RIGHT VENTRICULAR ANEURYSM DUE TO CONGENITAL MUSCULAR DEFECT IN AN ADULT

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A large right ventricular aneurysm was detected by an echocardiographic examination in a 50-year-old woman presenting with weakness, collapse and hypotension. At surgery, another smaller aneurysm, arising from the anterior infundibulum of the right ventricle, was found in addition to the large one. Both were resected. Histological studies on the resected specimen revealed that much of the myocardium was replaced by adipose tissue. As far as we know this is the first reported case in Japan in which two congenital aneurysms arose from the right ventricle as a result of muscular defect.

CONGENITAL ventricular aneurysm and diverticulum are rare anomalies of the heart. Those arising from the right ventricle have been said to be extremely rare, although at least 16 cases have been reported in world literature up to 1984¹. The following is a report of a 50-year-old woman with two ventricular aneurysms arising from the right ventricle which were detected by echocardiographic examination.

CASE REPORT

A 50-year-old woman was referred because of weakness, collapse and hypotension.

The patient was admitted to the Department of Orthopedics, Fukaya Red Cross Hospital, on Jan. 17, 1984, as a result of a traffic accident which caused fractures of both legs and blunt trauma to the chest. The hospital course was uneventful until Jan. 25, when she felt weakness while walking and collapsed. She was referred to

the Department of Internal Medicine.

The patient had been told that she had an enlarged heart at an annual X-ray examination 10 years ago. Since that time, she had occasionally experienced discomfort and fatigue during mild physical exercises but no palpitations or chest pains. The family history was not contributory.

The patient was a slender woman. Her blood pressure was 70/46 mmHg. The heart rate was 90 and regular. An ecchymosis, approximately 2 cm in a diameter, was observed on the left anterior chest. The lungs were clear; no rales, rhonchi, or pleural friction rub was heard. The cardiac dullness was 2 cm enlarged to the left. A grade 2/6 musical systolic murmur was heard at the lower left sternal border. The abdomen was normal; the liver, kidney and spleen were not palpable. Bruises were present on both legs.

The routine hematological and biochemical examinations were within normal limits except for a slight elevation of the serum CPK on admission.

The X-ray films of the chest showed cardiac enlargement without pulmonary congestion. The

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Fig.1. Chest X-ray films showing enlarged cardiac shadow. In lateral view, the cardiac shadow was enlarged posteriorly and inferiorly. A small amount of effusion was shown in the postero-inferior pleural space.

Fig.2. Electrocardiogram on admission revealed occasional ventricular premature beats. T waves were inverted in II, III, aVF and V₃ through V₆.

cardiothoracic ratio was 62.5%. In lateral view, a small amount of effusion was shown in the postero-inferior pleural space (Fig. 1).

The electrocardiogram showed a sinus tachycardia at a rate of 100 with occasional ventricular premature beats. The T waves were inverted in leads II, III, aVF and V₃ through V₆ (Fig. 2).

The echocardiographic examinations revealed an aneurysmal sack, 6.5 × 4.5 cm in size, at the apex of the right ventricle. This sack expanded slightly in the systole. Several cord-like echoes were observed at the connecting portion between the sack and the right ventricle (Fig. 3).

Because of low blood pressure and suggestive

Fig.3. Echocardiogram showing a large aneurysmal sac (X) protruding from the right ventricle (RV). Several cord-like structures were seen in the connecting hole between the right ventricle and the sack.
AO : aorta, LA : left atrium, LV : left ventricle, RV : right ventricle, X : aneurysmal sack.

Fig.4. Angiographic study showing a large aneurysmal sack protruding from the right ventricle.
RA : right atrium, RV : right ventricle, X : aneurysmal sack.

Fig.5. Surgical finding. Left: In addition to a giant aneurysmal sack protruding from the apex of the right ventricle, a smaller sack was found at the anterior infundibulum of the right ventricle. Right: Inside view of the right ventricle.

