Case Reports

Congenital Aortic Arch Interruption

Report of a Case, Surgically Treated with Success

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THE congenital interruption of the aortic arch is a very rare anomaly and only 105 autopsied or clinical cases including 4 successfully surgically treated ones, have been reported in the literature by 1965.¹⁾ Almost of these cases were associated with various anomalies, such as ductus arteriosus, ventricular septal defect, atrial septal defect, hypoplastic ascending aorta and various left heart anomalies, and usually died within a few months after birth. It is interesting fact that there has been found only 2 reports of the interruption without any other anomalies in the literature.^{2), 3)} Recently the authors experienced the successful surgery of the isolated interruption and so will present the case report herewith.

CASE REPORT

A 12-year-old well nourished Japanese girl was admitted to the University

Fig. 1. The preoperative chest X-ray film. The arrow shows the rib-notching.

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Hospital under the diagnosis of the coarctation of the aorta with complaints of headache and palpitation with exercise. She had never been cyanotic and did not demonstrate clubbing in fingers and toes.

The chest examination revealed no deformity nor thrill, but the early systolic murmur (1/6) at the cardiac base and the systolic bruits at the left supraclavicular groove were heard.

Fig. 2

Fig. 3

Fig. 4. The operative findings. A tape and Nelaton's catheters show the dilated ascending aorta, the brachiocephalic artery, the left common carotid artery and the thoracic descending aorta, respectively.

Fig. 5

The blood pressure was 210/70 mm.Hg in the right arm, 120/60 mm.Hg in the left arm and 90/70 mm.Hg in the leg, although the femoral pulsation was difficult to palpate. The blood pressure of the retinal vessels was 71/43 mm.Hg in the right side and 51/37 mm.Hg in the left. The hematologic findings and the liver and the renal functions were within normal range.

The chest X-ray picture showed a slight widening of the cardiac base without cardiomegaly and notching of the right 9th rib (Fig. 1). The electrocardiography indicated the marked left ventricular hypertrophy (Fig. 2). The retrograde aortography by the catheter inserted from the right brachial artery revealed the obstruction of the aortic arch probably at the proximal portion of the left subclavian artery and marked collateral channels (Fig. 3). The catheter, inserted from the femoral artery to the aorta, ran toward the neck and the left upper arm, but the left subclavian artery and the left common carotid artery were not opacified. She was operated upon as the coarctation of the aorta probably with the obstruction at the proximal portion of the left subclavian artery in May 6, 1966.

Operative findings and procedures: The left lateral thoracotomy was performed through the 4th intercostal space. The thoracic descending aorta was small and thin-walled and its outside diameter was 9 mm. It was originated at the upper thoracic level from two branches which descended from the neck. Then, the median sternotomy was performed. The ascending aorta was enlarged (32 mm. in diameter) and bifurcated. No connection was detectable between the ascending and descending aortae. Two branches of the ascending aorta were the brachiocephalic trunk and the left common carotid artery (Fig. 4). The electroencephalogram and the left brachial pulsation were not affected by occlusion of the left common carotid artery.

A woven Teflon graft of 12 mm. in diameter was anastomosed to bypass between the ascending aorta and the thoracic descending aorta (Fig. 5). The postoperative course was smooth and uneventful.

Fig. 6

Fig. 7. The postoperative left-sided angiogram reveals the good patency between the ascending aorta and the thoracic descending aorta.

The blood pressure rose to 130/90 mm.Hg in the left arm and 120/90 mm.Hg in the leg, but could not be measured in the right arm due to the preoperative catheterization. The blood pressure of the retinal vessels decreased to 64/42 mm.Hg in the right side and 51/34 mm.Hg in the left.

The postoperative angiocardiology was performed for reconfirmation of this anomaly and evaluation of the bypass grafting. The graft was completely patent. The descending aorta, the left vertebral artery and the left subclavian artery were well opacified and the intracardiac anomaly and the ductus arteriosus were not demonstrated. It was reconfirmed that no connection between the ascending aorta and the thoracic descending aorta in the thoracic cavity, except the bypass graft, was present (Fig. 6, 7).

