INTRAUTERINE MYOCARDIAL INFARCTION

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In infancy myocardial disease other than myocarditis is undoubtedly rare, but infarction resulting from coronary artery occlusion and glycogen storage disease of the heart occasionally occur. Still more unusual apparently is the lesion first reported in 1952 by Uhl in which the right ventricular myocardium is virtually absent, and this curious defect is likely to present serious diagnostic difficulty. Uhl (1952) regarded it as representing an intra-uterine failure in development of the primordium of the right ventricular myocardium, but our observations of the case to be reported lead us to take a different view of its pathogenesis.

Case Report

A. MacL. was the third child of healthy parents. Her two sibs were healthy. She was born after a normal pregnancy and weighed 2.95 kg. She cried immediately after birth and was not then cyanosed. Aged 8 days on dismissal from the maternity hospital, she seemed well except for a slight mucoid nasal discharge. During the next 10 days she continued to feed hungrily but vomited occasionally. Persistent cyanosis was first noticed at the age of 19 days and this was increased by feeding or crying. She became irritable and during the three days preceding her admission to hospital at least half of every feed was vomited. When admitted at the age of 26 days she was well developed, vigorous and afebrile, but had marked central cyanosis and dyspnoea at rest. The heart rate was 150 per minute. There was no oedema but the liver edge was felt two and a half fingerbreadths below the costal margin. No cardiac pulsation was palpable. The heart sounds at all areas were of poor quality but no murmur was heard. In the electrocardiogram the waves and complexes were small in all the recorded leads, particularly in V4R, and there was no evidence of ventricular hypertrophy (Fig. 1). A radiograph of the chest disclosed a large heart shadow, some concavity of the pulmonary segment and clear lung fields (Fig. 2). No methaemoglobin was detected in the blood.

On clinical grounds transposition of the great arteries was suspected but the radiological appearances raised the alternative possibility of Fallot's tetrad. The electrocardiographic features, however, were not in accord with either diagnosis.

During the next five days the cyanosis and dyspnoea persisted without substantial change except for accentuation of cyanosis during feeding and crying. The average respiratory rate was 50 per minute. She took feeds eagerly but vomited occasionally. Oxygen was administered after feeds with beneficial effect on her colour. Suddenly and quite unexpectedly she died on the sixth day after admission at the age of 32 days.

Findings at Autopsy. The body was that of a normally developed 5-week-old female child weighing 3,458 g. There was no external sign of disease. Neither lungs nor liver showed congestion.

The only remarkable anatomical observation was made in the heart which weighed 24 g. and was normal in shape. On routine examination a clot was found at the apex of the right ventricle adherent to the wall. The ventricular wall was thin but not excessively so. The foramen ovale seemed competently closed by a valve. The ductus arteriosus was still patent but probably closing. There was no anomaly of the coronary vessels.

Several blocks of the heart were made for examination. Neither in the left atrium nor left ventricle was any muscular lesion found. Nowhere was a coronary artery lesion found. The striking feature was absence of muscle from the right ventricular wall, which is well shown in Fig. 3, and an attempt was made to determine whether this was a developmental defect or the result of some catastrophe. The sharp demarcation between the normal muscle of the left ventricle and its absence in the right is seen in Fig. 4. Furthermore, in the right ventricular wall (Fig. 4) a collection of deposited calcium and giant cells was recognized; in Fig. 5 they can be seen at a higher magnification. In spite of the presence of calcium these multinucleated giant cells are not of the usual foreign body reaction type; it was thought possible that they could be the remains of muscle cells, but repeated attempts to demonstrate cross-striation failed. From a general examination of the apex of the right ventricle the firm impression was gained that the tissue examined was in fact granulation tissue, and Fig. 6 shows the high vascularity of the tissue of a pattern typical of granulation tissue. Here again after extensive examination no cross-striation was recognized and therefore no evidence of muscle cells. The apex of the right ventricle was proved to have an adherent clot showing extensive organization with the right ventricular wall;

FIG. 1.—All waves and complexes are small and there is no evidence of hypertrophy of either ventricle.

FIG. 2.—The teleradiogram shows cardiomegaly, some concavity of the upper left heart border and clear lung fields.

this granulation tissue reaction could be found for some 3 cm. above the apex. Also throughout the right ventricular wall at the apex were small collections of round cells, again suggesting an inflammatory basis for the lesion.

Discussion

A myocardial defect closely similar if not identical to that reported here has been described by Uhl (1952), Castleman (1952) and Taussig (1960) in patients whose ages at death were 8 months, 24 years and 6 years respectively. Congestive heart failure preceded death in each instance. Taussig

FIG. 3.—Apex of heart (P.A.H. $\times 6.3$). This shows absence of muscle in the right ventricle.

