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CASE REPORT

CLINICAL CASE

Interarm Blood Pressure Difference Revealing a Right-Sided Aortic Arch and Occluded Aberrant Left Subclavian Artery

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ABSTRACT

We present the case of a 33-year-old patient with a rare combination of a right-sided aortic arch and occluded asymptomatic aberrant left subclavian artery diagnosed after the coincidental finding of an interarm blood pressure difference. Because there were no symptoms of local compression or subclavian steal, conservative management was suggested. (J Am Coll Cardiol Case Rep 2024;29:102246) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 33-year-old male patient was referred by the general practitioner because of a significant blood pressure (BP) difference between arms, with the left arm lower than the right. There were no cardiac symptoms and no hoarseness or swallowing problems. There were also no symptoms of claudication in the left arm at rest or during exercise, and there were no other symptoms suggestive for subclavian steal. Physical examination revealed right arm BP of 132/83 mm Hg and left arm BP of 101/74 mm Hg. The right radial pulse was strong, but the radial pulse in the left arm was barely palpable. The heart and lung sounds were normal, and the left arm did not show any atrophic changes.

LEARNING OBJECTIVES

- To understand the importance of always measuring the blood pressure in both upper extremities as well as in the lower extremities during clinical examination.
- To be able to make a differential diagnosis of an interarm blood pressure difference.
- To understand the clinical importance of congenital anomalies of the aortic arch because the anomalies may be related to vessel rings or other congenital cardiovascular conditions.

PAST MEDICAL HISTORY

The patient's previous medical history consisted of ventriculoperitoneal shunting for hydrocephalus.

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ABBREVIATIONS AND ACRONYMS

ALSA = aberrant left subclavian artery

BP = blood pressure

CTA = computed tomography angiography

LSA = left subclavian artery

RAA = right-sided aortic arch

DIFFERENTIAL DIAGNOSIS

In younger people, a BP difference between both arms is often caused by muscular compression or by a structural problem that prevents smooth blood flow through an artery.[1] In older people, it is usually caused by a blockage arising because of atherosclerosis.[1] Other causes are subclavian artery stenosis, aortic coarctation, aortic dissection or aneurysm, vasculitis (eg, Takayasu disease), connective tissue disorders, fibromuscular hyperplasia, and thoracic outlet compression.[1]

INVESTIGATIONS

Transthoracic echocardiography showed normal cardiac function, but suprasternal imaging was suspicious for a right-sided aortic arch (RAA), and the origin of the left subclavian artery (LSA) could not be visualized. Computed tomography angiography (CTA) revealed a RAA with a focal aneurysmal dilatation of the descending aorta at the origin of an aberrant subclavian artery, matching with Kommerell diverticulum (Figure 1). The proximal segment of the LSA was hypoplastic. A fibrous string appeared to be present from the Kommerell diverticulum to the bifurcation of the left subclavian and left vertebral arteries, presumably a remnant of the occluded LSA (Figure 2).

MANAGEMENT

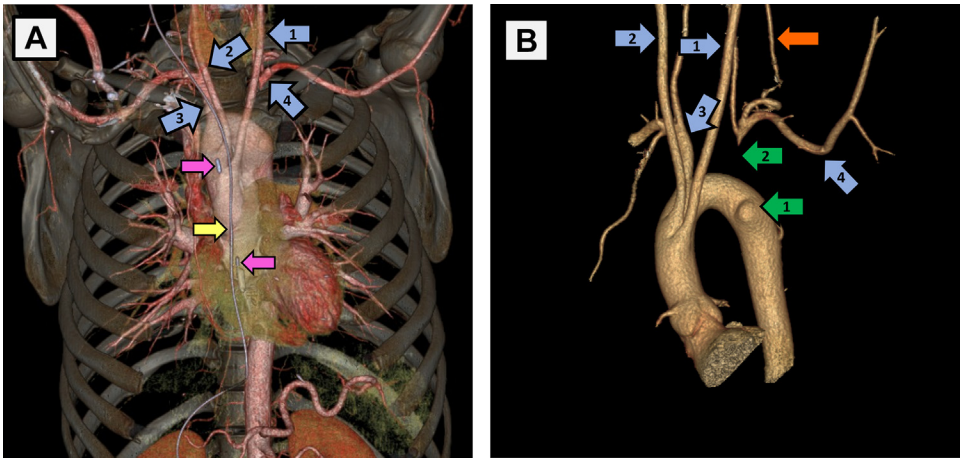
Because our patient denied any symptoms, conservative management was suggested until the development of complaints due to local compression or subclavian steal.