DISCUSSION

Celoria⁴⁾ classified the complete interruption of the aortic arch into three main types (A, B, C), based on the sites of origin of the branches of the aortic arch. This anomaly appears to result from regression or atrophy of the dorsal aorta (A), failure of formation of the left fourth arch (B) or the third and the fourth arches (C). All cases previously reported as the complete interruption or absence of the aortic arch were accompanied always with the pulmonary-ductus-descending aorta trunk if it to survive after birth, like as the fetal circulation. Among other various associated anomalies, ventricular septal defect is the most frequent, and the combination of aortic arch interruption, ductus arteriosus and ventricular septal defect is well known as the trilogy or triad of Everts-Suarez and Carson.⁵⁾

As compared with the frequently reported features of the complete interruption of the aortic arch, the anomaly of the present case is different at the following points. (1) Her prognosis was exceptionally good inspite that the pulmonary-ductus-descending aorta trunk was not accompanied, although the longest survival was 14 years in the case with this trunk in the literature.⁶⁾ (2) The ascending aorta was not hypoplastic and rather enlarged with hypertension of the upper half of the body. The relative hypertension in the arm, compared with the leg, is infrequent, because the pressure in the pulmonary trunk and descending aorta is usually similar to that in the ascending aorta,⁷⁾ if there is the large pulmonary-ductus-descending aorta trunk. However, recently the cases of the complete interruption without significant hypoplasia of the ascending aorta have been reported, and 2 cases, successfully operated upon by Merrill⁹⁾ and Villalobos⁶⁾ had shown preoperatively the relative hypertension in the arm (160/80 mm.Hg and 220/120 mm.Hg, respectively). Therefore, these findings seem not always uncommon in this anomaly, especially in the case with the small pulmonary-ductus-descending aorta trunk. (3) The

differential cyanosis of the lower half of the body or clubbed toe and the intracardiac anomaly were not found, though they are not constant symptoms nor pathognomonic findings.

The "cervical loop aortic arch", one of the anomalies of the aortic arch reported by Mahoney and Manning,¹⁰⁾ presents some similarity to the present case, but this condition is the upward elongation of the aortic arch and can be differentiated by the angiography from the interruption.

The present anomaly is rather similar to the coarctation of the aorta clinically and hemodynamically. However, the obstruction proximal to the left subclavian artery (Karnell's type IV of the coarctation) is uncommon^{11),12)} and it is essentially different from the coarctation that the ascending aorta ended with the left common carotid artery without any connection, even a remnant, with the thoracic descending aorta.

Only 2 cases identical to the authors' case which was not associated with any other anomaly have been reported in the literature,^{2),3)} and one of them³⁾ who was a 16-year-old girl, revealed the same clinical feature to ours and was successfully operated upon by bypass grafting. Pillsbury³⁾ described the possible embryologic explanation of this defect as follows: Normal regression of the left-sided dorsal aorta between the left third and fourth aortic arches did not occur. This vessel persisted as the large vessel seen connecting the upper part of the descending thoracic aorta with the carotid vessels and is functionally the left common carotid artery. There was an abnormal regression of the left third, fourth, and sixth aortic arches, which explains the interruption of the aortic arch between the innominate artery and the functional left common carotid artery (persistent left dorsal aorta), and the absence of the left-sided ductus arteriosus. There was a right-sided ductus arteriosus, which became obliterated. Pillsbury's explanation may be also applicable to our case, although the interrupted position of his case was different from ours.

It is very interesting that these 3 cases were 32,²⁾ 16³⁾ and 12 years of age respectively and the prognosis of the anomaly without the pulmonary-ductus-descending aorta trunk is better than the anomaly with this trunk which has been thought as the staff of life. At all events, this type of the aortic arch interruption is a very rare anomaly and the authors' case is the third case in the literature and this is the fifth case with successful surgical correction of the aortic arch interruption.

SUMMARY

The clinical case of a 12-year-old girl with the interruption of the aortic

arch without the pulmonary-ductus-descending aorta trunk was reported. Surgical correction of the interruption was successfully achieved by anastomosing to bypass a Teflon prosthesis between the ascending aorta and the thoracic descending aorta.

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