

The right aortic arch with mirror-image branching of brachiocephalic arteries and aplasia of the left brachiocephalic trunk: surgically cured rare cause of cerebrovascular insufficiency.

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Abstract

A clinical case with rare anomaly: right aortic arch with mirror-image branching of brachiocephalic arteries and aplasia (isolation) of the left brachiocephalic trunk, causing cerebrovascular insufficiency and left arm claudication, is described. Despite the clear neurologic manifestations of cerebrovascular insufficiency and absence of the pulse on the left arm, the pathology was not diagnosed until 35 years of age. The anomaly was diagnosed and surgically cured in the Department of Vascular Surgery of Vilnius University Emergency Hospital. Using transsternal median thoracotomy approach, 12 mm diameter PTFE vascular graft, arising from the ascending aorta and connected to the angle, made of branching left common carotid and left subclavian arteries, was implanted into the position of missing left brachiocephalic trunk. 3 years after surgery patient feels healthy and is working. To our knowledge, this is the first reported case of surgical replacement of missing left brachiocephalic trunk in the right-sided aortic arch with mirror-image branching of brachiocephalic arteries.

Key words: Anomalies of the aortic arch and its branches, Surgery of brachiocephalic arteries, Cerebrovascular insufficiency, Subclavian steal syndrome.

Introduction

Right aortic arch with mirror-image branching of brachiocephalic arteries is a rare clinical and pathological entity [1,2]. Only few cases are reported in the literature with this anomaly and isolation of the left brachiocephalic trunk [3,4]. It is unusual to have right-sided aortic arch without coexisting cardiac congenital defects [2,4,5].

B. Singh and coauthors [3] described a female, 36 years of age, who had vertigo, dizziness and left hand claudication. The cause of these symptoms was right-sided aortic arch with isolation of the left brachiocephalic trunk, steal through the left vertebral artery into the left subclavian artery. Our case is very similar to that case.

Case report

35 year old female was seeking medical help because of disabling dizziness, vertigo episodes, syncopal episodes, fast tiredness of the left arm and hand. She had these symptoms from the childhood. Physical work with the left hand aggravated the symptoms. She was not able to work for several years due to these symptoms. Absence of the pulse on the left arm was determined and the patient was referred to vascular surgeon. At examination no external body anomalies were detected. Absence of the pulse on the left arm was confirmed. There was no pulse on the left common carotid artery as well. Other peripheral pulses were normal. The systolic arterial pressure difference between arms was 60 mm Hg: 120 on the right and 60 on the left side. There was only right hemisindromus with elevation of tendon reflexes on the right side, no other neurological symptoms were found at rest, but postischemic hyperemia test of the left arm provoked pronounced dizziness, ataxia, horizontal nystagmus. Duplex studies showed low, poststenotic flow in the left brachial artery, retrograde flow with subclavian steal in the left vertebral artery and very slow antegrade flow in the left common and internal carotid arteries. Right common and internal carotid arteries, as well as right vertebral and subclavian arteries, had normal direction and velocity of blood flow. Anomaly of aortic arch branches or Takayasu disease was suspected and patient was admitted to the Department of Vascular Surgery of Vilnius University Emergency Hospital.

Angiography revealed the right-sided aortic arch with mirror-image branching of brachiocephalic arteries and aplasia (isolation) of the left brachiocephalic trunk (fig. 1, 2).

