

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Complete interruption of aortic arch diagnosed in adulthood: a case report [☆]

Abhishek Sharma, MBBS^{*}, Nirmal Prasad Neupane, MD, Prakash Dhakal, MD, Suraj Sharma, MD, Asim Sitaula, MD, Kritisha Rajlawot, MD

Sahid Gangalal National Heart Center, Kathmandu, Nepal

ARTICLE INFO

Article history:

Received 14 July 2022

Revised 21 July 2022

Accepted 23 July 2022

Keywords:

Computed tomography angiography

Congenital heart disease

Collateral circulation

Interrupted aorta

ABSTRACT

Few patients with interrupted aortas survive into maturity, and the majority of instances are diagnosed in young children. There are only a few cases of this extremely rare total aortic interruption that survives into maturity, necessitating the substantial growth of collaterals to supply the descending aorta. Here, we describe a rare instance of an interrupted aorta in a 43-year male that presented in late adulthood with complete interruption of the aortic arch. The patient has remained symptom-free and without treatment. This case gives us an idea about how one can survive into adulthood with complete interruption of the aortic arch given that extensive collaterals are formed.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

A full blockage of the aortic lumen between the ascending and descending aortas is referred to as an interrupted aortic arch. The majority of IAA cases is identified during infancy or the neonatal period and treated [1]. It is a severe aortic arch deformity that affects 3 per million live births. Based on where the interruption occurs, IAA has been divided into 3 categories: type A for interruptions distal to the left subclavian artery origin, type B for interruptions between the left subclavian artery and the left common carotid artery, and type C for interruptions between the innominate and the left common carotid artery [2]. Reviewing the documented instances of adult pre-

sentations of IAA reveals that type A interruptions account for 79% of the reported cases. When adult presentation does occur, it is incredibly rare and almost always the result of severe aortic coarctation, which interrupts blood flow [3].

Case presentation

A 43-year-old man who had been diagnosed with hypertension 2 years' prior presented to the outpatient department of our hospital with uncontrolled hypertension. He had been taking lisinopril for the previous 2 years. On admission, his right and left arm blood pressures were 160/102 and 164/100

Abbreviations: CT, Computed tomography; CTA, Computed tomography angiography; ECG, Electrocardiogram; IAA, Interrupted aortic arch.

[☆] Competing Interests: The authors have no competing interest to report.

* Corresponding author.

E-mail address: sharmaabhishek2052@gmail.com (A. Sharma).

<https://doi.org/10.1016/j.radcr.2022.07.084>

1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

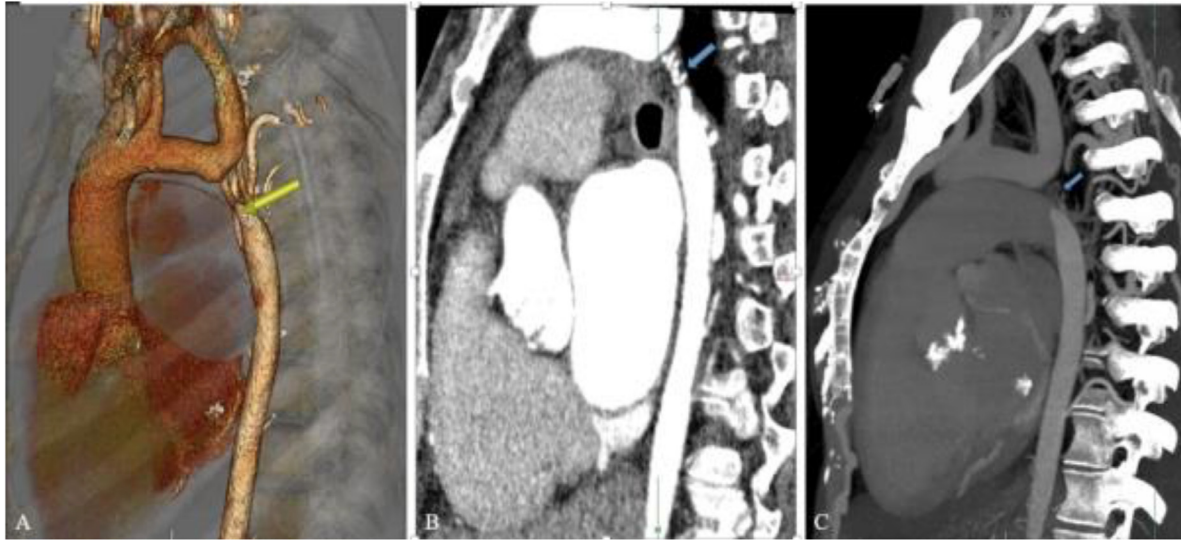


Fig. 1(A) – A3D volume rendering image, (B) postcontrast sagittal image, and (C) sagittal maximum intensity projection; demonstrating the absence of aorta distal to the origin of left subclavian artery (interrupted aorta) with multiple collaterals in the region of the absent aorta (arrow).

