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Case Report

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Unusual Case Report of Subclavian Steal Syndrome Caused by Rare Congenital Vascular Anomaly - Left Subclavian Artery Agenesis with Auto Bypass

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Abstract

The typical branching pattern of the aortic arch is present in approximately 65% of the population. Aortic arch anomalies occur in 35 % of people. Aortic arch and subclavian artery anomalies might occur during embryogenesis, when some parts of the primitive aorta persist or are not formed from the very beginning. In some cases, the causative factor remains unknown, malformation itself is an incidental finding and patients show no symptoms. A few anatomical variants of the aortic arch and the subclavian artery have been described but vascular agenesis seems to be an exceptional finding as it has been reported in only few references, which report cases of unilateral or bilateral congenital absence of the internal carotid artery, vertebral artery agenesis or right subclavian artery agenesis.

In this article, the authors present a case of a 51-year-old patient with the subclavian steal syndrome caused by left subclavian artery agenesis, who suffered from undefined arm discomfort and occasional syncope.

Keywords: Subclavian Artery, Agenesis, Subclavian Steal Syndrome, Autobypass, Aortic Arch, Anomaly

Introduction

The subclavian arteries are major arteries of the human upper body, which receive blood supply from the aortic arch and supply blood to the head, neck, thorax and upper extremities. They are paired, asymmetrical structures of high clinical importance due to their aortic origin, wide diameter and significant anatomical role. What happens then when one part of this crucial artery is missing ? A few anatomical variants of the subclavian artery, although infrequent, have been described. On the other hand, congenital anomalies of the subclavian arteries appear to be an extreme rarity on a worldwide scale – to the knowledge of the authors of the article.

In this article, the authors present an unusual case of a patient with the subclavian steal syndrome caused by left subclavian artery agenesis – a congenital anomaly described only in few references.

Case Report

A 51-year-old patient reported to the Angiology Department, with the history of poorly defined discomfort in his left arm persisting for years. The patient was not able to describe the character of the discomfort but noted that it started to interfere with his daily activities, which was the reason for seeking medical help. In the past, there were incidents of vertigo and syncope.

On physical examination, still undefined discomfort in the left arm aggravated by movement was found. No neurological deficits, such as paresis, muscle weakness, paresthesia or other symptoms were present. Brachial, radial and ulnar pulses were palpable. Currently, there were other complaints.

For further diagnostics, the patient was referred for computer tomography angiography (CTA) of the thoracic aorta, carotid and vertebral arteries and subclavian arteries, with the initial diagnosis of the subclavian steal syndrome.

CTA imaging revealed:

- agenesis of the first part of the left subclavian artery (LSA), /image1/
- auto bypass from the medial aspect of the descending thoracic aorta supplying the left subclavian artery, /image2 and 3/
- hypoplasia of the left internal thoracic artery (LITA), which turned out to be a part of the auto bypass and originated directly from the thoracic aorta
- hypoplasia of the left vertebral artery (LVA) with an atypical ostium at the top of the part of the left subclavian artery.
- aneurysmatic dilatation of the proximal ascending thoracic aorta /image4/

