Cervical aortic arch with aortic obstruction: report of two cases

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ABSTRACT The occurrence of aortic obstruction in patients with cervical arch is very rare. The clinical and angiocardiographic findings of two patients with this combination of defects are presented. One of these patients, in whom tricuspid atresia with reduced pulmonary blood flow was also present, had a successful resection of the obstruction together with construction of a Waterston shunt.

Cervical aortic arch is a very rare congenital anomaly. The malformation is usually isolated but may be associated with intracardiac defects. Aortic obstruction occurring between a cervical arch and the descending thoracic aorta has been recently reported. Our purpose in this paper is to describe two patients with the combination of cervical aortic arch and aortic obstruction. One of these patients, a 10-month-old infant, in whom tricuspid atresia with reduced pulmonary blood flow was present, had a successful resection of the obstruction together with construction of a Waterston shunt.

Case reports

Case 1
A 10-month-old infant was admitted to our hospital with a history of increasing cyanosis. A cardiac murmur was noted at birth. On clinical examination the baby appeared severely cyanosed, and had tachycardia and dyspnoea. A harsh systolic murmur was heard along the left sternal border. The left brachial pulse was prominent but the right brachial and the femoral pulses were absent. Blood pressure (160/100) could be measured in the left arm only. The ECG findings were consistent with right atrial and left ventricular hypertrophy. The QRS axis was $-10^\circ$. The chest radiograph showed cardiomegaly with decreased pulmonary vascularity. Cardiac catheterisation and angiography (fig 1a, b) showed tricuspid atresia with normally related great arteries and reduced pulmonary blood flow. There was a right-sided cervical aortic arch, and severe aortic obstruction was present between the arch itself and the descending thoracic aorta. The first branch to arise from the arch (fig 2) was a left brachiocephalic artery which gave rise to the left common carotid artery and the left subclavian artery. A hypoplastic right common carotid artery arose from the arch just above the obstruction.

A right thoracotomy was performed through the third intercostal space. The aortic arch was dissected free and followed through the mediastinal pleura upwards above the level of the right clavicle. The anatomical findings were in keeping with the clinical data presented. The right subclavian artery was found to arise from the thoracic aorta, which was retro-oesophageal, and continued as a left-sided descending aorta. The right common carotid artery was ligated at its origin and divided. The stenotic segment of the descending thoracic aorta was resected between clamps, and an end-to-end anastomosis was performed. A Waterston shunt 3 mm in size was then constructed. Good femoral pulses and a continued anastomotic murmur were present at the end of the operation. No neurological complications were observed during the postoperative course, which was uneventful. The patient remains in good general condition two years and a half years after the operation.

Case 2
A 12-year-old girl was referred to our hospital for evaluation of a pulsating mass in the left...
supraclavicular fossa. She was known to have had a systolic thrill with “abnormal” arterial pulsations in the left side of the neck since the age of 5 years. Cardiac catheterisation performed at the age of 8 years showed a left-sided cervical aortic arch. At that time a 1×1 cm pulsating mass was present in the left supraclavicular fossa. The chest radiograph (fig 3a) was unremarkable. She was asymptomatic, and the pertinent findings were limited to the cardiovascular system. The left brachial pulse was prominent but the right brachial and femoral pulses were barely palpable. Systolic blood pressure in the left arm (100 mmHg) was 50 mmHg greater than in the right arm and legs. A pulsating mass was present in the left supraclavicular fossa in the angle between the sternocleidomastoid muscle and the upper border of the clavicle. The pulsations were arterial in quality and timing and a systolic thrill was palpable over the mass. A grade 3/4 ejection systolic murmur loudest at the second left intercostal space radiated up into the mass. The ECG was normal. Chest radiography at this time showed a prominent aortic knob situated high above the left clavicle (fig 3b). Cardiac catheterisation showed no intracardiac abnormalities. Angiography confirmed the previous diagnosis of left cervical aortic arch (fig 4a, b). The ascending aorta gave rise through a single branch to both the right and left common carotid arteries and to the left subclavian artery (fig 5). A moderate narrowing was present between the aortic arch and the descending thoracic aorta, which showed a marked poststenotic dilatation. The right subclavian artery
arose from the descending aorta. Surgical treatment was suggested but it was refused by the parents of the patient who is now followed up in the outpatient clinic.

Fig 2  Case 1. Diagram summarising the key angiographic findings. $AAo$=ascending aorta; $DAo$=descending aorta; $LBA$=left brachiocephalic artery; $LSA$=left subclavian artery; $LCC$=left common carotid artery; $RCC$=right common carotid artery; $RSA$=right subclavian artery; other abbreviations as in fig 1.

Discussion

Surgical treatment of patients with cervical aortic arch is commonly indicated for relief of symptoms resulting from compression of the trachea and oesophagus or for correction of associated intracardiac defects. Successful repair of rare associated anomalies such as aneurysm of the cervical arch itself or of the descending thoracic aorta and aortic obstruction have also been reported.

Aortic obstruction occurring between the cervical arch and the descending thoracic aorta was present in both our cases. This was an unexpected finding in our first case, a 10-month-old infant, who underwent cardiac catheterisation because of the presence of severe cyanosis. Tricuspid atresia with reduced pulmonary blood flow was found to be associated with a right cervical aortic arch. Successful end-to-end resection of the obstruction and construction of a Waterston anastomosis were performed at the same time. The second patient presented with a long history of a pulsating mass in the left supraclavicular fossa. The interesting feature in case 1 is the occurrence of severe aortic obstruction in a cyanotic congenital heart defect in the absence of any subvalvar or valvar aortic stenosis. The obstruction could well be related to the abnormal development of the arch which occurs in patients with cervical aortic arch.

In case 2 the rapid development of the cervical mass was thought to be related to the progressive dilatation of the descending thoracic aorta distal to the obstruction. Surgical treatment in this instance was advised to avoid the possible development of an aneurysmal dilatation with compression of the brachial plexus, stellate ganglion, recurrent nerve and even jugular or innominate vein and apex of the lung. Traumatic rupture or perforation resulting from its exposed position was another threat. Resection of the stenotic segment of the cervical arch and of the post-stenotic dilatation would have been a relatively simple procedure since both the common carotid arteries arose from the ascending aorta proximal to the obstruction. However, surgical treatment was refused.

The origin of the head vessels was unusual in both our cases. In the first patient, who had a right aortic arch, the right subclavian artery was the last branch to arise from the descending thoracic aorta. This is not in keeping with the findings reported by Mullins et al in which the last branch of the thoracic aorta was always the subclavian artery contralateral to the side of the aortic arch. A single origin of both the right and left common carotid arteries and a thoracic aorta descending anterior to the oesophagus and on the same side of the aortic arch were the peculiar findings in the second case.

These findings show that the pattern of origin of the brachiocephalic arteries is not always a predictable pattern, and therefore this cannot be used as a clue to explain the morphogenesis of a cervical aortic arch.

Indeed it is still uncertain whether the cervical position of the aortic arch is the result of persistence of the embryonic third arch, or whether on the contrary, the aortic arch although normally derived is retained in the cervical region.

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Fig 3  Case 2. Postero-anterior chest radiographs showing (a) poorly defined aortic knob at the age of 8 years and (b) prominent high position of left aortic arch at the age of 12 years.

Fig 4  Case 2. (a) Postero-anterior and (b) left anterior oblique frames of a left ventriculogram showing a left-sided cervical aortic arch with (b) aortic obstruction (arrowed) AAo=ascending aorta; DAo=descending aorta.
Fig 5  Case 2. Diagram summarising the key angiographic findings. Abbreviations as in fig 2.

References