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Brief Report

Left cervical aortic arch with proximal obstruction

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Abstract Cervical aortic arch is a rare congenital anomaly. We present a case of left-sided cervical aortic arch, found in a patient with mitral regurgitation, mitral stenosis, and a regurgitant bicuspid aortic valve. There was atypical obstruction proximally within the arch. The obstructive segment was resected, and corrected by performing an end-to-side anostomosis. Mitral valvoplasty was performed in the same surgical procedure. To the best of our knowledge, a cervical arch has not previously been described with such atypical obstruction, and in association with multiple lesions involving the left heart.

Keywords: Aortic arch; aortic coarctation; mitral regurgitation; mitral stenosis; aortic regurgitation

THE CERVICAL AORTIC ARCH IS A RARE CONGENITAL anomaly with a prevalence of less than 1 in each 10,000 live births.[1] With this anomaly, the aorta is elongated superiorly and is located cervically above the clavicle. It is rare to find the cervical aorta in association with coarctation and anomalies involving the left heart. In most cases, it is an isolated condition, and is usually clinically silent. We present here a patient in whom a left-sided cervical aortic arch was obstructed proximally in the presence of multiple malformations involving the left side of the heart.

Case report

An 18-year-old male patient had been referred to a cardiac centre complaining of palpitations and fatigue. Cardiac catheterisation revealed that pressures in the left carotid artery and ascending aorta were 190/90 mmHg, with corresponding pressures in the right carotid artery and transverse aorta being 120/70, and in the right ventricle 80/0–6 mmHg. A peak-to-peak pressure gradient of 70 mmHg was measured between the left and right carotid arteries, and it was noted that the left carotid artery originated from ascending aorta as its first branch. He was referred to our centre for surgical correction.

On physical examination, a pulsatile mass was noted in the right supraclavicular region, the upper and femoral pulses were weak, and systolic blood pressure as measured in the arms was 100 mmHg as assessed by palpation of the radial pulses. A loud systolic murmur was heard in the right upper chest and the right side of the neck, with an accompanying palpable thrill, with a systolic regurgitant murmur of grade 3/6 heard at the apex, with transmission to the left axilla. Chest radiography showed a high aortic knob, with a right apical mass (Fig. 1). Electrocardiography showed left ventricular hypertrophy, with inversion of the T waves in the left precordial leads.

Transthoracic echocardiography revealed an obstructed cervical aortic arch, moderate mitral regurgitation, mild mitral stenosis, a bicuspid and mildy regurgitant aortic valve, and pulmonary hypertension. Magnetic resonance imaging confirmed the cervical location of the aortic arch, revealing stenosis proximal to the origin of the right carotid artery. The origin of the brachiocephalic arteries was also shown to be abnormal. The left common carotid artery was the first branch from the ascending aorta, originating proximal to the lesion narrowing the arch. The right carotid artery arose as the second artery, distal to the obstruction, with an aberrant right subclavian artery arising from the transverse aorta, and the left subclavian artery

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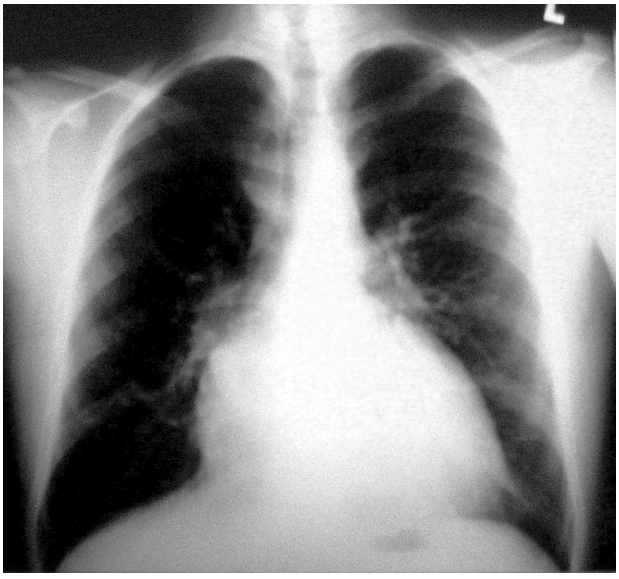

Figure 1.
Chest radiograph showing an abnormal shadow in the right clavicular region.