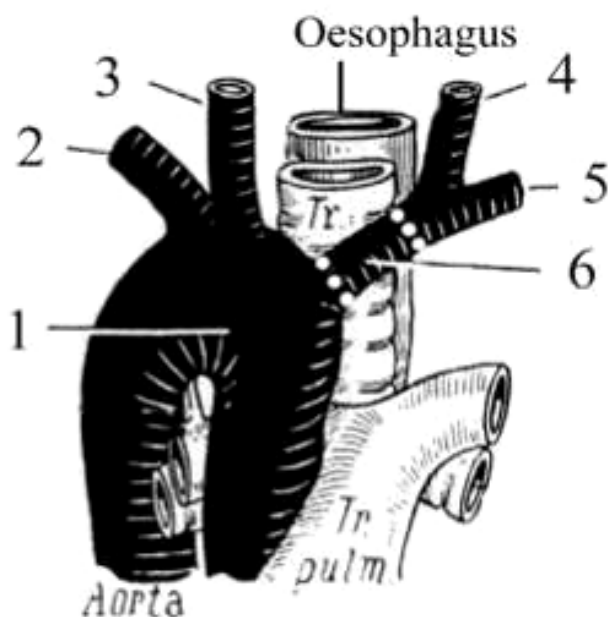


Fig. 1

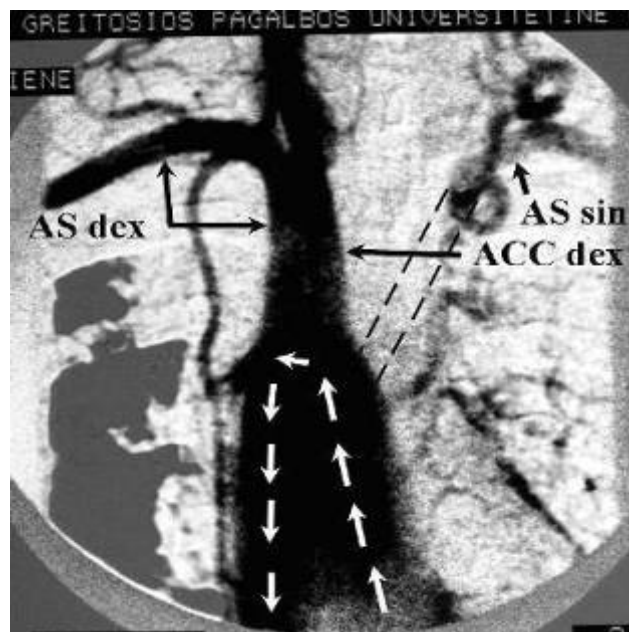


Fig. 2 A

Fig.1. Schematic diagram of the right aortic arch with right-sided aorta and mirror-image branching of brachiocephalic arteries. Segment 6, excluded with white dotted lines, was absent in our patient. Tr – trachea, 1- aortic arch, 2 – right subclavian artery, 3 – right common carotid artery, 4 – left common carotid artery, 5 – left subclavian artery, 6 – left brachiocephalic trunk.

Fig. 2 A. Aortic arch angiogram. White arrows indicate the direction of aortic arch. Right common carotid and right subclavian arteries arise from the aortic arch as separate trunks (they overlap in this angiographic view). Left subclavian artery is filled with contrast through the collaterals (internal thoracic artery, thyrocervical trunk). Missing left brachiocephalic trunk is depicted by broken black lines. AS – a. subclavia, ACC – a. carotis communis.

The left cerebral hemisphere was supplied by the blood through the very long and complicated route: the blood was flowing up by the right vertebral artery, some of the blood was distributed in the territory of basilar artery (fig. 2 B), remainder of it flows down through the left vertebral artery into the left subclavian artery and into the left common carotid artery (fig. 2 C).

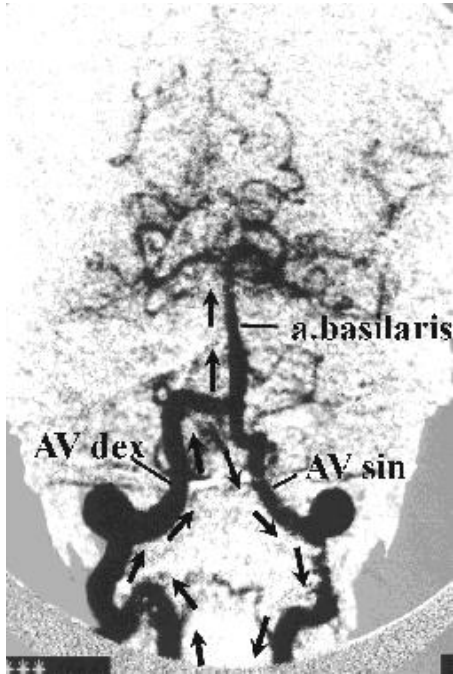


Fig. 2 B

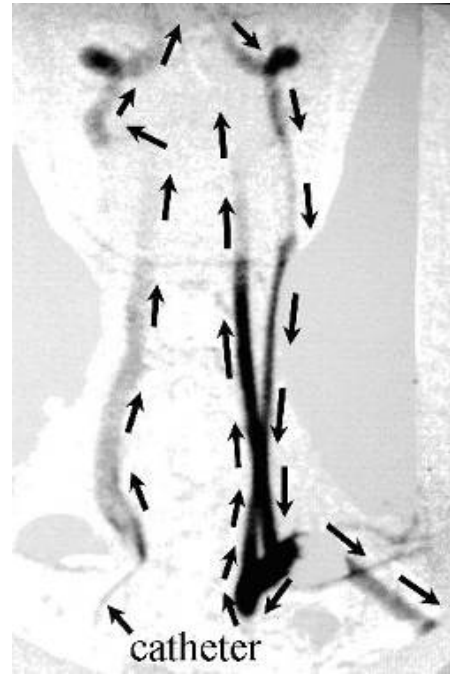


Fig. 2 C

Fig. 2 B. Distribution of blood flow in vertebrobasilar territory. Blood is flowing up by the right vertebral artery and going down by the left vertebral artery. Some of the blood is distributed in the territory of basilar artery. Arrows indicate the direction of blood flow. AV – a. vertebralis.