Computed Tomography Images

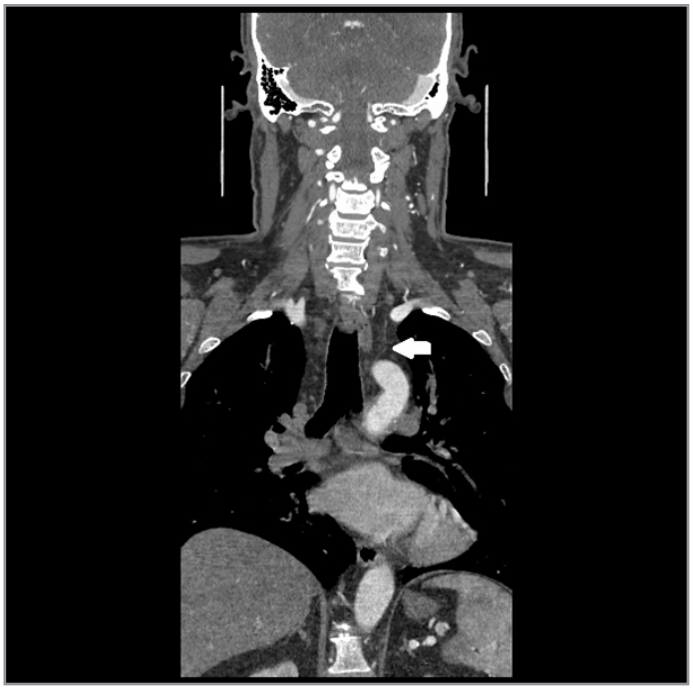

Figure 1: Computed tomography angiography of the thorax, MPR reconstruction, frontal view - agenesis of the first part left subclavian artery (LSA) - white arrow.

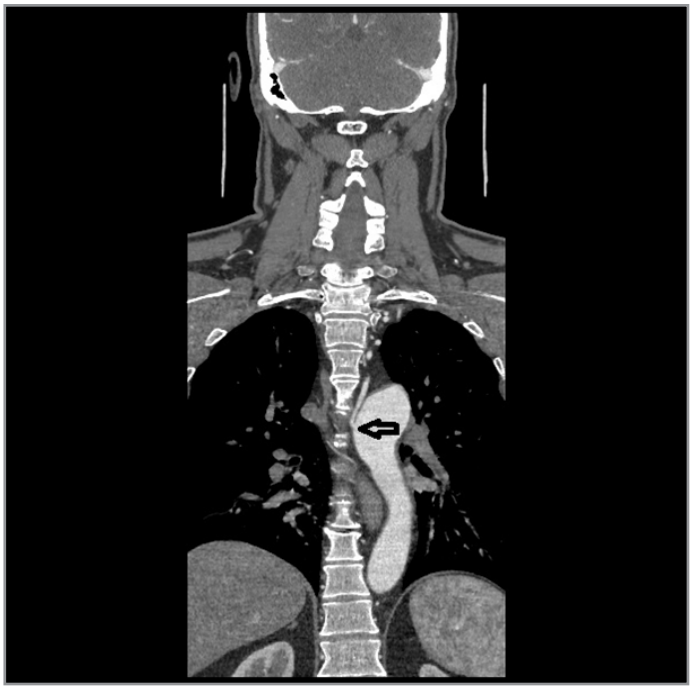

Figure 2: Computed tomography angiography of thorax, MPR reconstruction, frontal view - auto bypass from the medial aspect of the descending thoracic aorta supplying the left subclavian artery – black arrow.

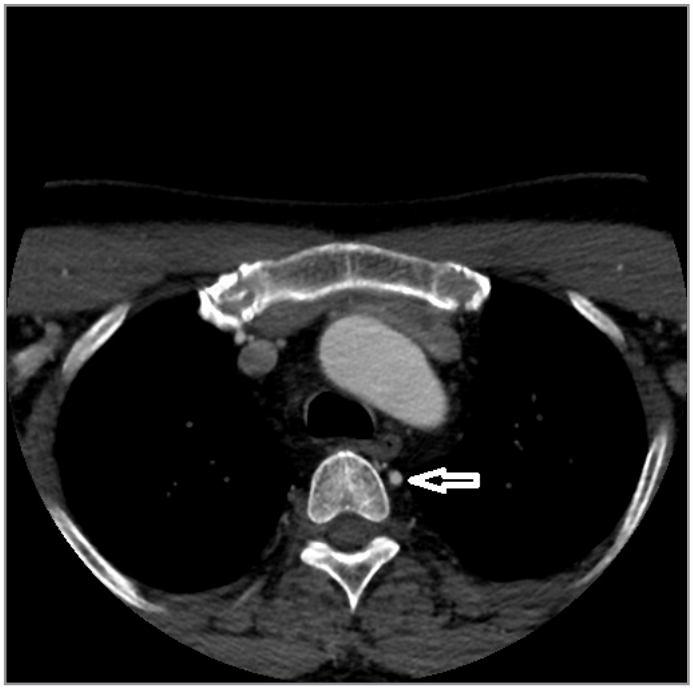

Figure 3: Computed tomography angiography of thorax, axial scan - autobypass from the descending thoracic aorta - white arrow.

