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Right Aortic Arch and Arterial Flow Steal: Case Report

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Abstract: The aortic arch develops around the seventh week of gestation from the migration and regression of the branchial arches. The fourth arch gives rise to the aortic arch after the regression of the right dorsal root. When this regression occurs on the left side, it may result in a right aortic arch. This study reports a 32-year-old female patient with a right aortic arch who presented to the outpatient clinic with complaints of tingling and numbness in the left upper limb. On physical examination, there was a decrease in pulses in this limb during thoracic outlet maneuvers. Computed tomography identified the aortic arch and thoracic aorta on the right side, with tracheal stenosis and a retroesophageal course. CT angiography revealed more than 50% stenosis at the origin of the left subclavian artery. Duplex scan demonstrated flow with a retrograde meso-systolic component in the left vertebral artery, a finding compatible with a subclavian steal phenomenon. This is a rare case of right aortic arch with an aberrant left subclavian artery without Kommerell's diverticulum. The case highlights the importance of correlating clinical findings with imaging studies in the investigation of vascular symptoms, contributing to accurate diagnosis and appropriate therapeutic decision-making.

Keywords: Right aortic arch; Cardiovascular malformation; Computed tomography.

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1. Introduction

Anomalies of the aortic arch can be classified based on their anatomy. They include: anomalies of the left aortic arch with variations in the branching of the great vessels; anomalies of the right aortic arch, in which the arch crosses the main bronchus and passes to the right of the trachea and esophagus, the descending aorta may be found on either the left or the right side [1]; double aortic arch, in which two aortic segments (right and left) encircle the trachea and esophagus, forming a complete vascular ring; and cervical aortic arch, in which the aortic arch is located above the clavicle [2].

The right aortic arch stands out for presenting different anatomical configurations. Among the most frequent are right aortic arch with mirror-image branching, right aortic arch with aberrant left subclavian artery, and right aortic arch with isolation of the left subclavian artery [2–6]. In right aortic arch with mirror-image branching, the left subclavian artery originates together with or just before the left common carotid artery, reproducing in an inverted way the normal anatomical patterns, with the following branching sequence: left subclavian artery, left common carotid artery, and a common trunk of the right subclavian and right common carotid arteries [7]. Its course lies between the superior vena cava and the right side of the trachea and esophagus, without forming a vascular ring in most cases. This anomaly results from regression of the distal left dorsal aorta beyond the origin of the seventh intersegmental artery, so that the left fourth arch becomes the proximal subclavian artery instead of the definitive arch [8].

In right aortic arch with aberrant left subclavian artery (ALSA), the branching sequence follows this order: left subclavian artery, left common carotid artery, right common carotid artery, and right subclavian artery [4]. The left subclavian artery may emerge as the last branch of the right aortic arch or from a localized dilation in the aorta known as Kommerell's diverticulum (retroesophageal diverticulum), which represents a remnant of the left dorsal aorta. This latter situation results from regression of the left fourth aortic arch between the left common carotid and left subclavian arteries, usually with persistence of the left sixth arch [2].

Right aortic arch with isolation of the left subclavian artery consists of isolation of branches of the aortic arch and results from two ipsilateral interruptions in the arch system. The term "isolation" refers to the fact that one of the arch vessels, in this case the left subclavian artery, originates exclusively from the pulmonary artery via the ductus arteriosus or ligamentum arteriosum, without any direct connection to the aorta. This pattern results from regression of two segments of the left aortic arch: one between the left common carotid and left subclavian arteries, and another distal to the left ductus arteriosus and left subclavian artery [2,5].

Regarding clinical aspects, although these anomalies are often asymptomatic, they may have clinical repercussions depending on their morphology and the adjacent structures involved. The main findings include respiratory symptoms such as stridor and dyspnea, and gastrointestinal symptoms such as dysphagia, resulting from tracheobronchial and esophageal compression by complete vascular rings or anomalous courses. Furthermore, hemodynamic changes, such as subclavian steal phenomenon, may compromise cerebral perfusion and cause neurological symptoms such as dizziness, syncope, or upper limb claudication. In other cases, these anomalies may be associated with congenital heart disease, requiring a multidisciplinary clinical and surgical approach [2,5].

