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Case Report

Type a Interrupted Aortic Arch: An Unusual Cause of Resistant Arterial Hypertension in Adults

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Abstract: Type A interrupted aortic arch is a rare congenital heart disease characterized by a complete loss of continuity between the ascending and descending portions of the aorta; presentation in adults is exceptional due to its high mortality. This article presents the case of a 57-year-old woman with resistant arterial hypertension since the age of 15, treated with multiple therapeutic regimens, presenting long-standing nonspecific symptoms. CT angiography and aortography were performed, confirming the diagnosis of type A interrupted aortic arch. She was referred to a tertiary care center where a left subclavian artery–descending aorta bypass was performed with an extra-anatomic graft, with good clinical evolution; she is currently asymptomatic under follow-up. Interrupted aortic arch is a rare congenital heart disease with low prevalence in adults due to its high mortality, of which type A is the most frequent and is associated with greater survival due to the formation of collateral arteries. Multiple studies have compared imaging methods to evaluate sensitivity, with modalities such as computed tomography and magnetic resonance imaging being superior to echocardiography.

Keywords: Interrupted Aortic Arch, Refractory Arterial Hypertension, Aortic Bypass.

INTRODUCTION

Type A interrupted aortic arch is a rare congenital heart disease characterized by a complete loss of continuity between the ascending and descending portions of the aorta [2].

Most cases are diagnosed during childhood, which has a crucial impact since, without treatment, most patients die [2]. Diagnosis at later stages is unusual due to low survival, and when it occurs, it is generally secondary to the formation of collateral arteries that connect both segments and generate hemodynamic compensation [5].

Clinical presentation is variable, ranging from asymptomatic patients to headache, decreased pulses in the lower extremities, or resistant arterial hypertension. Due to nonspecific and very common findings, it may go unnoticed for a long time, which can significantly impact the patient's prognosis [5].

Surgical repair is the treatment of choice for aortic arch disorders, since medical therapy alone has been associated with lower survival [3].

CASE PRESENTATION

A 57-year-old woman with a history of systemic arterial hypertension diagnosed at the age of 15, classified as refractory, receiving various treatment regimens. She presented to the cardiology clinic in February 2024 reporting long-

standing nonspecific symptoms characterized by tinnitus, holocranial headache 5/10, and adynamia, in addition to uncontrolled hypertension despite quadruple therapy, with systolic blood pressure levels of 190 mmHg.

On physical examination, rhythmic heart sounds of adequate intensity and frequency were noted, normal first and second heart sounds, no third or fourth heart sound, and no murmurs.

In March 2024, thoracoabdominal-pelvic CT angiography was performed, reporting aortic coarctation with a luminal area of 1 cm (Figure 1), for which she was scheduled for aortic angiography and percutaneous resolution (Figure 2). However, due to inability to advance the guidewire, aortography was performed, documenting interruption of the aortic arch distal to the origin of the left subclavian artery, as well as the formation of a large collateral artery connecting the proximal and distal portions of the aorta, therefore she was referred to thoracic surgery for surgical treatment.

