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Brief Report

Left cervical aortic arch associated with multiple vascular anomalies

José Luiz Balthazar Jacob

Department of Hemodynamic and Interventional Cardiology of the Instituto de Moléstias Cardiovasculares de São José do Rio Preto, São Paulo, Brazil

Abstract Cervical aortic arch is a rare anomaly occasionally associated with other cardiovascular abnormalities. We present a case of tortuous left cervical aortic arch associated with hypoplastic transverse arch, coarctation of the aorta, and right brachiocephalic arteries arising below the coarctation and stenotic origin of the left subclavian artery. These multiple anatomic anomalies, which are associated in our case, have not been described in a single patient previously.

Keywords: Congenital anomalies of the aorta; congenital heart disease; vascular anomalies

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Cervical aortic arch is a rare anomaly in which the aortic arch is situated above the clavicle.[1] The exact cause of this anomaly is uncertain, but embryologic errors of the second, third, and fourth dorsal arches may be responsible for this malformation.[1] Cervical aortic arch is occasionally associated with other cardiovascular abnormalities.[1–3] We report a patient with left cervical aortic arch who had hypoplastic transverse arch, stenosis of origin of the left subclavian artery, and aortic coarctation and anomalous origin of the right brachiocephalic arteries. Transseptal technique was mandatory for catheterisation of the left ventricle and ascending aorta.

Case report

An 8-year-old girl was referred to our Cardiologic Center for evaluation for percutaneous treatment of an atypical coarctation of the aorta. The physical examination revealed her blood pressure to be 130/80 millimetres of mercury in the left upper, 90/60 in the right upper, and 90/60 in both lower limbs. A loud systolic murmur was audible in the right and left upper chest, in the suprasternal notch, and in the left side of the neck with an accompanying palpable thrill and pulsatile vessel in this area. Electrocardiogram showed left ventricular hypertrophy. Radiologic study of the chest showing the left aortic arch above the clavicle suggested the presence of a left cervical aortic arch. Transthoracic echocardiography revealed features of the left cervical aortic arch, tricuspid aortic valve, and severe aortic coarctation. The conclusion of the magnetic resonance imaging performed in another hospital described severe coarctation of the aorta, anomalous origin of the right brachiocephalic arteries, and large and tortuous vessels above aortic coarctation, but the images were not sent to us. Cardiac catheterisation was attempted in the other centre of cardiology using the right brachial and right femoral arterial approach, but was inconclusive because the ascending aorta was not catheterised.

We performed cardiac catheterisation using transseptal technique to reach the left ventricle and ascending aorta. Pressure in the ascending aorta was 172/100 millimetres of mercury. The angiographic study revealed that the right carotid was the first branch of the ascending aorta, followed by the left carotid. Both arteries emerge proximal to the hypoplastic transverse arch. The left cervical aortic arch was very tortuous, but not aneurysmatic (Fig 1a and 1b). The left subclavian artery was stenotic

Correspondence to: Dr J. L. Balthazar Jacob, MD, PhD, Rua Castelo D'Água 3030, CEP 15015-210, São José do Rio Preto, São Paulo, Brazil. Tel: 55 17 32034010; Fax: 55 17 32034000 R 239; E-mail: jljacob@cardiol.br; josejacobliteratura@netsite.com.br

