Received: 2015.08.31
Accepted: 2015.09.18
Published: 2016.02.23

Authors' Contribution:

- A** Study Design
- B** Data Collection
- C** Statistical Analysis
- D** Data Interpretation
- E** Manuscript Preparation
- F** Literature Search
- G** Funds Collection

Right Aortic Arch with Aplasia of the Left Brachiocephalic Trunk Presented as Systolic Blood Pressure Difference Between Upper Extremities

Anna Babińska^{ABCEFG}, Wojciech Wawrzynek^{ACD}, Karolina Kukawska-Sysio^{ACD}, Jarosław Skupiński^{FG}, Patrycja Nowak^{FG}

Department of Diagnostic Imaging, Hospital of Traumatology, Piekary Śląskie, Poland

Author's address: Anna Babińska, Department of Diagnostic Imaging, Hospital of Traumatology, Bytomska 62 Str, 41-940 Piekary Śląskie, Poland, e-mail: annababinska88@gmail.com

Background:

The right aortic arch with mirror-image of branching arteries without coexisting congenital heart disease is a very rare anomaly.

Case Report:

We report a case of the right-sided aortic arch with aplasia of the left brachiocephalic trunk in a 64-year-old woman, presenting difference in systolic blood pressure between upper extremities. The history of the patient and angio-CT findings were described and visualized with images.

Conclusions:

The knowledge of vascular variations is important for the clinical and therapeutic aspects.

MeSH Keywords:

Aortic Arch Syndromes • Cardiovascular Abnormalities • Subclavian Steal Syndrome

PDF file:

<http://www.polradiol.com/abstract/index/idArt/895826>

Summary

Background

The extensive use of diagnostic imaging, especially computed tomography for most various clinical problems, has increased the number of incidentally found rare vascular anomalies. Despite the lack of clinical symptoms, uncommon arterial variations could be important for diagnostic or therapeutic issues.

The right aortic arch is divided into three major sub-groups classified according to branching patterns: (1) with an aberrant left subclavian artery, which is the most common; (2) with mirror-image branching of the major arteries (left brachiocephalic trunk, right common carotid artery, right subclavian artery); and (3) with the left subclavian artery isolated from the arch [1,2].

We report a case of the right aortic arch with mirror-image branching of the major arteries and aplasia of the left brachiocephalic trunk, without accompanying congenital cardiac defect, with left subclavian steal syndrome through vertebral artery and collateral circulation extending from the descending aorta, which makes our configuration extremely rare. To the best of our knowledge, this is the

first reported case with this type of aortic anomaly coexisting with collateral circulation through the posterior intercostal artery.

Case Report

A 64-year-old woman with a significant difference in systolic blood pressure and pulse asymmetry of the upper extremities was consulted by a cardiologist and after ultrasound Doppler study of carotid and vertebral arteries was referred to further diagnostics due to a suspicion of the left subclavian steal syndrome. Outpatient ultrasound imaging showed significant asymmetry of carotid arteries and reversed blood flow in vertebral arteries.

Patient underwent CT angiography (Discovery CT750 HD, GE) of the aortic arch and supra-aortic arteries in the Department of Diagnostic Imaging of the Hospital of Trauma Surgery in Piekary Śląskie. CT-angiogram revealed the right aortic arch with mirror-image branching and right-sided descending thoracic aorta. The first branch extending from the arch was the right common carotid artery and the second one was the right subclavian artery. Aplasia of the left brachiocephalic trunk was recognized

Figure 1. 3-D reformat images of angio-CT depict: (A) a front view of the right aortic arch with mirror-image branching and aplasia of the left brachiocephalic trunk, (B) collateral circulation connecting the descending aorta with the left subclavian artery, (C) a back view of the anomaly.

and a connection between the left subclavian artery and the left common carotid artery was visualized. The examination revealed also a single, expanded vessel leaving the descending aorta- a posterior intercostal artery, passing into a paraspinal network of small, sinuous vessels and connecting with the left subclavian artery through the internal thoracic artery. CTA presented asymmetry of subclavian arteries and common carotid arteries, significantly narrower on the left side, which suggests reduced blood flow. Both vertebral arteries arise properly from the subclavian arteries (Figures 1, 2).