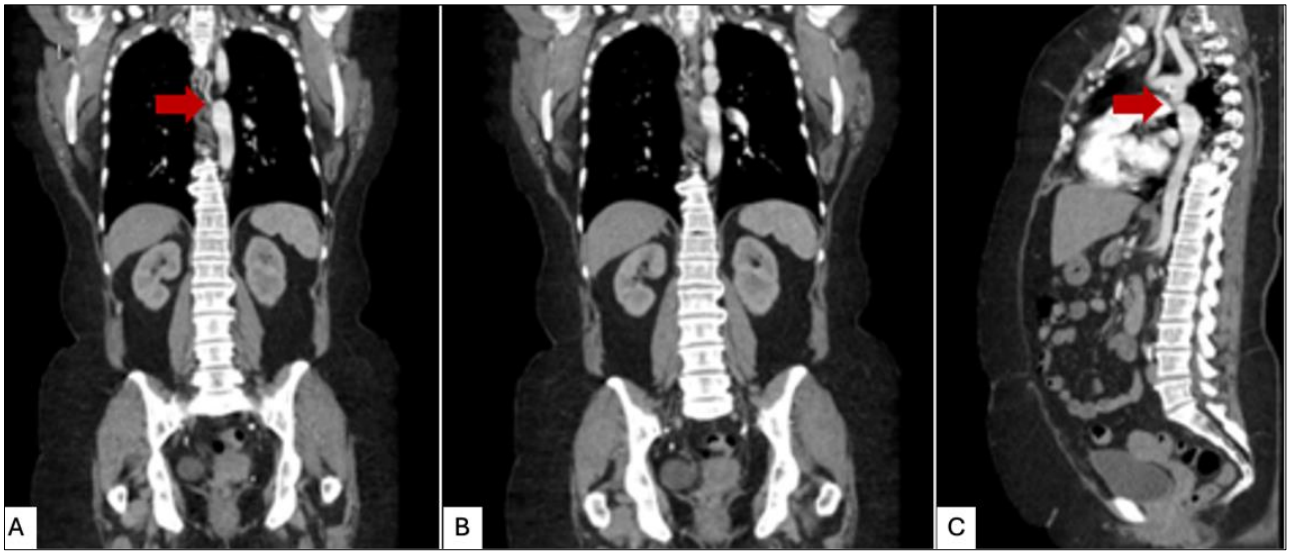

Figure 1: Contrast-enhanced CT angiography of the chest in coronal (A and B) and sagittal (C) sections showing type A interrupted aortic arch, evidencing loss of continuity between the aortic arch distal to the origin of the left subclavian artery and the descending thoracic aorta (arrows)

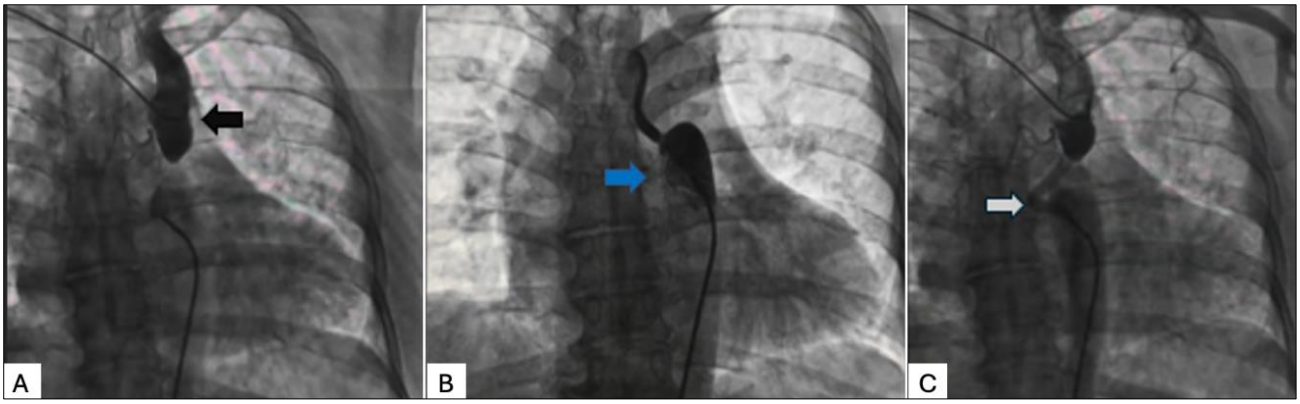

Figure 2: (A y B) Aortic angiography showing absence of contrast passage from the proximal portion (black arrow) to the distal aorta (blue arrow); (C) retrograde filling of the descending aorta is observed originating from a large collateral artery arising from the left subclavian artery (white arrow)

Surgical correction was performed by placing a left subclavian–descending aortic isthmus bypass with an extra-anatomic graft (Figure 3), with a hospital stay of 10 days, showing improvement and subsequent discharge. She continued follow-up in the outpatient clinic and is currently asymptomatic, with controlled blood pressure without antihypertensive treatment. An echocardiogram showed no alterations in wall thickness or ventricular geometry, preserved systolic function with LVEF 60%, as well as CT angiography with 3D reconstruction showing a functional bypass.