Fig.6. Microscopic section from the wall of the right ventricular aneurysm. While the endocardium was thickened, the myocardium was replaced by adipose tissue. (HE stain)

signs of low perfusion, $4 \mu\text{g/Kg/min}$ of dopamine was infused intravenously. There were no other symptoms indicating acute myocardial infarction or acute myocarditis. The systolic pressure rose to 110 mmHg, and the patient did not complain of weakness thereafter. She was transferred to the First Department of Surgery, Saitama Medical College on Jan. 27, 1984, for further evaluation and operation.

An angiographic study performed before the surgical operation revealed a giant aneurysmal sack protruding from the apical portion of the right ventricle inferiorly and posteriorly (Fig. 4). At the right heart catheterization, the mean pulmonary artery wedge pressure was 15 mmHg, while the right ventricular pressure was 39/4–10 mmHg. The pressure in the aneurysmal sack was 38/4 mmHg, virtually the same as in the main cavity of the right ventricle. No oxygen step up was demonstrated.

At surgery, a giant aneurysmal sack arising from the right ventricular apex, $8 \times 6 \times 5.5$ cm in size, was observed. This sack evidently had been moved into the right ventricular cavity through a wide hole with a diameter of 4 cm. Several tricuspid tendons were attached to the wall of the sack through this connecting hole. In addition, a smaller sack measuring approximately 2 cm in a diameter was also found at the anterior

infundibulum of the right ventricle (Fig. 5). Both sacks were resected. The coronary arteries were normal.

Histological findings: The wall of the resected sack was mainly composed of adipose tissue. Only a few scattered hypertrophied myocardial cells surrounded by fibrous tissue were observed. On the whole, degenerative change was not obvious and there was no evidence of inflammatory reaction (Fig. 6).

DISCUSSION

A large aneurysmal sack arising from the right ventricle was detected by echocardiographic studies and another smaller sack arising from the anterior infundibulum of the right ventricle was found at surgery in this patient. Both were surgically resected. We initially suspected traumatic aneurysm, since other possibilities of acquired ventricular aneurysms were thought to be unlikely. However, histological studies on the resected specimen revealed almost total absence of the sack myocardium that did not have obvious degenerative change. Only few scattered myocardial cells remained. Thus, our final diagnosis was congenital aneurysm of the right ventricle due to muscular defect.

The differentiation between congenital aneu-

rysm and diverticulum is not very distinct. Treistman² noted that the term congenital diverticulum has been used when the point of connection to the ventricle is narrow and there are midline defects; the term congenital aneurysm has been used when the point of ventricular connection is wide and no midline defects are present. On the other hand, Gueron and associates³ did not consider the presence or absence of thoraco-abdominal defects to be critical, because there were diverticula that are not associated with these defects. According to these authors, a congenital aneurysm is one that results from a muscular defect and is connected to the ventricular cavity by a wide opening. As for diverticula they subdivided that to 3 types: (1) muscular diverticula with thoraco-abdominal defects, (2) fibrous type of diverticula and (3) others. In the present case, the connections between the sacks and the right ventricular cavity are wide, while the myocardium is almost entirely replaced by adipose tissue. Therefore, congenital ventricular aneurysm seems to be the appropriate term.

The above histological findings were also compatible with Uhl's anomaly. Since the first description of an infant with almost total absence of the myocardium of the right ventricle by Uhl⁴ in 1952, several investigators^{5,6} have reported this rare malformation of the heart. Although the epicardium and endocardium were said to lie adjacent to each other in the 1952 case, the myocardium may be replaced by adipose tissue or by fibrous tissue in this anomaly. Moreover, a partial type of this anomaly^{7,8} characterized by the partial absence of the right ventricular myocardium, has been reported^{9,10,11}. So, the present case may be called a partial type of Uhl's anomaly with ventricular aneurysm. There have been at least 4 cases of Uhl's anomaly with right ventricular aneurysm^{7,12,13,14}. Of these, two had giant aneurysm^{7,14}. A case similar to ours was reported by Marcus and associates, who found two aneurysms, one at the apex and another at the infundibulum of the right ventricle.

The exact cause of hypotension, weakness and collapse, which necessitated medical evaluation in this case, was not obvious. The giant aneurysm of the right ventricle may have impeded the right

ventricular function and resulted in low cardiac output.

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