Clinical Case Report

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Isolated interrupted of aortic arch diagnosed using CT angiography

A case report and literature review

Guang-wen Chen, MS, Hang Li, MS, Hong Pu, MD*

Abstract

Rationale: Interrupted aortic arch (IAA) is defined as a complete luminal and anatomical interruption between the ascending and descending aortas. The majority of patients with IAA rarely reach adulthood. In most cases, IAA is associated with an intracardiac malformation, such as ventricular septal defect, bicuspidaortic valve, and patent ductus arteriosus. We reported a 70-year-old man with isolated IAA diagnosed using computed tomography angiography (CTA) without previous intervention.

Patient concerns: The patient presented with edema of the lower extremities and productive cough.

Diagnoses: CTA displayed an interruption of the aortic arch distal to the left common carotid artery (type B) and extensive collateralization.

Interventions: The patient refused surgery and underwent conservative therapy with follow-up examinations because of the extensive collateral vascularization.

Outcomes: The patient is still currently undergoing conservative therapy with follow-up examinations.

Lessons: IAA can present in adulthood with different clinical symptoms. Careful physical examination of the lower and upper peripheral pulses in adults with a chief complaint of hypertension is of outmost importance. CTA is a useful noninvasive imaging modality for the morphologic evaluation of aortic interruption and collateral circulation.

Abbreviations: CTA = computed tomography angiography, IAA= interrupted aortic arch.

Keywords: collateral circulation, computed tomography angiography, interrupted aortic arch

1. Introduction

Interrupted aortic arch (IAA) is a severe congenital malformation of the aortic arch, which involves 3 per million live births.[1] IAA is defined as a complete interruption of aortic lumen between the ascending and descending aortas.[2] It is usually detected in early childhood or neonatal period, but there will be a few adult cases with IAA and other intracardiac malformations, such as ventricular septal defect and patent ductus arteriosus.[3] Therefore, it rarely occurs as an isolated lesion. Previous studies presented an isolated IAA case in a < 60-year-old individual without extensive collateral circulation on computed tomography angiography (CTA).[4,5] In this case report, we present a case of a 70-year-old man with isolated IAA diagnosed using CTA.

Editor: N/A.

GWC and HL both equally contributed to this study.

The authors have no funding and conflicts of interest to disclose.

Department of Radiology, Sichuan Provincial People's Hospital, 32# second section of first ring road, Chengdu, Sichuan, China.

** Correspondence: Hong Pu, Department of Radiology, Sichuan Academy of Medical Sciences and Sichuan Provincial People's Hospital, 32# Second Section of First Ring Road, Qingyang District, Chengdu, Sichuan 610072, China (e-mail: ph196797@163.com).*

Medicine (2018) 97:21(e10569)

Received: 22 December 2017 / Accepted: 5 April 2018

http://dx.doi.org/10.1097/MD.0000000000010569

2. Case report

The institutional review board of our institution approved this study, and written informed consent was obtained from the patient. A 70-year-old man with edema of the lower extremities and productive cough was admitted to the hospital. He had a history of hypertension for 20 years and chronic obstructive pulmonary disease for 10 years. On admission, his blood pressure was 182/108 and 123/62 mm Hg in the right and left arms, respectively. Heart rate was between 60 and 135 bpm. Grade 2 systolic murmur was heard at the 3 to 4 left intercostal space. Inspiratory rales were remarkable upon chest auscultation. Severe pitting edema was noted on the lower extremities. The echocardiogram showed dilatation of the ascending aorta (62 mm), left ventricular enlargement, moderate systolic dysfunction of the left ventricle, and moderate mitral regurgitation.