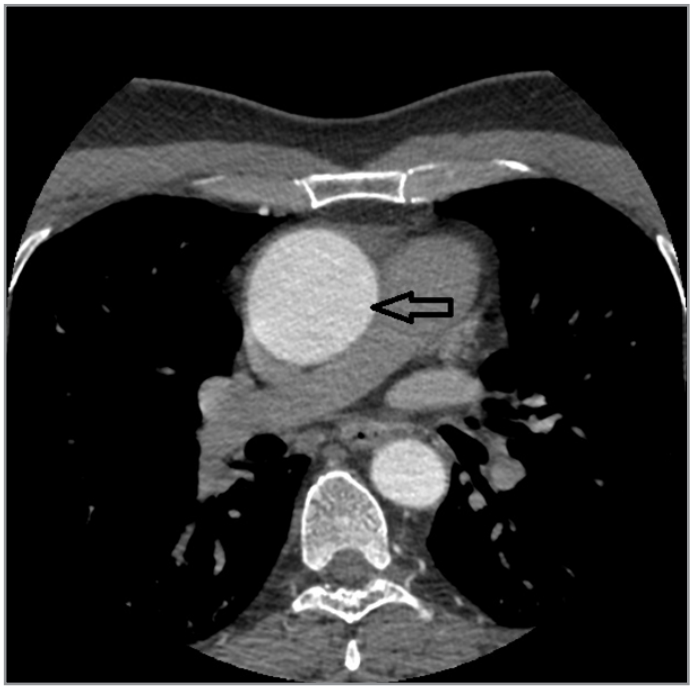

Figure 4: Computed tomography angiography of thorax, axial scan - aneurysmatic dilatation of the proximal ascending thoracic aorta - black arrow.

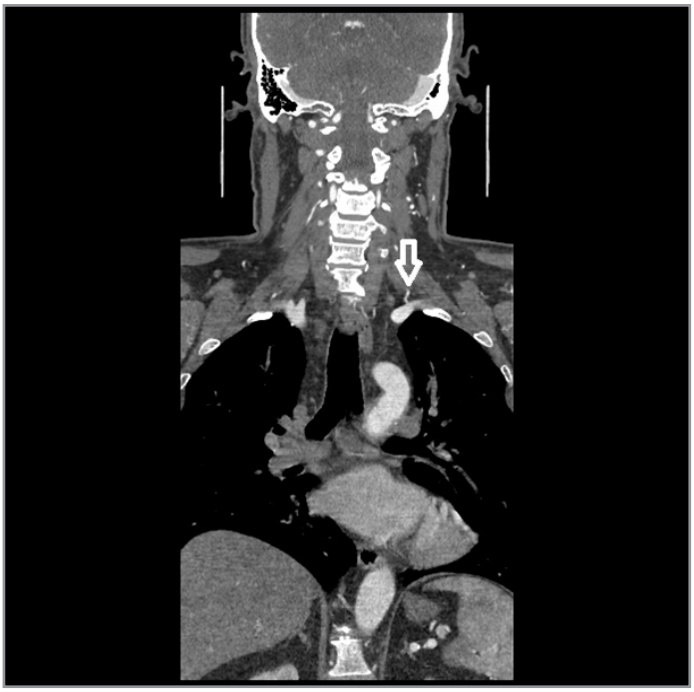

Figure 5: Computed tomography angiography of thorax, axial scan - aneurysmatic dilatation of the proximal ascending thoracic aorta - black arrow.