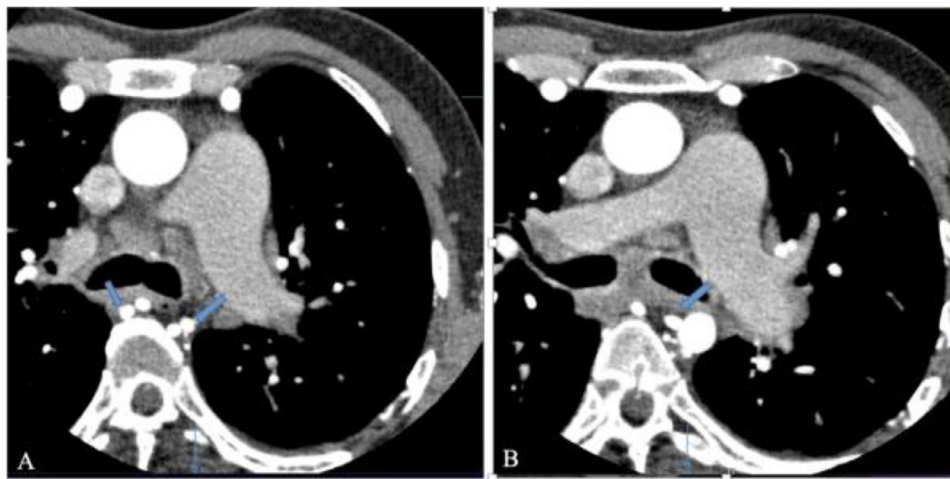


Fig. 2 – (A and B) Axial CT angiogram images showing absence of descending aorta in image A, the area is replaced by multiple collateral vessels (arrow), B (just distal to image A) showing descending aorta with multiple collaterals(arrow).

mm Hg, respectively. Heartbeats per minute ranged from 60 to 102. At the third to the fourth left intercostal area, a grade 2 systolic murmur was audible. There was no radioradial or radiofemoral delay. Dorsalis pedis pulse was palpable. T wave alteration and complete right bundle branch block were both visible on the electrocardiogram. A normal right atrium and right ventricle were seen on a color Doppler echocardiogram, along with an enlarged left atrium and left ventricle. Around the descending aorta, there were multiple collateral branches and a discontinuous aortic arch distal to the left subclavian artery. The abdominal aorta's flow pattern revealed a modest, sluggish wave. Blood test results yielded normal results with no significant finding on the renal function test. Coarctation

of the aorta was suspected and CT angiography was sent for (Fig. 1), (Fig. 2).

Due to the presence of extensive collaterals and being asymptomatic other than hypertension the patient chose to undergo noninvasive treatment and was managed with anti-hypertensive medication Figs. 1 and 2.

Discussion

IAA is a complex and uncommon congenital cardiovascular disorder. It often first appears in children, and adult patients are exceedingly rarely affected. The first IAA case was de-

scribed by Steidele et al [4] in 1778. A review done in 2021 by Jiang et al [5] reported only 25 cases of isolated IAA reported in the literature in the past 20 years. We present a rare case of isolated IAA presenting with hypertension.

IAA can be divided into 3 categories clinically based on where the interruption occurs. Type A (40%): The left subclavian artery's distal end has an aortic arch disruption. Type B (55%-60%): the interruption occurs between the left subclavian artery and the left common carotid artery and is closely linked to the DiGeorge syndrome and the deletion of chromosome 22q11.2. Type C (4%): There is an obstruction between the left common carotid artery and innominate artery [3].

Nearly all instances have a neonatal diagnosis and have severe congestive heart failure with quick clinical decline. 90% of affected infants die during the first year of life if untreated, the majority within the first few days. The presentation in the rare accounts in adults differs significantly from that in children and might range from no symptoms to refractory hypertension, headache, claudication, malaise, differential blood pressure in the arms and legs, and congestive heart failure [6].

