THE CERVICAL AORTIC ARCH

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SUMMARY

A patient with a cervical aortic arch associated with a coarctation of the aortic arch and a Tetralogy of Fallot presented to the General Hospital, Kuala Lumpur.

INTRODUCTION

Congenital malformations of the aortic arch are commonly encountered.[1] We report here a case where the aortic arch was located high in the neck and associated with a coarctation of the aortic arch and a Tetralogy of Fallot. Such a presentation is rare.

CASE HISTORY

NAB, a nine-year-old Malay female, presented at the age of five months with poor weight gain and recurrent upper respiratory tract infection; it was diagnosed as Tetralogy of Fallot. She had no hypercyanotic spells but was severely restricted in her activities. She was a full-term, spontaneous vertex delivery. Antenatal and postnatal periods were uneventful and her milestones were normal. She was small for age, blood pressure 90/60 mmHg; clinically she was not distressed but was cyanosed and clubbed. Her pulse rate was 100/min but her peripheral pulses were not well felt in comparison with the carotid pulse. The praecordium was relatively quiet. Both S1 and S2 were heard, S2 was single. A high pitched long systolic murmur (3/6) was audible at the mid-left sternal edge. No significant diastolic murmurs were heard. Electrocardiography showed right axis deviation, right ventricular hypertrophy with strain and also some left ventricular hypertrophy. Chest X-ray showed cardiomegaly, empty pulmonary artery bay and a right aortic arch. Her haemoglobin was 17.4g%.

2-D Echocardiography showed a Tetralogy of Fallot with an aortic arch anomaly (with a right-sided aortic arch). Barium swallow showed displacement of the oesophagus to the left side by the right-sided arch. No other anomalies were detected. Cardiac catheterization (Fig. 1) showed the aortic arch in an abnormal position at the cervical area on the right. The neck vessels were in the following order: left innominate artery, right internal carotid artery, right external carotid artery, right external carotid artery and right subclavian arterty; there was a coarcted segment just before the origin of the right-sided neck vessels (just before the origin of the right internal carotid artery). Other cardiac catheterization findings were consistent with the presence of a Tetralogy of Fallot.

DISCUSSION

In this condition, the ascending aorta extends in such a fashion that the aortic arch is situated high in the neck on either side.

Embryology

A double aortic arch forms the basic concept to explain the development of the cervical aortic arch. There is an embryonic aortic arch on each side and interruption of either the right or left arch at one or more points can explain the basis of the maldevelopment.

Normally, the aortic arch represents the persistence of the fourth branchial arch and some

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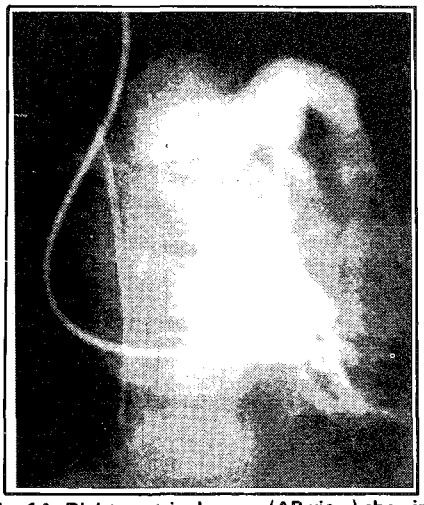

Fig. 1A Right ventriculogram (AP view) showing features of Tetralogy of Fallot.

Fig. 1C Aortic arch angiogram (AP view) showing its location in the cervical area.

