Congenital subclavian steal syndrome with multiple cerebellar infarctions caused by an atypical circumflex retroesophageal right aortic arch with atretic aberrant left subclavian artery

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A right-sided aortic arch is a rare anomaly with an incidence of 0.1% worldwide and is usually associated with a mirror image of all supra-aortic branches or an aberrant left subclavian artery. The latter is often associated with a Kommerell diverticulum, although it can rarely be hypoplastic or atretic and lead to congenital subclavian steal. In most patients, the situation is well-tolerated. In this report, we present a case of subclavian steal syndrome with multiple cerebellar infarcts in a patient with an atypical right-sided aortic arch and an atretic aberrant left subclavian artery arising from a left-sided descending thoracic aorta. (J Vasc Surg 2014;60:776-9.)

CASE REPORT

A 60-year-old man with a left-sided subclavian steal syndrome was referred to us for a subclavian reconstruction after an episode of vertigo, headache, temporary right-sided ataxia, and hemihypesthesia 30 days earlier. A magnetic resonance tomography showed an acute right cerebellar infarction, multiple older bilateral cerebellar infarctions (more pronounced on the left), and diffuse ischemic atrophy of the cerebral stem with complete lack of ischemic lesions of the cerebral hemispheres (Fig 1, *A*). The cerebral arterial anatomy presented both vertebral arteries forming the basilar artery with a complete circle of Willis.

A magnetic resonance angiography revealed a circumflex retroesophageal right-sided aortic arch (RAA) with an occlusion of an aberrant left subclavian artery (aLSA) (Fig 1, *B* and *C*). A retrograde perfusion of the left vertebral artery and a difference in arterial pressure of 30 mm Hg between the upper extremities were documented.

A ligamentum arteriosum connected the left pulmonary artery with the atretic origin of the LSA (Fig 2, *A-C*). Furthermore, a disturbed flow in the right vertebral artery after induced reactive hyperemia of the left arm could be demonstrated, showing the direct influence of increased perfusion of the left arm upon the entire vertebral circulation. Cardiogenic embolism was ruled out with a transesophageal echocardiogram, and 72-hour electrocardiograph monitoring showed a stable sinus rhythm.

The findings described above were compatible with a chronic vertebrobasilar ischemia. Microemboli originating from the mechanical heart valve or from the right subclavian artery aneurysm correlated to neither the distribution of ischemic lesions predominantly in the left cerebellar hemisphere nor to the diffuse cerebellar ischemic atrophy.

Four months earlier, the patient had another episode of presyncope with vertigo and nausea, which were repeated five times within 3 hours. An echocardiogram also revealed normal findings in this case. His medical history often included episodes of vertigo and a fractured hip after a fall 4 years earlier. At the age of 34 years, a difference in arterial pressure between both arms led to the diagnosis of a LSA occlusion and a high grade insufficiency of a bicuspid aortic valve, which was replaced with a mechanical valve. The subclavian occlusion, well-tolerated at that time, was left untreated. The patient had been taking oral anticoagulants since that time.

We performed a transverse supraclavicular incision carried through the clavicular head of the sternocleidomastoid muscle and dissected the left common carotid artery down to its ostium from the aortic arch (Fig 3, *A*). The LSA was found much deeper and dorsal to the left carotid artery (Fig 3, *B*). The proximal segment of the LSA exhibited a wineglass-shaped hypoplasia ending in a string-like atretic segment at the ostium from the descending aorta. The string-like segment was ligated, and the normal segment of the subclavian artery with the ostium of the left vertebral artery was separated with cranioventral rotation (Fig 3, *C*) to allow a dorsolateral anastomosis with the posterior wall of the left carotid artery (Fig 3, *D*).