Because of suspicion of ascending aorta aneurysm or coarctation of the aorta, he was referred to the radiology department to confirm the aortic morphologic condition. CTA showed an interruption of the aortic arch distal to the left common carotid artery (type B) (Fig. 1). The interruption consisted of a 2-mm thick septum. The enlargement of the vertebrobasilar artery, left subclavian artery, right internal mammary artery, bilateral intercostal arteries, bilateral inferior phrenic artery, thoracoabdominal wall artery, and other arterial branches provided collateral flow to the distal aorta and large branch arteries of the aorta. For example, the coeliac trunk was provided with collateral circulation from the right internal mammary and left hepatic arteries; and the right external iliac artery was provided with collateral circulation from the thoracoabdominal wall and right superficial epigastric arteries (Fig. 2). CTA also demonstrated dilatation of the ascending aorta

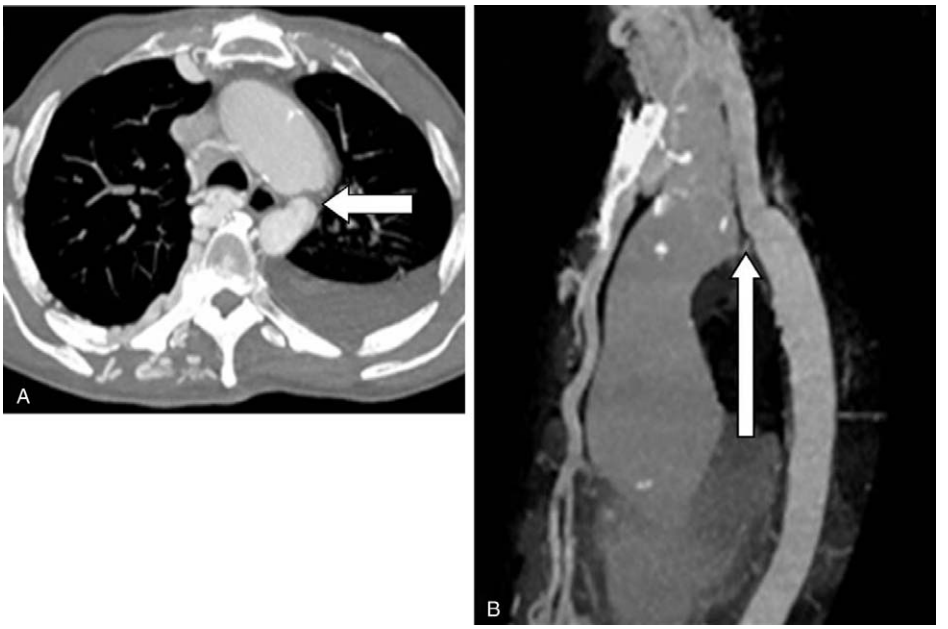

Figure 1. Axial and oblique sagittal maximum intensity projection images in a 70-year-old man who is subsequently diagnosed with type B interruption of the aortic arch. Axial (A) and oblique sagittal (B) maximum intensity projection images of the thoracic aorta show an interruption of the aortic arch (white thick arrow) distal to the origin of the left common carotid artery.