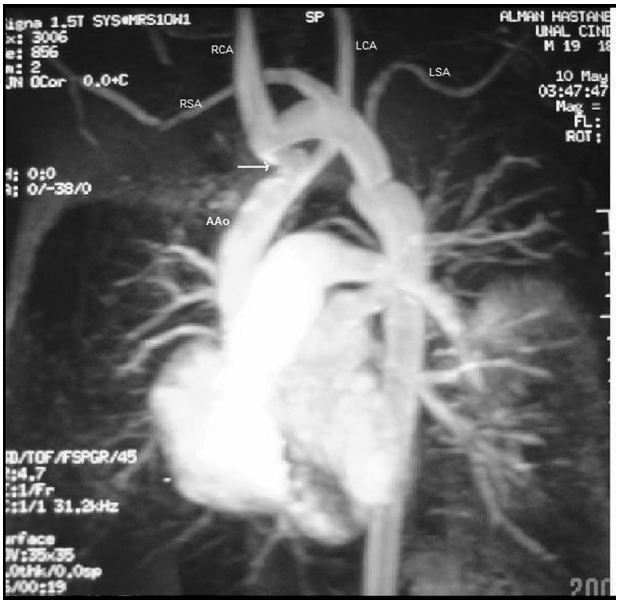

Figure 2.
Magnetic resonance angiography showing that the first vessel arising from the aortic arch is the left carotid artery. There is a narrow segment between the left and right carotid arteries, aberrant origin of the right subclavian artery from the transverse aorta, with the left subclavian artery arising from the distal part of the transverse aorta. AAo: ascending aorta; RCA: right carotid artery; LCA: left carotid artery; RSA: right subclavian artery; LSA: left subclavian artery.

originating from the distal part of the transverse aorta. There was a long narrowed segment between the origins of the left and right carotid arteries (Fig. 2).

Surgery was performed via a median sternotomy. The obstructive segment was resected and corrected by end-to-side anostomosis. Stenosis was noted at the supero-posterior end of the zone of apposition between the leaflets of the mitral valve, with elongation of the mural leaflet. We performed a DeVega valvoplasty as part of the operative procedure. The repair was successful, with no perioperative complications. Histopathologic examination of the resected specimen showed severe stenosis of the aorta, with intimal and medial thickening with myxoid degeneration. At follow-up the patient was well. The blood pressure was now measured at 110/80 mmHg in all limbs. Echocardiographic examination showed mild residual stenosis proximally within the arch, with mild aortic and mitral regurgitation.

Discussion

The cervical aortic arch, regardless of obstruction or associated lesions, is a rare anomaly in itself.[1–8] It is generally thought that such a cervical location represents persistence of the embryonic third arch, with regression of the fourth arch on that side. Alternatively, it is speculated that the cervical arch is simply a high fourth arch, with failure of normal descent.[2–5,9] Although the cervical aortic arch is slightly more commonly found in right-sided position, when obstructed, or showing aneurysmal transformation, the arch is mostly left-sided.[3]

It is usual for the cervical arch not to produce symptoms, but some patients may present with dyspnoea, dysphagia, and recurrent infection due to compression of the trachea or oesophagus by a vascular ring, and/or discrepancy of arterial blood pressures between the upper and lower limbs. In our patient, the symptoms of palpitation and fatigue were more probably due to the mitral regurgitation.

The cervical aortic arch may be associated with coarctation, with such a complication found in one-tenth of reported cases.[2–4] Such obstructions, however, have previously been described only in the transverse or distal arch. We have found no previous description of narrowing at the proximal arch. We hesitated at describing our obstruction in the proximal arch rather than the ascending aorta, but opted for the proximal arch because the narrow segment was distal the first brachiocephalic vessel.

Structural abnormalities of brachiocephalic arterial branching are relatively common when the aortic arch is located in the neck.[2,3,6–8] Direct aortic origin of the external and internal carotid arteries on the same side of the arch are particularly common, as in our patient. Although an isolated anomaly in most cases, about one-third of patients have associated lesions, such as tetralogy of Fallot, ventricular septal defect, or double outlet right ventricle.[9] We have found only one report, however, of anomalies

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involving the left heart, specifically with mitral and aortic regurgitation and pseudocoarctation.[8] Thus, although various congenital anomalies, including cardiovascular malformations, occasionally complicate the cervical aortic arch, we are unaware of previous reports of a cervical arch associated with proximal obstruction and multiple lesions involving the left heart.

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