Understanding congenital anomalies of the aortic arch is essential for the differential diagnosis of nonspecific symptoms, especially when associated with vascular changes identified on imaging examinations. Well-documented case reports are valuable tools for teaching, research, and enhancing clinical reasoning, particularly when dealing with rare anatomical entities with systemic repercussions. Thus, the present study aims to report the case of a patient with the right aortic arch. It is therefore a clinical-observational study, in the form of a case report, conducted through the analysis of diagnostic tests.

2. Case Report

A 32-year-old woman, with a normal BMI and no comorbidities such as arterial hypertension, diabetes mellitus, dyslipidemia, or smoking, sought outpatient care in vascular surgery with complaints of tingling and numbness in the left upper limb. These symptoms had started approximately six months earlier, were intermittent, and were triggered by arm elevation and relieved when the limb was at rest.

On physical examination, she presented decreased pulses in the left upper limb during thoracic outlet maneuvers, without audible bruits. The neurological examination revealed no central neurological abnormalities. Imaging tests previously performed as part of routine evaluation revealed a congenital anomaly of the aortic arch. Computed tomography identified the aortic arch and thoracic aorta located on the right side (Figure 1), with tracheal stenosis and a retroesophageal course. CT angiography showed more than 50% stenosis at the origin of the left subclavian artery. Atherosclerosis was ruled out as the cause of the stenosis, as was extrinsic compression. Duplex scan demonstrated flow with a retrograde meso-systolic component in the left vertebral artery, a finding compatible with subclavian steal phenomenon involving the left subclavian artery (Figure 2). Transcranial Doppler was not performed.

Figure 1. Carotid duplex scan image showing retrograde flow in the left vertebral artery, compatible with subclavian steal phenomenon involving the left subclavian artery.

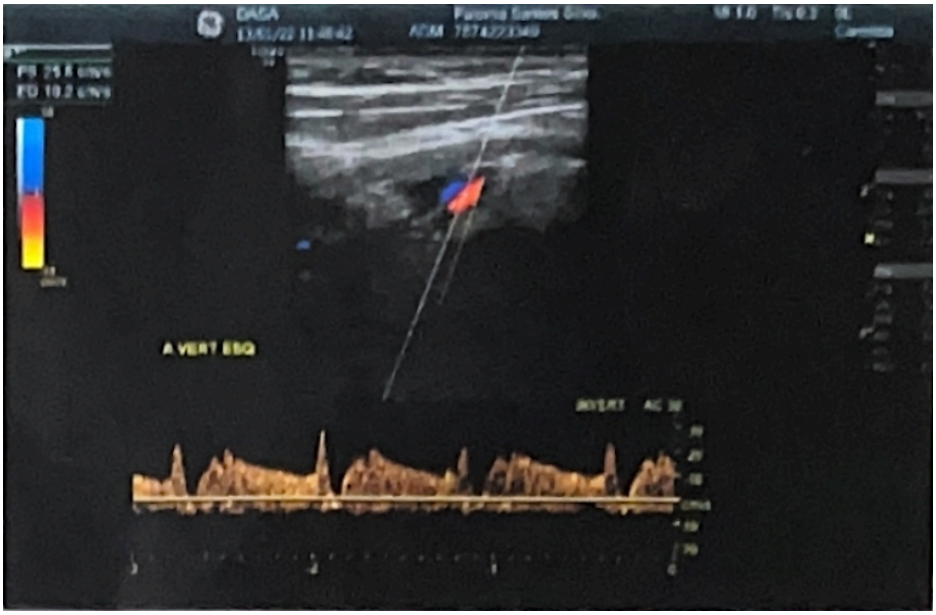
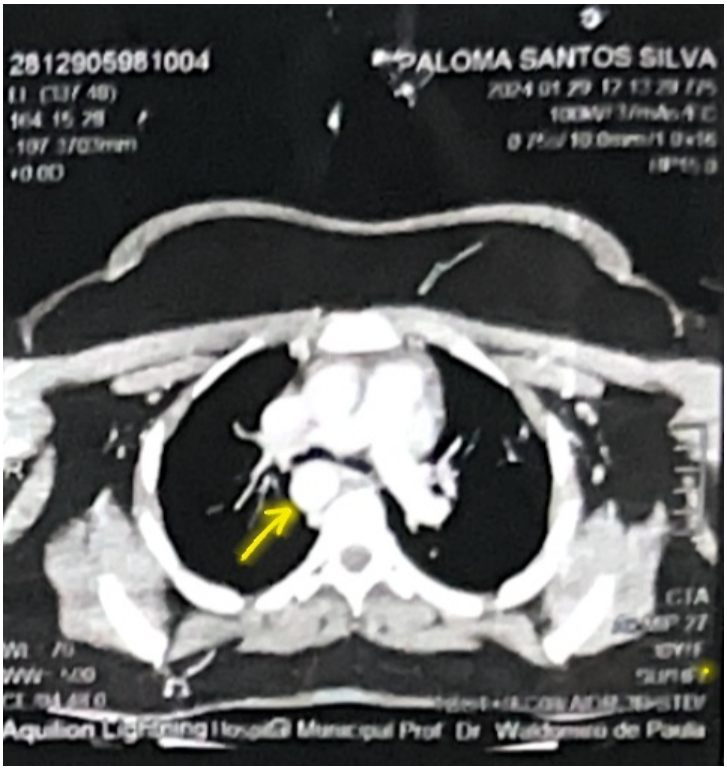


