A CASE REPORT OF ANOMALOUS ORIGIN OF A SINGLE CORONARY ARTERY FROM THE PULMONARY ARTERY ASSOCIATED WITH MULTIPLE CARDIAC MALFORMATIONS

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Anomalous origin of a single coronary artery from the pulmonary artery is exceedingly rare. Clinical and necropsy findings in a 17-month-old boy with anomalous origin of a single coronary artery from the pulmonary artery are described. In our case, no areas of infarction or ischemic changes were found as well as in 4 of the 5 previously reported cases. It is most important that surgical treatment be performed without delay.

ANOMALOUS origin of either one or both coronary arteries from the pulmonary artery is rare. Especially, anomalous origin of a single coronary artery from the pulmonary artery is exceedingly rare. To the best of our knowledge, only 5 cases have been reported in the literature¹⁻⁵. We report on a 17-month-old boy who had anomalous origin of a single coronary artery from the pulmonary artery associated with multiple cardiac malformations: ventricular septal defect, patent foramen ovale, aortic bicuspid valve, coarctation of aorta and scimitar syndrome.

CASE REPORT

The patient was born weighing 2.9 kg in an uncomplicated pregnancy, labor, and delivery. The patient was first found to have a heart murmur and congenital heart disease at one week

of age. There was no family history of congenital heart disease. He was affected by respiratory infection with cough and dyspnea at the age of 6, 7 and 9 months. In April 1980, he was admitted to Miyazaki Prefectural Hospital because of cough and dyspnea. After 2 days, cardiac and respiratory arrest developed and resuscitation was done. After resuscitation he was transferred to Miyazaki Medical College Hospital by ambulance, where he died after 2 hours.

Examination on admission to Miyazaki Medical College Hospital revealed a heart rate of 140/min, respiratory rate of 40/min (mechanical respiration). The patient was unconscious and the peripheral pulses were very poor in all extremities. There was clubbing of the fingers with cyanosis of the nails. As external anomalies, there were hypospadias and bilateral retentio testis. Deformity of the anterior wall of the chest was not present. Breath sounds were decreased in the right lung field and piping rales were heard over the whole lung field. No heart murmur was heard on admission. The liver was palpable one cm below the right costal margin, smooth and of normal consistency. Spleen and kidneys were not palpable.

Key Words:

Single coronary artery from the pulmonary artery
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Coarctation of aorta

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Fig.1. Plane chest X-ray. Arrows show scimitar-sign.

Plane chest X-ray showed situs solitus, hypoplasia of the right lung, dextroposition of the heart, and a scimitar-shaped anomalous vein along the right heart border (Fig. 1).

An electrocardiogram revealed the pattern of right ventricular hypertrophy: the mean electrical QRS axis was $+210^\circ$ and the QRS wave in all precordial leads were RS pattern (Fig. 2). To our regret, echocardiography, cardiac catheterization and angiocardiology were not performed because the patient died soon after admission.

NECROPSY FINDINGS

Except for hypospadias and bilateral retentio testis, abnormal findings were confined to the heart and lungs. Each lung showed lobular abnormality, consisting of one lobe. In comparison with the left lung, the right lung and the right pulmonary artery showed a decrease in size. An anomalous vein from the right lung descended into the inferior vena cava below the diaphragm.

The aorta and the pulmonary artery were in the normal position. A patent foramen ovale was in the atrium and a ventricular septal defect, measuring 4×5 mm in diameter, was in the papillary muscle of the conus. A ductus arteriosus was obstructive and juxtaductal coarctation of the aorta was slightly seen. A single coronary ostium was present in the left sinus of the pulmonary artery (Fig. 3). Autopsy revealed no coronary ostia in the aorta, the valve of which was bicuspid. The right aortic valve was markedly thick (Fig. 4). The main distribution of the single coronary artery is shown in Fig. 5. Its ostium was found in the left pulmonary sinus. About one cm from its origin, it gave off a branch corresponding to the left circumflex artery. Then it divided into 2 branches. One was the right coronary artery which reached the right atrioventricular groove across the right ventricular wall. The other branch was the left anterior descending coronary artery. It descended over the anterior aspect of the heart in the interventricular sulcus, following the usual course for the anterior descending artery. No areas of infar-

Fig.2. Electrocardiogram

Fig. 3. A single coronary ostium was present in the left sinus of the pulmonary artery (arrow).

tion or ischemic changes were found. The lung and liver were slightly congested. The other organs were normal.