DISCUSSION

A BP difference between arms is a frequently encountered phenomenon. An interarm difference is defined as a variance in systolic BP of >10 mm Hg and can be both physiological and pathological.[1] It is important to detect an interarm BP difference for further vascular assessment and management of risk factors.[1]

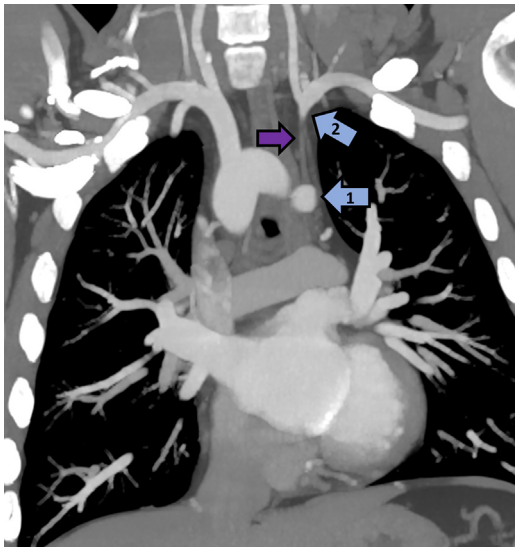
A RAA is a rare congenital aortic arch variant that has been reported to occur in 0.05% to 0.1% of the general population.[2] RAAs have been classified into 3 types according to the branching pattern of the arch vessels (Figure 3): a RAA with mirror image branching, a RAA with an aberrant left subclavian artery (ALSA), and a RAA with isolation of the LSA in which the LSA does not have a connection with the aorta but is connected to the pulmonary artery by a left ductus

FIGURE 1 Computed Tomography Angiography Volume Rendering Reformations of the Aortic Arch With and Without Subtraction of the Bony Structures



Computed tomography angiography (CTA) volume rendering reformations of the aortic arch (B) with and (A) without subtraction of the bony structures. The first branch arising from the aortic arch is the left carotid artery (blue arrow 1) followed by the right carotid artery (blue arrow 2), right subclavian artery (blue arrow 3), and the aberrant left subclavian artery (blue arrow 4). The pink and yellow arrows depict the patient's first and second ventriculoperitoneal shunt, respectively. The Kommerell diverticulum is visible at the origin of the aberrant subclavian artery from the descending aorta (green arrow 1). The proximal segment of the left subclavian artery exhibited hypoplasia (green arrow 2). Computed tomography angiography (CTA) also revealed a prominent and tortuous left deep cervical artery (orange arrow), a potential collateral arterial supply for the subclavian artery.

FIGURE 2 **Computed Tomography Angiography Coronal Maximum Intensity Projection Reformation of the Aortic Arch**

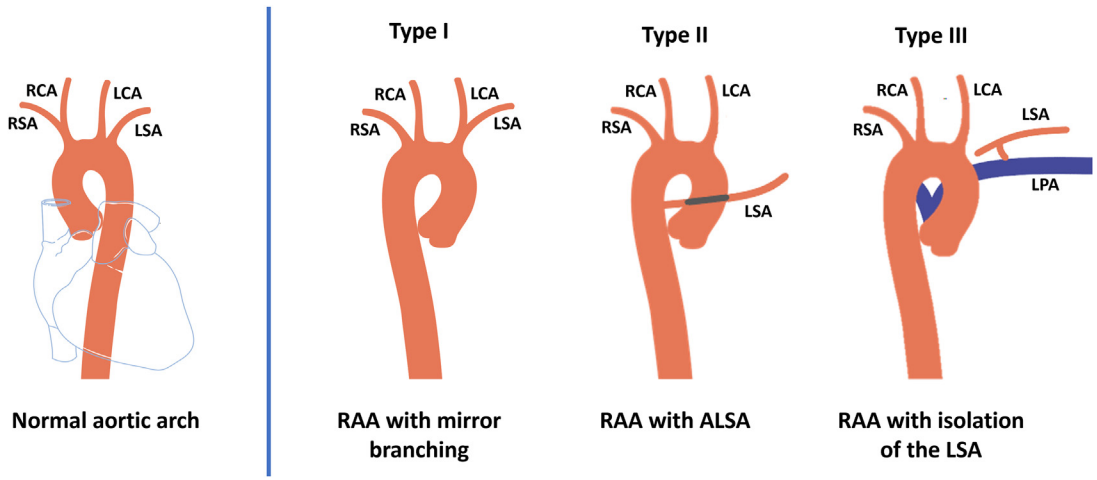


A fibrous string (purple arrow) is visible ranging from the Kommerell diverticulum (blue arrow 1) to the left subclavian artery just inferior of the left vertebral artery origin (blue arrow 2).

arteriosus.[3] Our patient was diagnosed with the second type of RAA (a RAA with ALSA) (Figure 1), which is the most common RAA type and is rarely associated with other cardiovascular abnormalities. This anomaly results from interruption of the dorsal segment of the left arch between the left common carotid and left subclavian arteries with regression of the right ductus arteriosus in the hypothetical double aortic arch.[3] This anomaly rarely produces symptoms and is usually an incidental radiologic finding.[3] Nevertheless, this ALSA courses across and may compress thoracic structures on its way to the left thoracic outlet.[2] Compression on local structures may also arise from the Kommerell diverticulum, and together with the left pulmonary artery and left ductus arteriosus, a RAA with an ALSA may form a complete vascular ring. Such local compression may result in changes in voice, difficulties in swallowing, and respiratory symptoms.[2,3] In addition, patients may also present for the first time with a rupture of a thoracic aneurysm.[3]