Figure 2. Computed tomography image showing the descending aorta to the right of the vertebral column, associated with a right aortic arch with an aberrant left subclavian artery. Relevant for the diagnosis of the vascular anomaly.



The adopted management was conservative treatment, with the use of an antiplatelet agent (ASA) and physical exercise. The patient was only monitored, maintaining a stable clinical condition during follow-up.

3. Discussion

Right aortic arch represents a rare anomaly of the aorta, with an estimated prevalence of 0.2% [9]. Right aortic arch with aberrant left subclavian artery (ALSA) is the most common variant and, although often an incidental radiological finding as it rarely causes symptoms, it may be associated with esophageal compression symptoms, particularly in older individuals, especially in the presence of ectasia, tortuosity, or aneurysm of the aberrant left subclavian artery [2,10]. Respiratory symptoms due to tracheal compression may be present in pediatric patients. It is rarely associated with other cardiovascular anomalies [11].

When this type of anomaly is present, the ductus arteriosus may originate from the aberrant left subclavian artery, leading to aortic dilatation forming an aneurysm known as Kommerell's diverticulum [12]. Less frequently, patients who do not present dilatation at the origin of the left subclavian artery are classified as having right aortic arch with aberrant left subclavian artery without Kommerell's diverticulum. This anomaly results from regression of the left fourth and sixth aortic arches, preventing the contribution of the left dorsal aorta to the formation of the descending aorta [13].

In the present case, the patient has a right aortic arch with an aberrant left subclavian artery without Kommerell's diverticulum, with a symptomatic presentation related to subclavian steal phenomenon, a condition that resulted in hemodynamic changes and prompted the search for medical care. This phenomenon occurs due to proximal stenosis of the subclavian artery that generates retrograde blood flow in the ipsilateral vertebral artery, diverting blood from the posterior cerebral circulation to the upper limb. Clinically, it manifests with paresthesias, upper limb claudication, dizziness, and in severe cases, vertebrobasilar insufficiency. In the case described, the complaints were limited to effort-induced paresthesias, without episodes of syncope or central symptoms. The absence of significant ischemic changes in the upper limb and the mild clinical presentation justified the choice of conservative management, with no indication for surgery or endovascular intervention. Furthermore, the exclusion of atherosclerosis and extrinsic compression reinforced the diagnosis of hemodynamically significant stenosis of the left subclavian artery as the cause of the steal phenomenon. Clinical follow-up is essential, as some patients may progress with symptom worsening, making it necessary to reassess the need for intervention.

Current guidelines for the management of congenital anomalies of the aortic arch and subclavian steal syndrome recommend an individualized approach, taking into account the presence and severity of symptoms as well as the functional impact on the patient's quality of life. Recognition of these anomalies through imaging tests such as computed tomography and duplex scan is fundamental for proper clinical assessment and management planning. The literature shows that although many of these malformations do not require intervention, symptomatic cases such as the one presented here may need specialized follow-up and, in some contexts, surgical or endovascular treatment [9,14].

The detailed discussion of clinical and radiological findings, as in this case, contributes to the understanding of the clinical manifestations of aortic arch anomalies and reinforces the importance of clinical–radiological correlation, enhancing academic training and diagnostic reasoning of students and healthcare professionals.

4. Conclusion

The present study reported a rare case of a patient with a right aortic arch with an aberrant left subclavian artery and no Kommerell's diverticulum, associated with subclavian steal phenomenon involving the left subclavian artery. Conservative management with antiplatelet therapy and exercise proved effective, reinforcing the importance of individualized management based on clinical severity and imaging findings.

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