DISCUSSION

Anomalous origin of either one or both coronary arteries from the pulmonary artery is rare.⁷⁻⁹ The variations of this malformations are as follows: 1) anomalous origin of the left coronary artery arising from the pulmonary artery (Bland-White-Garland syndrome), 2) anomalous origin of the right coronary artery from the pulmonary artery, 3) anomalous origin of both coronary arteries from the pulmonary artery and 4)

Fig. 4. Autopsy revealed no coronary ostia in the aorta. The aortic valve was bicuspid. RAV = right aortic valve, LAV = left aortic valve.

Ao: Aorta, PA: Pulmonary Artery
RAA: Right Atrial Appendage
LAA: Left Atrial Appendage
RCA: Right Coronary Artery
LAD: Left Anterior Descending Artery

Fig. 5. The main distribution of the single coronary artery.

TABLE I ANOMALOUS ORIGIN OF SINGLE CORONARY ARTERY FROM THE PULMONARY ARTERY

Author	Year	Age at Death	Sex	Associated Cardiovascular Anomalies	Origin of Single Coronary Artery	Other Findings	Ischemic Change	Autopsy
1. Tow ¹	1931	5 months	F.	PTA, Cor biloculare, Common bicuspid valve	Left pulmonary artery		—	+
2. Colmers et al. ²	1963	33 days	M.	None	Left pulmonary sinus		+	+
3. Feldt et al. ³	1965	7 years	F.	VSD, PDA, PFO, MS	Left pulmonary sinus	Absence of gallbladder	—	+
4. Ogden ⁴	1969	6 days	F.	PDA, PFO	Left pulmonary sinus		—	+
5. Monselise et al. ⁵	1970	1 year	M.	VSD, PFO	Right pulmonary sinus		—	+
6. Present case		1-year-5-months	M.	VSD, PFO, Co/Ao, Scimitar syndrome, Aortic bicuspid valve	Left pulmonary sinus	Hypospadias, Retentio testis	—	+

PTA = persistent truncus arteriosus, VSD = ventricular septal defect, MS = mitral stenosis, PDA = patent ductus arteriosus, PFO = patent foramen ovale, Co/Ao = coarctation of aorta.

anomalous origin of a single coronary artery from the pulmonary artery. Over 90%⁹ of the cases of abnormal origin of a coronary artery are Bland-White-Garland syndrome: approximately 200 cases⁷ have been reported. Other cases [2)–4)] are rare and several cases have been reported. Anomalous origin of a single coronary artery from the pulmonary artery is extremely rare, only 5 cases having been reported in the literature^{1–5} (Table I).

The prognosis of these cases is related to the presence of additional cardiac defects and a well developed coronary tree that allows increase of coronary flow and perfusion. These cases were associated with other cardiac anomalies, such as a persistent truncus arteriosus, ventricular septal defect, patent ductus arteriosus and patent foramen ovale, etc.

The age at death was within one year of birth in 4 of the 5 cases; 2 of them died in 6 and 33 days of life respectively. Only one case survived until 7 years. In this 7-year-old girl, described by Feldt,³ her coronary circulation was maintained by a single coronary artery arising from the pulmonary trunk. The presence of patent ductus arteriosus and ventricular septal defect allowed perfusion of the single coronary artery at high pressure. Correction of these cardiovascular defects, however, removed the sources of high pressure in the right heart and led rapidly to her

death by decreasing the coronary perfusion pressure. Our child survived until 17 months due to the presence of ventricular septal defect, patent foramen ovale, and right pulmonary venous drainage to the inferior vena cava, which increased the oxygen saturation of the pulmonary arterial blood and elevated the pulmonary artery pressure.

It is interesting to have found myocardial infarction in only one of the 5 previously reported cases. No ischemic changes were noted in 4 cases. In our case, also no areas of infarction or ischemic changes were found.

Anomalous origin of the coronary arteries from the pulmonary trunk is thought to be due to defective development of the septum dividing the truncus arteriosus. Blake⁶ stated that if the anlage of the coronary artery arises low, its origin will lie in the aortic segment. On the other hand, if the take-off is high, it will originate from the pulmonary side. According to Blake's theory, this anomaly can possibly occur if the anlage of the coronary artery is single and its take-off is high.

In 5 of 6 cases, including the present case, the single coronary artery was found to be the left coronary artery. Ogden⁴ stated that the left coronary artery is usually the vessel affected when one coronary artery arises from the pulmonary artery and one arises from the aorta.

These cases run the risk of sudden death from coronary insufficiency. It is most important that cardiac catheterization and surgery (e.g., direct anastomosis between aorta and single coronary artery, aorto-single coronary saphenous vein bypass, anastomosis between left subclavian artery and single coronary artery) be performed without delay.

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