Nevertheless, the anatomy in our patient differs from the typical second type of RAA because the proximal segment of the LSA was occluded. Because of the young age of the patient, the concurrent presence of a congenital anomaly of the aorta, and the fibrous string visible on CTA, this occlusion is most likely caused by congenital hypoplasia rather than atheromatous plaque deposition.[4] This combination of anomalies is extremely rare and has only been previously described in 2 case reports.[2,4] However, only a BP difference between both arms was present in an otherwise asymptomatic patient. In the previously described cases, the patients presented with recent changes in their voice because of local

FIGURE 3 **Overview of the Classification of the 3 Types of RAA**



ALSA = aberrant left subclavian artery; LCA = left carotid artery; LPA = left pulmonary artery; LSA = left subclavian artery; RAA = right-sided aortic arch; RCA = right carotid artery; RSA = right subclavian artery. This figure was partly generated using Servier Medical Art, provided by Servier, licensed under a Creative Commons Attribution 3.0. unported license.

compression[2] and with an ischemic stroke in the left posterior circulation area resulting from subclavian artery steal.[4] The term "subclavian steal" refers to a reversed blood flow in the vertebral artery distal and ipsilateral to a hemodynamically significant stenosis or occlusion of the proximal subclavian artery.[5] The common characteristic of congenital subclavian steal syndrome is an episode of ischemic symptoms during childhood. However, some patients develop neurologic steal symptoms or arm claudication with aging, with arteriosclerosis usually being the cause of the decompensation of the collateral circulation.[4] Progressively worsening left arm claudication has also been described in a similar anatomical variant presenting with a focal stenosis at the origin of the ALSA.[6] In cases of left subclavian artery occlusion, blood supply to the left arm classically occurs via the left vertebral artery. However, CTA in this patient also demonstrated a prominent and strikingly tortuous deep cervical artery, suggesting that at least part of the collateral flow originates from this artery. However, no additional imaging was performed to study this phenomenon in more detail.

To the best of our knowledge, this is also the first time that a combination of a RAA and ALSA occlusion with hydrocephalus has been described. Like a RAA, hydrocephalus is also a rare condition with an overall global prevalence of approximately 85 per 100,000 individuals.[7] In the literature, cases of hydrocephalus of various origins in combination with other aortic abnormalities such as an interrupted, tortuous, or double aortic arch or aortic arch coarctation have been described, often in a syndromic context or with an underlying genetic pattern (eg, Fryns syndrome). However, genetic testing was never performed in our patient, and the cause of the hydrocephalus is unknown.

FOLLOW-UP

A follow-up consultation with new CTA is planned within 5 years or sooner if complaints arise because of local compression or subclavian steal.

CONCLUSIONS

Congenital anomalies of the aortic arch are uncommon and are usually an incidental radiologic finding in asymptomatic patients. However, they are of clinical importance because the anomalies may be related to vascular rings or other congenital cardiovascular conditions. We presented an extremely rare case of a combination of a RAA and ALSA occlusion. The presenting symptom was a blood pressure difference between the 2 upper limbs. Because the patient denied any symptoms of local compression or subclavian steal, conservative management was suggested.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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REFERENCES

1. Gopalakrishnan S, Savitha AK, Rama R. Evaluation of inter-arm difference in blood pressure as predictor of vascular diseases among urban adults in Kancheepuram District of Tamil Nadu. *J Family Med Prim Care*. 2018;7(1):142–146.

2. Margolis J, Bilfinger T, Labropoulos N. A right-sided aortic arch and aberrant left subclavian artery with proximal segment hypoplasia. *Interact Cardiovasc Thorac Surg*. 2012;14(3):370–371.

3. Turkvatan A, Buyukbayraktar FG, Olcer T, Cumhur T. Congenital anomalies of the aortic arch: evaluation with the use of multidetector computed tomography. *Korean J Radiol*. 2009;10(2):176–184.

4. Tempaku A, Kuroiwa T, Nishio A. Aberrant left subclavian artery occlusion in right-sided aortic artery associated with left cerebral infarction: a case report. *Interv Neuroradiol*. 2018;24(3):322–326.

5. Potter BJ, Pinto DS. Subclavian steal syndrome. *Circulation*. 2014;129(22):2320–2323.

6. Batheeb NA, Habbab LM, Qattan NM. Symptomatic stenosis of left subclavian artery from Kommerell's diverticulum. *Asian Cardiovasc Thorac Ann*. 2015;23(9):1068–1071.

7. Koleva M, De Jesus O. *Hydrocephalus*. StatPearls; 2023.

KEY WORDS aberrant left subclavian artery, blood pressure difference, congenital subclavian steal, Kommerell diverticulum, right-sided aortic arch