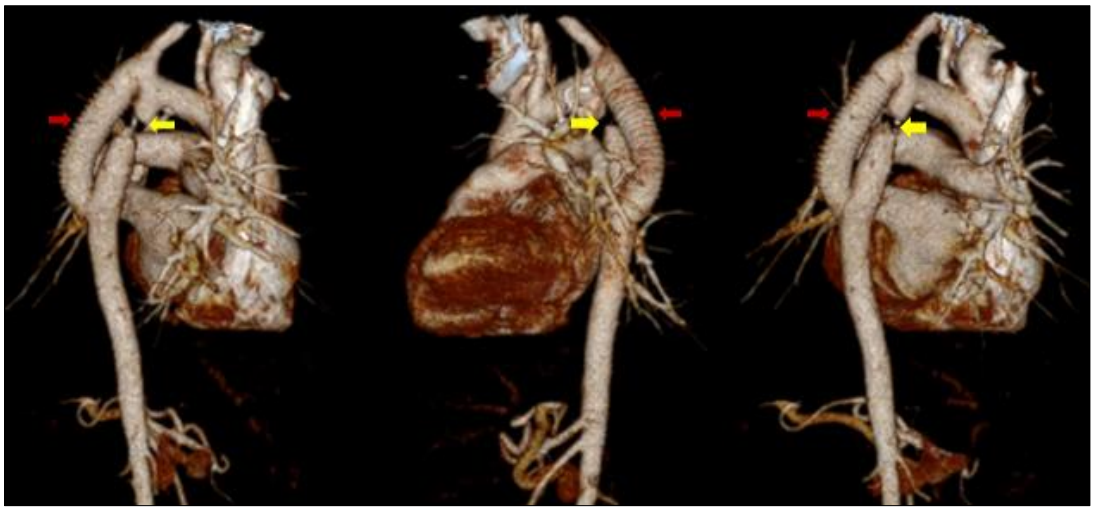

Figure 3: Three-dimensional reconstruction in right and left sagittal sections showing interruption of the aortic arch (yellow arrows) and surgical reconstruction using a subclavian–terminal aortic bypass with an extra-anatomic graft (red arrows)

DISCUSSION

Interrupted aortic arch (IAA) is a rare congenital anomaly characterized by loss of continuity between the ascending and descending aortic arch and represents 1% of congenital heart diseases [2]. According to the classification of Celoria and Patton, type A is the second most frequent type, and the defect occurs distal to the origin of the left subclavian artery, representing approximately 30% of reported cases [1-6].

Interrupted aortic arch typically manifests clinically within the first days of life, most commonly presenting on the first day [7].

Diagnosis in adulthood is exceptional, as most untreated patients die during the first year of life due to heart failure or associated complications [4]. Among anatomical variants, type A is the most frequently reported in adults, which has been attributed to the development of collateral circulation that maintains blood flow to the descending aorta and distal territories.

Clinical presentation in adults is variable and may range from asymptomatic patients to nonspecific symptoms such as headache, decreased pulses in the lower extremities, or resistant arterial hypertension. Refractory hypertension has been described in approximately 70% of cases, followed by claudication in 13% and aortic insufficiency in 10%, findings consistent with the clinical presentation of our patient [4-6].

Imaging studies play a fundamental role in diagnosis. CT angiography has demonstrated greater diagnostic accuracy compared to echocardiography, as it allows detailed anatomical evaluation and identification of collateral circulation. Therefore, computed tomography and cardiovascular magnetic resonance are considered the imaging methods of choice for anatomical evaluation and therapeutic planning due to their high sensitivity and specificity, which is consistent with what was observed in our patient [3-5].

As reported by Zhou JM *et al*., this case highlights the importance of considering secondary causes of hypertension, particularly in patients with early onset or poor blood pressure control despite multiple pharmacological treatments. It also emphasizes that congenital cardiovascular anomalies should remain within the differential diagnosis even in adult patients [3].

CONCLUSIONS

Interrupted aortic arch is a rare congenital anomaly that occurs mainly in the pediatric population and, exceptionally, in adult patients, in whom survival depends on the development of collateral circulation. It may be asymptomatic or present with nonspecific symptoms, which makes timely diagnosis difficult.

This case highlights the importance of proper clinical evaluation, as well as the use of imaging studies, to identify secondary causes of resistant arterial hypertension. Furthermore, it emphasizes that clinical suspicion in patients with difficult-to-control hypertension, especially when associated with discrepancies in pulses or blood pressure between limbs,

should prompt a targeted search for structural etiologies, in which computed tomography angiography and magnetic resonance imaging play a fundamental role in diagnosis and therapeutic planning. Timely recognition of this condition allows appropriate management to be established and may potentially improve long-term prognosis.

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Conflict of Interest: The authors declared no conflict of interest related to this work.

ETHICAL CONSIDERATIONS
Ethical Approval: According to the policies of our institution, case reports do not require ethics committee review.

Consent Statement
Informed consent was obtained from the patient for publication of this case report and any relevant images during the diagnostic and therapeutic process.

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