Figure 2. Axial images of angio-CT show (A, B) collateral circulation through an expanded posterior intercostal artery, that extends from the right-sided descending aorta.

Discussion

Right-sided aortic arch with mirror-image branching and aplasia of the left brachiocephalic trunk is a rare clinical case [3,4]. Only a few cases have been reported in the literature [4–6]. The incidence in the population of dextro-position of the aortic arch is approximately 0.1% [7]. It is rare among normal individuals and usually it is linked with congenital heart defects, for example tetralogy of Fallot. Normal formation of the major arteries depends on the transformation of the aortic sac and the arch vessel during prenatal development [1]. Regression in the left IV arch vessel results in the right-sided arch of the aorta [8].

Singh et al. [5] described a 36-year-old woman, suffering from vertigo and left hand claudication, caused by dextro-position of the aortic arch with isolation of the left brachiocephalic artery and a steal syndrome through the left vertebral artery into the left subclavian artery. A very similar vascular anomaly was reported [6] in a 49-year-old female with symptoms of subclavian steal syndrome. Pauliukas [4] showed a case of a 35-year-old patient with the same pathology, surgically treated, presenting neurological manifestations of cerebrovascular insufficiency and absence of pulse on the left arm.

References:

1. Rea G, Valente T, Iaselli F et al: Multi-detector computed tomography in the evaluation of variants and anomalies of aortic arch and its branching pattern. *Ital J Anat Embryol*, 2014; 119: 180–92
2. Shouford WH, Sybres RG, Edwards FK: The three types of right aortic arch. *Am J Roentgenol*, 1970; 109: 67–74
3. Mc Elhinney DB, Hoydu AK, Gaynor JW et al: Patterns of right aortic arch and mirror-image branching of the brachiocephalic vessels without associated anomalies. *Pediatr Cardiol*, 2001; 22: 285–91
4. Paulikas PA: 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. *Angio Sosud Khir*, 2005; 11: 131–34
5. Singh B, Satyapal KS, Moodley J, Pajaruthnam P: Right aortic arch with isolated left brachiocephalic artery. *Clin Anat*, 2001; 14: 47–51
6. Kokotsakis J, Anagnostakou V, Nenekidis I, Lioulis A: Right aortic arch with mirror image aplasia of brachiocephalic trunk presented as subclavian steal syndrome. *J Cardiothorac Surg*, 2011; 40: 1029–30
7. Kanne JP, Godwin JD: Right aortic arch and its variants. *J Cardiovasc Comput Tomogr*, 2010; 4: 293–300
8. Niszczoła C, Kolečnik A: The embryogenesis and morphology of the aortic arch malformations. *Pediatr Pol*, 2013; 88: 80–91

In our case, poor clinical symptoms and patient's late age diagnosis are associated with efficient collateral circulation, connecting the descending aorta and the left subclavian artery, through a network of small vessels. Ultrasound examination suggested vascular pathology. However, CTA clearly revealed the problem. The knowledge of types of pathology is important for the clinical and therapeutic reasons. Arterial anomalies could cause problems during invasive procedures or coronary interventions. In our patient sudden interruption in the blood flow to the left upper extremity, due to for example a closure of one of the vessels from the collateral circulation, could cause symptoms of cerebrovascular insufficiency and upper limb ischemia.

Conclusions

Medical knowledge of uncommon vascular malformations is still expanding because of the development in diagnostic imaging and extensive use of examinations. It is important for the clinical and therapeutic reasons to be aware of all the variations of asymptomatic arteries to avoid complications during invasive procedures.