A computed tomography angiography confirmed the technical success of the procedure (Fig 3, *E* and *F*). The patient had

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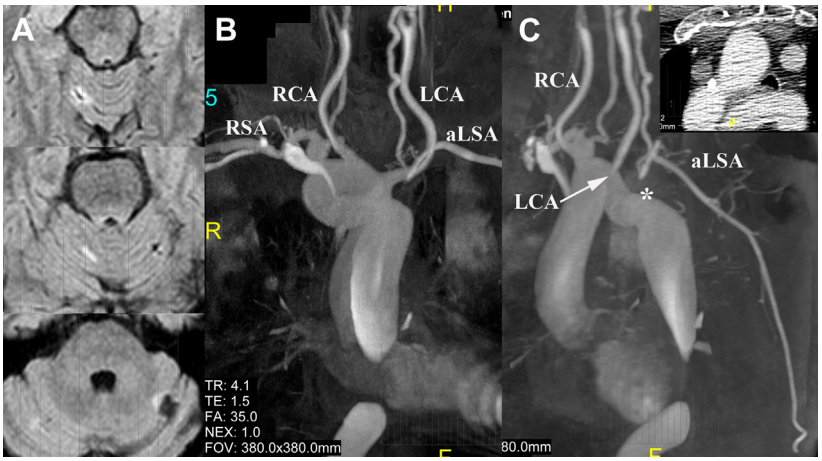

Fig 1. A, Magnetic resonance tomography scan of the brain with multiple infarctions in both cerebellar hemispheres, more pronounced on the left side. Furthermore, diffuse chronic ischemic alterations with atrophy of the brain stem could be demonstrated. In stark contrast, both brain hemispheres were completely free of any pathological findings. The findings described above, combined with the functional duplex-ultrasound examination, were compatible with a chronic vertebrobasilar ischemia. **B** and **C,** Magnetic resonance angiography of the thoracic aorta showing a circumflex right-sided aortic arch (anteroposterior **[B]** and left lateral view **[C]**). The first supra-aortic branch was the left common carotid artery (*LCA*) followed by the right common carotid artery (*RCA*) and the right subclavian artery (*RSA*; with a 17-mm aneurysmatic proximal segment). An aberrant left subclavian artery (*aLSA*), proximally occluded, originated from the proximal descending aorta (*). The aortic arch had a retroesophageal segment (*inset*) without a pronounced diverticulum of Kommerell.

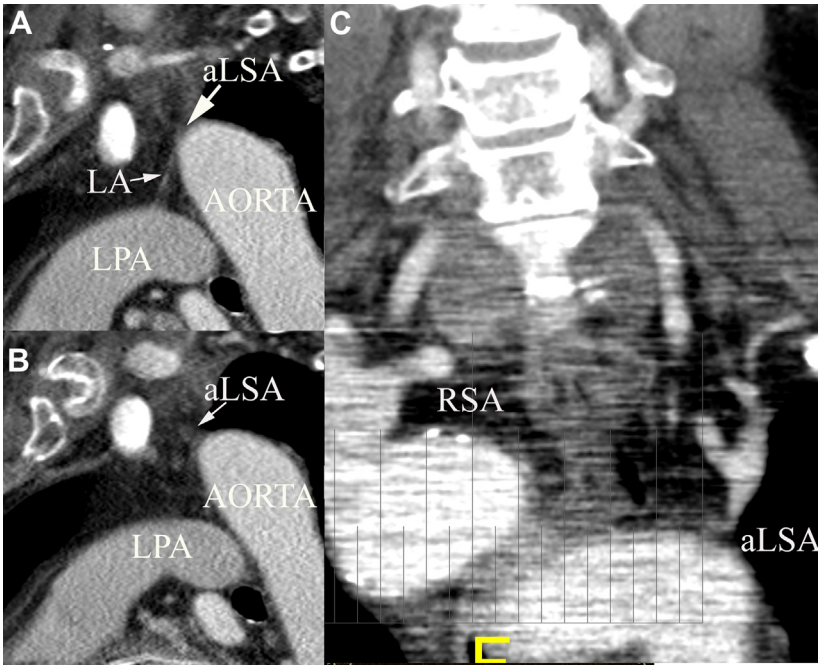

Fig 2. A, Computed tomography angiography showing a ligamentum arteriosum (*LA*) between the left pulmonary artery (*LPA*) and the origin of the aberrant left subclavian artery (*aLSA*). **B,** The origin of the aLSA from the descending aorta showed no pronounced diverticulum of Kommerell. **C,** Anteroposterior computed tomography reconstruction of the proximal aLSA with the ostium of the vertebral artery. The proximal segment of the right subclavian artery (*RSA*) is also visible.