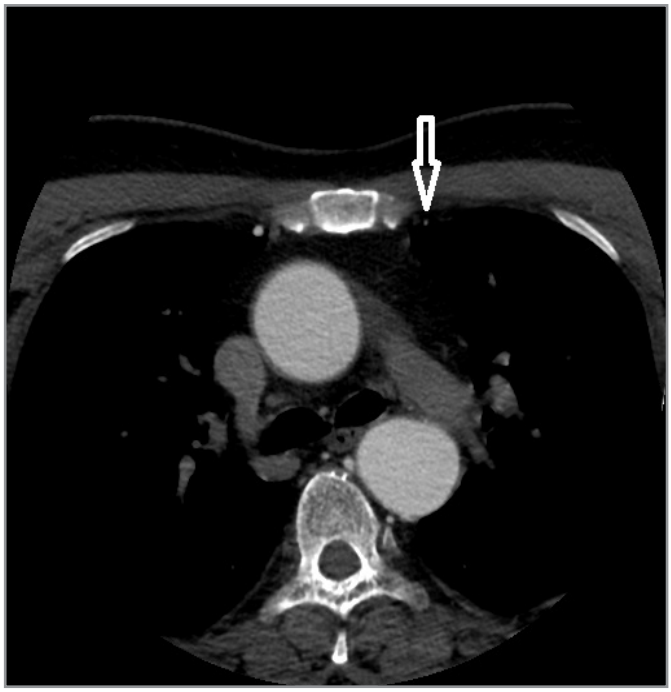

Figure 6: Computed tomography angiography of thorax, axial scan - aneurysmatic dilatation of the proximal ascending thoracic aorta -white arrow.

The patient has been referred to the University Clinical Hospital in Wroclaw to schedule vascular surgeon consultation and to establish further treatment.

Discussion

Discussing subclavian artery anomalies and clinical consequences, the authors consider it is appropriate to remind anatomy and embryogenesis of the aortic arch and its branches including subclavian arteries.

Subclavian arteries are branches of the aortic arch but with different origin sites - the right subclavian artery emerges from the brachiocephalic trunk along with the right common carotid artery, whereas the left subclavian artery arises directly from the aortic arch. Both left and right subclavian arteries leave the thorax through the upper thoracic outlet. Each subclavian artery is divided into three segments, depending on its relation to the anterior scalene muscle: 1. prescalane segment, 2. scalene segment and 3. postscalane segment.

Subclavian arteries give off five branches, in the following order: the vertebral artery, the internal thoracic artery, the thyrocervical trunk, the costocervical trunk, and the dorsal scapular artery. During embryogenesis, the innate structure of forming aorta is the aortic sac, which gives rise to the paired aortic arches that go into pharyngeal arches, and the dorsal aorta, that creates seven intersegmental arteries. Fourth aortic arch contributes to the formation of the proximal right subclavian artery while the seventh intersegmental arteries form the subclavian arteries bilaterally [1, 2].

The typical branching pattern of the aortic arch is present in approximately 65% of the population. Aortic arch anomalies occur in 35 % of people. Most frequent one (10-20% of the population), referred as a bovine arch, occur when the brachiocephalic trunk gives off the right subclavian artery, right common carotid artery and left common carotid artery, whereas the left subclavian artery originates directly from the aortic arch. Other variants exist in less the 3% of people and include :

- shortened brachiocephalic trunk bifurcating into the right subclavian and right common carotid arteries with the left common carotid artery branching off from the aortic arch close to the brachiocephalic trunk and the left subclavian originating from the aortic arch in a usual manner
- the left vertebral artery arising directly from the aortic arch, between the left common artery and left subclavian artery
- the right-sided aortic arch with the aberrant right subclavian artery, named the arteria lusoria.

The last variant has its serious clinical implications due to compression of the esophagus in 5% of patients and possibility of causing congenital left subclavian steal syndrome, due to the retrograde blood flow from the left vertebral artery to the isolated left subclavian artery [3, 5].