Fig. 2 C. Angiographic appearance of blood steal from the left vertebral artery into the left subclavian and left common carotid arteries. Arrows indicate the direction of blood flow and sequence of filling of these arteries by contrast material. Catheter is inserted into the right vertebral artery. Blood flows up by the right vertebral artery, goes down by the left vertebral artery and only then goes up by the left common carotid artery. Part of the blood goes into the left arm by the distal left subclavian artery.

Symptoms were corresponding to this anomaly and logical operation, grafting of missing left brachiocephalic trunk, was suggested for patient.

2001 01 16 the patient was operated. Using median sternotomy approach, 12 mm diameter PTFE vascular graft was implanted into the position of missing brachiocephalic trunk: proximal end of the graft was connected to the ascending aorta and the distal end of it was sutured to the angle made by left common carotid and left subclavian arteries (fig. 3).

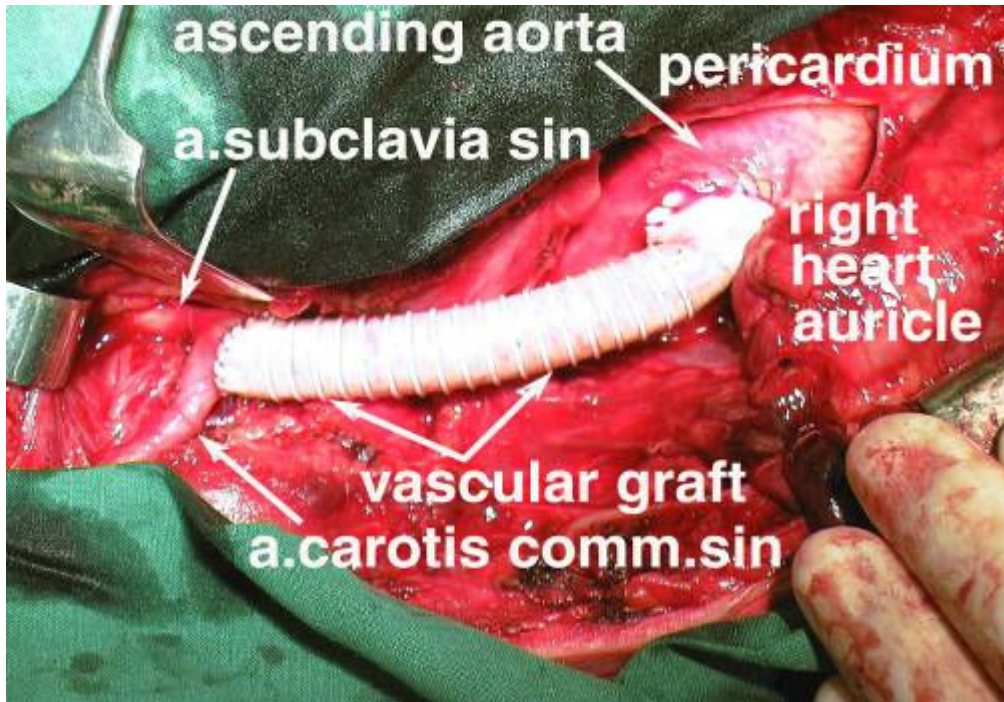


Fig. 3. Operative photograph, made after reconstruction. 12 mm diameter PTFE ringed vascular graft is implanted into the position of missing left brachiocephalic trunk. The proximal end of the graft is connected to the ascending aorta and the distal end – to the angle made by left common carotid and subclavian arteries.

Postoperative course was smooth, except of headache, characteristic for cerebral hyperperfusion syndrome, which dissappeared at the end of hospital stay. Postoperative duplex and transcranial doppler studies showed elevated blood flow velocities over the normal values in the left common and internal carotid arteries and in the left middle cerebral artery. This can be explained by undeveloped autoregulative mechanism of cerebral blood flow in the left hemisphere, which had congenital insufficient blood flow. The direction and velocity of blood flow was normal in both vertebral arteries, both subclavian arteries, both common and internal carotid arteries. On the 7 –th day patient was discharged from the hospital.

Patient was examined every 6 months. After 6 months patient feels healthy, all symptoms disappeared. Blood flow in the left internal carotid and left middle cerebral arteries came down to normal. Follow – up after 1 year: feels healthy, started to work. After 3 years: healthy, working.

Discussion

Our patient had a rare anomaly: right-sided aortic arch, right sided aorta, mirror-image branching of brachiocephalic arteries and aplasia of the left brachiocephalic trunk. Duplex blood flow studies suggested this pathology, however standard angiography clearly revealed the complex vascular pathology and the way, the left cerebral hemisphere gets the blood supply. The operation performed was logical and curative. There were no congenital cardiac defects detected in our patient, what is unusual in this anomaly.

References:

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