Persistent Fifth Aortic Arch

An Ignored and Underestimated Disease

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SUMMARY

We report two unique cases of persistent fifth aortic arch with a systemic-to-pulmonary connection. All previously reported cases with such a connection in the literature have either been cases of pulmonary atresia or an aortic arch anomaly, and the existence of a fifth aortic arch was a benefit to the underlying great vessel anomaly. However, our two cases did not have this associated great vessel anomaly, and the fifth arch resulted in a large left-to-right shunt with severe pulmonary hypertension and heart failure. The first case was misdiagnosed preoperatively; an accurate diagnosis was made after cardiac surgery. Because of its rarity and complexity, a persistent fifth aortic arch is often ignored and misdiagnosed. (*Jpn Heart J* 2000; 41: 665-671)

Key words: Persistent fifth aortic arch, Systemic-to-pulmonary connection, Complex great vessel anomaly

PERSISTENT fifth aortic arch is an extremely rare congenital cardiovascular malformation. It is an abnormal arterial branch originating from the distal ascending aorta proximal and opposite to the ostium of the innominate artery.¹⁾ Most cases are associated with severe cardiac malformation and diagnosed fortuitously.²⁾ By 1989, only 21 cases had been reported in the literature.³⁾ Among them, 15 cases had a systemic-to-systemic connection, and six cases had a systemic-to-pulmonary connection. All six cases were associated with a variety of complex great vessel anomalies such as pulmonary atresia, aortic atresia or interruption of the aorta. The fifth arches in the reported cases were actually of some hemodynamic benefit to the patients. Recently, however, we encountered two unique cases of persistent fifth aortic arch with systemic-to-pulmonary connection without pulmonary atresia or an aortic arch anomaly. The fifth aortic arch caused a catastrophic event in both of these patients.

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CASE 1

A 4-year-old Taiwanese boy was transferred to our hospital because of a heart murmur and increasing exercise intolerance. This case has previously been reported in detail elsewhere, so we will describe his condition here only briefly.⁴⁾ After echocardiography, angiography and magnetic resonance imaging study, an abnormal vascular connection between the distal ascending aorta and pulmonary artery was well visualized. The huge abnormal vessel caused a large left-to-right shunt and severe pulmonary hypertension. He was regarded as having a right-sided aortic arch with giant atypical patent ductus arteriosus or distal aortopulmonary window. At surgery, a huge short duct-like vessel, arising from the distal ascending aorta and proximal to the innominate artery, was connected to the superior margin of the bifurcation of the pulmonary trunk (Figure 1A). Under temporary cardiopulmonary bypass and profound hypothermic circulatory arrest, the pulmonary artery was vertically opened and a "tunnel" opening, measuring 2.8×2.8 cm was found above the bifurcation of the pulmonary trunk (Figure 1B). A 3.0×3.0 cm Dacron patch was sutured near the junction between the fifth aortic arch and pulmonary trunk. The main pulmonary artery was then closed by continuous suture (Figure 1C). The postoperative course was uneventful and he has been doing well during the 4-year follow-up period.

CASE 2

A one-day old female baby was transferred to our hospital due to a suspected imperforated anus. Cloacal anomaly with rectal agenesis was noted and transverse colostomy was performed the same day. Unfortunately, the patient could still not be weaned off mechanical ventilation one month after the operation. Two-dimensional color Doppler echocardiography at the parasternal area showed a perimembranous ventricular septal defect with a large left-to-right shunt and pulmonary hypertension. Since this sick baby was in critical condition under mechanical ventilation, no other examination was arranged for her before the operation. Because the colostomy site was in the upper middle abdomen and because there were other associated anomalies, a pulmonary artery banding was arranged for intractable congestive heart failure.

The operation was performed through a small left anterolateral thoracotomy. To our surprise, we found a huge vessel arising from the distal ascending aorta just beneath and opposite the origin of the innominate

Figure 1. Operative findings of case 1. **A:** A huge fifth aortic arch (arrows) communicating between distal ascending aorta (AO) and pulmonary trunk (PA). **B:** The distal opening of fifth aortic arch (arrow) can be found. **C:** Post-operative picture RPA = right pulmonary artery; LPA = left pulmonary artery.