As for the subclavian artery variations, three most common variants have been described: aberrant left subclavian artery (32.9%), right aortic arch with mirror-image branching (49.6%), and isolated left subclavian artery (17.4%). At the same time, the authors would like to draw reader's attention to the fact that in current literature there are few reports of a rare congenital anomaly, which is vascular agenesis. These were cases of unilateral or bilateral congenital absence of the internal carotid artery, vertebral artery agenesis and right subclavian artery agenesis. Aortic arch and subclavian anomalies might occur during embryogenesis, when some parts of the primitive aorta persist or are not formed from the very beginning. Some authors believe

that agenesis of the subclavian artery specifically, might occur due to intrauterine injury to the 7th intersegmental artery or the 4th aortic arch. In some cases, the causative factor remains unknown and malformation is an incidental asymptomatic finding [5-9].

In our case, the patient developed subclavian steal syndrome as a result of agenesis of the first, proximal part of the left subclavian artery. Subclavian steal syndrome is a phenomenon, in which retrograde blood flow from the ipsilateral vertebral artery happens, when subclavian artery is stenosed or occluded proximally to the origin of the vertebral artery – the arm „steals" blood from the head. In approximately 95 % cases, atherosclerosis of the subclavian artery is the main cause. Whereas the majority of the patient is asymptomatic, the remaining minority presents signs of ischemia in the affected arm, most commonly as an arm claudication, or vertebrobasilar insufficiency. In our patient's case, undefined arm discomfort and occasional syncope, both exacerbated by exercise, prevailed. What makes this case unique is a combination of congenital vascular anomalies in one patient that prevented him from developing severe symptoms. The remaining part of the left subclavian artery has been supplied by the internal thoracic artery with its unusual origin from the aortic arch, thus creating an auto bypass directly from the aorta. Therefore hypoplastic left vertebral artery maintained moderate blood flow to the head. In the case of such complex anatomical alterations, computed tomography angiography with three-dimensional reconstruction seems to be a suitable tool and the first-choice imaging method. It allows to detect vascular anomalies and create detailed reconstructions, useful for planning further treatment, including surgical interventions. CTA provides images of high resolution, which can be processed in the MPR (Multiplanar Reconstruction) or 3D reconstructions in a short time. Furthermore, new generation scanners are equipped with radiation dose reduction technologies to decrease radiation exposure. High pitch spiral mode allows to diminish cardiac pulsation artefacts. Magnetic resonance angiography offers comparable results to computed tomography angiography in terms of image resolution and reconstruction. In everyday practice it is used when access to CTA is restricted, CTA results are incoherent or for reasons of costs. In the future it might become the golden standard as long as the time between the sequences will be reduced, which makes space for further research and MR modality development. For the classical subclavian steal syndrome continuous wave Doppler and duplex ultrasonography might be a screening tool in patients with typical symptoms. This is a readily available, non-invasive method, which provides real time images of the reversed flow in the ipsilateral vertebral artery and typical parvus-tardus waveform and monophasic waveform in the distal subclavian artery. However, it seems insufficient to evaluate complex anomalies because of bony structures overlying examined vessels and limited ability to create image reconstructions [5, 10-13].

Conclusion

Case reports of rare congenital vascular anomalies such as the presented case of the subclavian artery agenesis increase awareness of such phenomena among medical specialists. This affects effectiveness of a diagnostic process of patients with those anomalies, who may remain asymptomatic or undiagnosed for many years due to atypical clinical image. Knowledge of the anatomy, embryological development and possible variants of the aortic arch and its branches allows one to understand the concept of congenital anomalies, their complexity and possible clinical implications. Imaging techniques, especially computed tomography angiography (CTA) with three-dimensional reconstructions, enable to detect the alternated anatomy as well as planning further management. In the future, new modalities such as magnetic resonance (MR) will gain in prominence in the course of further research and development to provide even more satisfactory results.

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