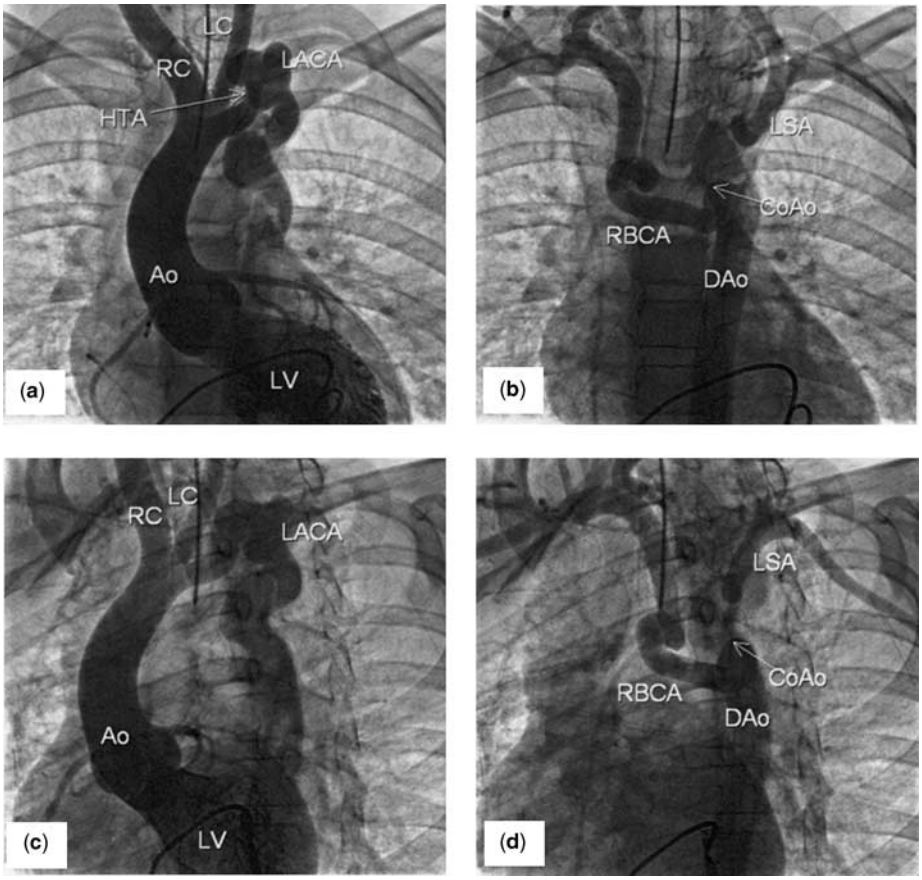

Figure 1.
*(**a**) Angiogram in the LV in the frontal view shows the Ao with an HTA (arrow). RC is the first vessel arising from the aortic arch, and the LC is the second vessel. Left cervical aortic arch shows a very tortuous pathway. (**b**) LSA emerges above the CoAo (arrow). RBCA emerge from the DAo below the coarctation. DAo shows late opacification. (**c**) Angiogram in the left anterior oblique view confirms the tortuous LACA. Right and left carotid arteries are the first and second vessels, respectively, arising from the aortic arch. (**d**) LSA emerges above the CoAo (arrow) and is stenotic in origin. Anomalous origin of the RBCA below the coarctation is visible. DAo shows late opacification due to severe coarctation (arrow) (LV = left ventricle; Ao = ascending aorta; HTA = hypoplastic transverse arch; RC = right carotid artery; LC = left carotid artery; LACA = left aortic cervical arch; LSA = left subclavian artery; CoAo = coarctation of the aorta; RBCA = right brachiocephalic arteries; DAo = descending aorta).*

in origin with late and slow opacification of the artery. Below the left subclavian origin there was severe aortic coarctation and the right brachiocephalic arteries emerge out of the descending aorta below the coarctation (Fig 1c and 1d). Surgical treatment was suggested but was refused by the parents.

Discussion

The cervical aortic arch is a rare anomaly that represents persistence of the embryonic third arch, with regression of the fourth arch on that side.[3,4] Coarctation of the aorta is associated in 10% and aneurysm of the cervical arch in 20% of reported cases.[2,3,5] Obstruction of the cervical aortic arch is generally in the transverse segment, and structural abnormalities of brachiocephalic arterial branching are relatively common when the aortic arch is located in the neck.[2–4] Discrepancy of arterial blood pressures between the upper and lower limbs described in our case was previously reported.[2,3]

Direct aortic origin of the right and left carotid arteries at the same side of the arch is common, but anomalous origin of the right brachiocephalic arteries of the descending aorta below the coarctation is a conspicuous feature in our patient.

It is usual for the cervical aortic arch not to produce symptoms, but some patients may present with dyspnoea, dysphagia, or recurrent infection due to compression of the trachea or oesophagus by a vascular ring.[3,4] Our patient was asymptomatic.

Resonance magnetic imaging is particularly useful in accurately defining this complex anomaly.[4] Many features not described in the magnetic resonance

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imaging in this case probably were due to unsuitable interpretation in the other centre of cardiology.

The tortuous pathway of the left cervical aortic arch with associated aneurysm has been described previously.[5] To our knowledge, the very tortuous pathway of the left cervical aortic arch with hypoplastic transverse arch and anomalous origin of the right brachiocephalic arteries below the aortic coarctation present in our patient has not been described previously in the literature. This report shows the importance of the atrial transseptal technique for catheterisation of the left ventricle and ascending aorta , which was impossible by the other catheterisation approach due to associated anomalies.

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