Figure 2. Two dimensional echocardiogram in case 2 demonstrated a huge fifth aortic arch (arrow) between distal ascending aorta (AO) and pulmonary trunk (PA).

artery and rejoining at the superior margin of the pulmonary trunk. We could do nothing but halt the operation. Another two-dimensional echocardiography from the subcostal left ventricular outflow tract area disclosed a large ventricular septal defect and a large communication between the left side of the distal ascending aorta and pulmonary artery (Figure 2). Cardiac catheterization revealed a total O_2 step-up of 40% from the right atrium to the pulmonary artery with high pulmonary artery pressure ($P_p / P_s = 1$). Aortography displayed a systemic-to-pulmonary connection between the distal ascending aorta and pulmonary trunk (Figure 3). The family refused to allow further surgical intervention and the patient expired at the age of 3 months.

Post-mortem examination of the heart showed the usual arrangement of the thoraco-abdominal organs and the atrial chambers with a concordant atrioventricular connection. A perimembranous ventricular septal defect measuring 5 mm in diameter was present with otherwise normal ventricular morphology. The ascending aorta and the main pulmonary artery were in their normal positions. However, a huge and short vessel of a diameter equal to that of the ascending aorta could be well visualized arising from the distal ascending aorta and ending at the superior margin of the main pulmonary artery (Figure 4). No arterial duct or ligament was identifiable.

Figure 3. An aortogram revealed an aorto-to-pulmonary shunt through the huge fifth aortic arch (arrow).

Figure 4. Post-mortem examination of case 2 revealed a huge fifth aortic arch (arrow) communicating the distal ascending aorta with the pulmonary trunk. RPA = right pulmonary artery; LPA = left pulmonary artery.

DISCUSSION

Due to difficulty in making the diagnosis, the real incidence of persistent fifth aortic arch may have been underestimated. Although Gerlis and his colleagues reported an incidence of about 1 in 330 in autopsy studies,³⁾ still only rare cases have been presented since their publication.⁴⁻⁷⁾ Almost half of the fifth aortic arches with systemic-to-pulmonary connection were misdiagnosed as either ductus arteriosus or aortopulmonary window,^{2,3)} just as happened in our first case. We had first interpreted the case as a giant atypical ductus arteriosus or distal aorto-pulmonary window. After surgery, however, because the distal openings of the persistent fifth aortic arches were at the superior margin of the bifurcation of the pulmonary trunk in both of our cases, they could not be regarded as being aortopulmonary windows. If the latter were the case, the distal extremity of this septum would lie at the proximal and inferior margin of the right pulmonary artery. The only possible structure that could communicate from the distal ascending aorta to the main pulmonary artery would be a persistent fifth aortic arch with systemic-to-pulmonary connection.³⁾ Although anatomically almost identical, the two conditions are embryologically quite distinct.

It is interesting to note that the potential hemodynamic consequence of a persistent fifth arch with systemic-to-pulmonary connection appears to be favorable in all six reported cases in the literature.³⁾ The fifth aortic arch provides a vital systemic-to-pulmonary shunt in four cases with pulmonary atresia and acts as a vital pulmonary-to-systemic shunt in two cases with aortic atresia or interruption of the aortic arch. Physiologically, the persistent fifth aortic arch seems to act as a collateral circulation in pulmonary atresia or aortic atresia. None are harmful to the patient. However, contrary to the previous reported cases, in our two cases, the fifth aortic arch caused or precipitated a large left-to-right shunt and severe heart failure. This demonstrated that the persistent fifth aortic arch may co-exist under different hemodynamic conditions.

Gerlis and his colleagues³⁾ stated that the persistent fifth aortic arch is by no means rare but only underestimated and misdiagnosed, and we agree with their conclusion. Awareness of the protean manifestations of the anomaly and some understanding of the embryological principles involved may facilitate more frequent and accurate diagnosis in the future. Thirty years after the first description of persistent fifth aortic arch, there is still much to be learned about this very interesting entity.

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