The ability to survive into adulthood depends on the creation of a vast collateral network, which is necessary for the preservation of distal flow. This collateral network must have a ductus arteriosus that gradually shuts during childhood to grow. The distinctions between children and adults mentioned above provide credence to the idea that in some instances, the initial defect is a coarctation that progresses to full occlusion of the lumen [7]. Clinical and anatomical similarities between Type A IAA and severe coarctation may exist, with aortic coarctation being significantly more common and accounting for 4%-6% of all congenital cardiac abnormalities [8]. Undiagnosed aortic coarctation may have progressed to total blockage given the form and location of the obstruction distal to the left subclavian artery. This can explain the formation of extensive collaterals supplying the descending aorta and the late presentation. Differences and similarities exist between adult type A IAA and CoA. The distinction between the two is whether the arterial lumen is continuous or not, but they are similar in that secondary hypertension and extensive collateral circulation are present as well [9].

Similar to neonates or infants, adults with IAA often have a single-staged operation that involves a deep hypothermic circulatory arrest, rebuilding of aortic arch continuity, and repair of heart abnormalities. However, surgery can be challenging when there is a myriad of collateral circulation because there is a significant chance of bleeding, and the collateral vessels are prone to atrophy and atherosclerosis, which increases the risk of bleeding [5]. Since the postoperative therapy of IAA is similar to that of coarctation of the aorta, Jiang et al evaluated the pertinent data for coarctation of the aorta and discovered that 68% of patients still have hypertension following surgery. The majority of documented IAA cases undergoing surgical reconstruction show an improvement in patient's symptoms following surgery. Eleven patients refused surgery and got antihypertensive therapy, but no problems were noted, according to the review by Jiang et al [5]. In other words, the therapeutic

effects of antihypertensive drugs are comparable to those of surgical correction. Antihypertensive medication without surgical intervention is also reasonable when taken into account, along with the fact that hypertension is the main symptom and those patients experience fewer other symptoms.

Conclusion

Complete interruption of the aortic arch in an adult is a rare diagnosis. The interruption can be due to congenital causes or complete blockage after coarctation of the aorta. However, both require a significant amount of collateral for the patient to survive. Although surgical management is warranted, numerous cases are asymptomatic and benefit from medical management provided that the collaterals are developed extensively.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

REFERENCE

- [1] Shirani S, Soleymanzadeh M. Diagnosis of aortic interruption by CT angiography. *Pol J Radiol* 2013;78(1):72–4.
- [2] Franconeri A, Ballati F, Pin M, Carone L, Danesino GM, Valentini A. Interrupted aortic arch: a case report. *Indian J Radiol Imaging* 2020;30(1):81–3.
- [3] John AS, Schaff HV, Drew T, Warnes CA, Ammash N. Adult presentation of the interrupted aortic arch: case presentation and a review of the medical literature. *Congenit Heart Dis* 2011;6(3):269–75.
- [4] Hudsmith LE, Thorne SA, Clift PF, de Giovanni J. Acquired thoracic aortic interruption: percutaneous repair using graft stents. *Congenit Heart Dis* 2009;4(1):42–5.
- [5] Jiang Y, Wang C, Jiang X, Chen S. Is surgery necessary for adults with isolated interrupted aortic arch?: Case series with literature review. *J Card Surg* 2021;36(7):2467–75.
- [6] Silva J, Guiomar N, Silva MP, Caeiro D, Gama V. Interrupted aortic arch in an adult. *Eur J Case Rep Intern Med* [Internet] 2017;4(9). [cited 2022 Jul 11] Available from: <https://www.ejcrim.com/index.php/EJCRIM/article/view/692>.
- [7] Messner G, Reul GJ, Flamm SD, Gregoric ID, Opfermann UT. Interrupted aortic arch in an adult single-stage extra-anatomic repair. *Tex Heart Inst J* 2002;29(2):118–21.
- [8] Hoffman JIE, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002;39(12):1890–900.
- [9] Ren SX, Zhang Q, Li PP, Wang XD. Difference and similarity between type A interrupted aortic arch and aortic coarctation in adults: two case reports. *World J Clin Cases* 2022;10(11):3472–7.