FIG. 4.—Junction of right and left ventricle at apex (P.A.H. $\times 26$). There is sharp demarcation between right and left ventricle; coronary vessel is patent. Calcium deposition in right ventricular wall.

FIG. 5.—Higher power of right ventricular lesion where calcium is deposited and giant cells have formed. No evidence of muscle. (Haematoxylin and eosin $\times 124$.)

FIG. 6.—Wall of right ventricle at apex. No evidence of muscle. A vascularized scar. (P.A.H. $\times 68$.)

(1960) described another example in which at 17 months of age the diagnosis was made by means of cardiac catheterization and angiocardiography. Ebstein's anomaly of the tricuspid valve was later proved to be present in addition. In the other cases the correct diagnosis was not suspected during life and the diagnostic possibilities entertained included Fiedler's myocarditis or congenital heart disease (Uhl, 1952), Fallot's tetrad (Taussig, 1960), idiopathic cardiac hypertrophy or myocarditis of unknown aetiology (Castleman, 1952).

In the present case the diagnosis which seemed most probable was Fallot's tetrad or transposition of the great arteries, but the absence of a heart murmur and, in particular, the electrocardiographic findings were considered to be exceptional features. Uhl's patient (1952) had no murmur and although Taussig (1960) heard an apical diastolic murmur and Castleman (1952) described a pulmonary systolic murmur, it is difficult to attach any diagnostic significance to these. As in the electrocardiogram of the present patient, QRS complexes of low voltage were observed by Uhl (1952) and by Castleman (1952), but tall P waves were also described by the former author and by Taussig (1960). In the present case the concave pulmonary segment of the heart on the radiograph, together with the clear lung fields led to the conclusion that infundibular stenosis was likely and lent support to the erroneous diagnosis of Fallot's tetrad. The cardiomegaly was, however, a suspicious feature. In the description of her second patient Taussig (1960) referred to a large heart shadow and clear lung fields on the radiograph. Her first patient diagnosed incorrectly as Fallot's tetrad seems likely also to have shown clear lung fields. Uhl (1952) described the heart shadow of his patient as being grossly enlarged and globular.

It is clear, therefore, that this defect of the right ventricular myocardium is yet another cyanotic type of congenital heart disease in which the electrocardiogram may not show right ventricular dominance and that this taken in conjunction with clinical evidence of right ventricular enlargement, cardiomegaly and clear lung fields should suggest the possibility of the correct diagnosis.

The cause of cyanosis in our patient is not certain, but although at autopsy the foramen ovale seemed soundly covered by a valve it is likely that in life right atrial hypertension could have caused the valve to open so permitting venous blood to pass into the left atrium. It is probable that peripheral stagnation of blood in a failing circulation contributed to the cyanosis. The ductus arteriosus can scarcely be incriminated as the site of a veno-arterial shunt, because it seems impossible that the

pulmonary artery blood pressure could have exceeded that in the aorta.

In her account of this lesion Taussig (1960) has described it as *parchment right ventricle* and by so doing recalls its naked-eye resemblance to the myocardial lesion of unknown pathogenesis which affected both ventricles of the heart of a middle-aged man in Osler's collection and which was referred to by Segall (1950) as *parchment heart*. Uhl (1952) thought that the extensive defect of the right ventricular myocardium which he described represented injury during intrauterine life and was most probably the result of a failure of growth of the right cardiogenic fold. He considered but rejected coronary artery occlusion and foetal endomyocarditis as causal. The present patient died at the age of 32 days the opportunity thus being given for the lesion to be studied at an earlier stage in its development. There can be no doubt that it originated during intrauterine life, but because of the replacement of the right ventricular myocardium by granulation tissue, the case for a defect in the development of the primordium of the right ventricle does not seem tenable. Nevertheless, the cause for an inflammatory lesion is difficult to produce. The possibility of foetal endomyocarditis is ruled out because there was little evidence of an active infection at the time of examination, the cellular reaction expected of a myocarditis was lacking and the lesion was sharply limited to the right ventricular myocardium. No

occlusion or aberration of coronary vessels was found, but we are of the opinion that the only tenable theory is that this clearly demarcated lesion in which granulation tissue was so abundant represents myocardial infarction as the result of severe ischaemia due to occlusion of the right coronary artery at some time during the course of intrauterine life.

Summary

An infant with cyanotic congenital heart disease who died at the age of 32 days is described. The right ventricular myocardium was found to be replaced by granulation tissue. The difficulty of clinical diagnosis is discussed. The lesion may reflect intrauterine infarction of the right ventricular myocardium.

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