular structures, most reminiscent of the embryonal kidney. The kidney tissue immediately next to the cyst showed chronic pyelonephritic changes with thickened Bowman's capsule and hyalinised glomeruli. The kidney tissue further away was normal.

The liver weighed 841 gm., was congested with a nut-meg appearance, and microscopically it showed centrilobular haemorrhage of the nut-meg type.

DISCUSSION

This is a case of fibroelastosis of the heart involving the aortic valve. There is a stenosis of the aortic valve with diffuse fibroelastosis of the endocardium in the left ventricle, with overgrowth of elastic fibres involving both valves and endocardium.

Whether the endocardial sclerosis is primary or secondary to hypertrophy of the left ventricle resulting from aortic stenosis is difficult to conclude. The degree of thickening of the endocardium of the left ventricle, marked proliferation of elastic tissue, absence of organised thrombus in the endocardium, lack of evidence of inflammation and fibrosis in the underlying myocardium and presence of other congenital anomalies however suggest that the endocardial fibroelastosis was primary in this case. The cardiac lesions in this case are identical with those described by Dushane and Edwards[5] and Gowing[6]. Cases of endocardial thickening secondary to diseased valves presents usually a different picture with amorphous organised thrombus formation, the thickening is mainly collagenous with little elastic tissue proliferation at the base and round the circumference. Endocardial pockets or pseudovalves may be present (Still 1961)[4]. Among the cases in which valvular disease was associated with endocardial fibroelastosis of the left ventricle the mitral valve was more commonly affected than the aortic valve and in some cases there was co-existent mitral and aortic stenosis. In this case the presence of severe aortic stenosis must bring up the consideration of this being rheumatic in origin especially in the presence of a radiological picture suggesting bi-ventricular hypertrophy. However the absence of history of rheumatic fever, the presence of marked endocardial sclerosis, absence of scarring of the myocardium and the presence of other congenital anomalies make rheumatic fever as the cause of aortic stenosis most unlikely and favour its congenital origin. The medial hypertrophy of the muscular arteries in the lung in this case is most likely to be due to pulmonary hypertension secondary to left ventricular failure. In a case like this the endocardial fibroelastosis has a binding effect on the left ventricle so that the probability of obtaining an adequate function of the left ventricle by the relief of the aortic stenosis is therefore of some doubt. The immediate cause of death in this case was congestive cardiac failure.

SUMMARY

A case of endocardial fibroelastosis involving mainly the left ventricle and the aortic valve is presented. The important clinical features included dyspnoea, a large heart, triple rhythm and a systolic thrill and murmur over the aortic area; the electrocardiogram showed marked left ventricular hypertrophy and the roengentrogram revealed bi-ventricular enlargement. The most important pathological findings were fibroelastosis involving the left ventricular cavity and the aortic valve.

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