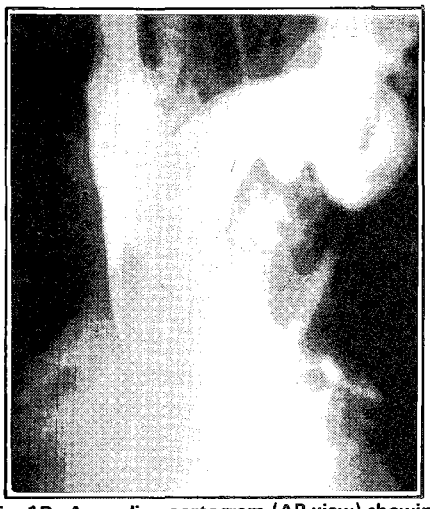

Fig. 1B Ascending aortogram (AP view) showing dilated left sided innominate artery. Aortic arch not seen.

Fig. 1D Ascending aortic arch angiogram (lateral view) showing the cervical aortic arch with the area of coarctation.

Note: The neck branches and the right subclavian artery were not well visualised in these still frames.

authors postulate that the cervical aortic arch may be derived instead from the second or third branchial arch. Fig. 2A shows the possibility of the development of the aortic arch from the second or third branchial arch. Fig. 2B shows the result of interruption of the left embryonic arch between the left external carotid artery and the right external carotid artery. A right aortic arch is formed and the first branch is the left innominate artery which gives rise to the left subclavian artery and the left common carotid artery.

The second branch is the right internal carotid artery, the right external carotid artery and the right subclavian artery. There is also a coarcted segment of the aortic arch. This case also illustrates the rare combination of Tetralogy of Fallot and coarctation of aorta. Embryologically, coarctation of aorta tends to occur in conditions with reduced ascending aortic flow during foetal life. To the contrary, Tetralogy of Fallot causes an increase in the ascending aortic flow during foetal life.

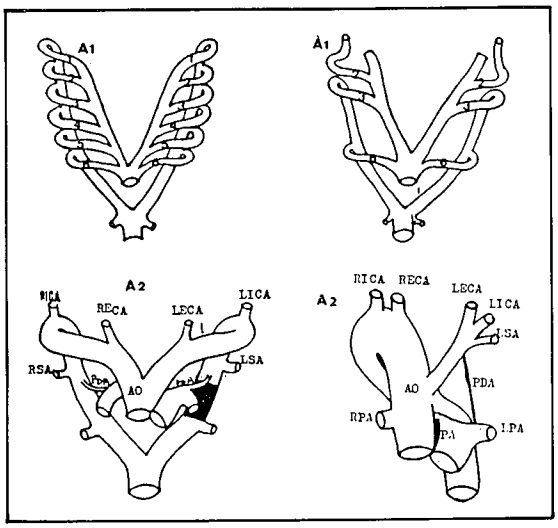

Fig. 2 Development of the cervical aortic arch. RICA = (R) internal carotid artery, RECA = (R) external carotid artery, LECA=(L) external carotid artery, LICA= (L) internal carotid artery, RSA= (R) subclavian artery, PDA= patent dustus arteriosus, AO= aorta, RPA=(R) pulmonary artery, LPA= (L) pulmonary artery. Areas in black indicate segments of regression.

Clinical Significance

From literature,[2] the presentation of the cervical aortic arch can be: patients are usually asymptomatic; a pulsating mass may be present in the neck with a murmur and thrill over the mass; femoral pulsation are most often normal or slightly diminished and with manual compression of the cervical arch, slowing of the pulse and decreases of the pulse pressure usually occurs; no significant murmur or thrill noted over the praecordium; X-ray may show a high mediastinal mass or compression of the barium filled oesophagus; aortography demonstrate the present of vascular anomaly.

The pulsatile mass in the neck above the clavicle may be mistaken for an aneursym of the subclavian artery, carotid or innominate artery or even for other neck swellings.

Surgery for this case would be staged starting with relief of the coarctation before total correction of the intracardiac abnormality.[3] Theoretically, relief of the coarctation would result in a drop in the pressure gradient across the right ventricular outflow tract and thus reducing the pulmonary blood flow. The surgeon should be prepared to create a form of a systemic-pulmonary shunt in anticipation of this problem.

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