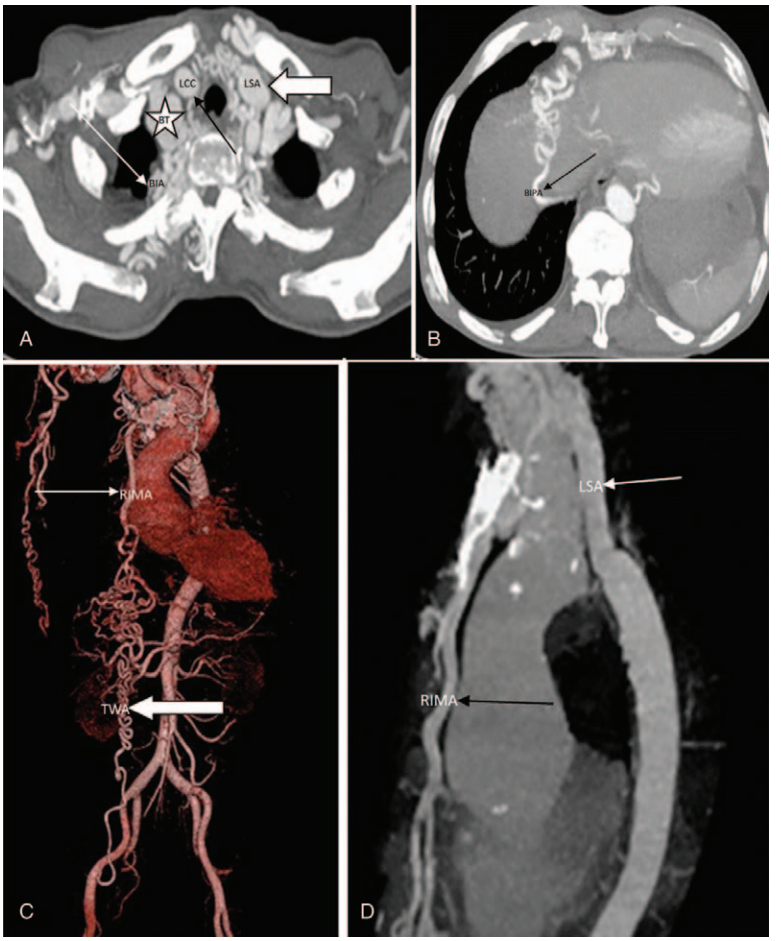

Figure 2. Axial and oblique sagittal maximum intensity projection and volume-rendered images in a 70-year-old man with type B interruption of the aortic arch. Axial maximum intensity projection (A, B) and volume-rendered from an anteroposterior projection (C) images show the dilated brachiocephalic trunk (BT) (A, white star), left common carotid artery (LCC) (A, black arrow), left subclavian artery (LSA) (D, white thin arrow; A, white thick arrow), right internal mammary artery (RIMA) (D, black arrow; C, white thin arrow), bilateral intercostal arteries (BIA) (A, white thin arrow), bilateral inferior phrenic artery (BIPA) (B, black arrow), and thoracoabdominal wall artery (TWA) (C, white thick arrow).

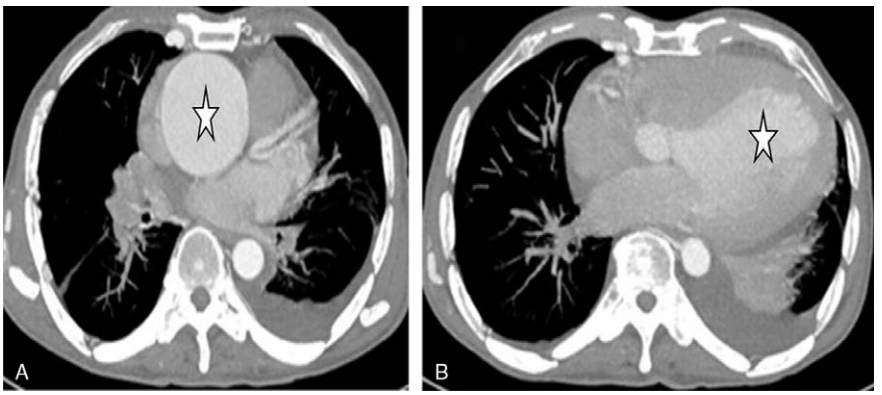

Figure 3. Axial maximum intensity projection image in a 70-year-old man with type B interruption of the aortic arch. Axial (A, B) maximum intensity projection image shows dilatation of the ascending aorta (A, white star) and left ventricular enlargement (B, white star).

and left ventricular enlargement (Fig. 3). However, the patient refused surgery and underwent conservative therapy with follow-up examinations because of the extensive collateral vascularization.

3. Discussion

IAA in an adult is extremely rare and often missed in patients with secondary hypertension. This case displays the complete interruption between the aortic arch and thoracic aorta. In general, this interruption of the blood flow in most cases is expected to be fatal. However, patients with isolated IAA may survive until adulthood because of the extensive collateral vessels joining the descending aorta.

Based on the position of the interruption, IAA is classified into three groups: Type A interruption is positioned distal to the left subclavian artery; Type B interruption is positioned distal to the origin of the left common carotid artery; and Type C interruption is positioned proximal to the origin of the left common carotid artery.[6] In neonates, 53% of the cases are type B, followed by types A (43%) and C (4%).[7] However, the presentation of IAA in adulthood was obviously different. In a review of 38 cases of IAA in adulthood, 79% of these patients were type A, followed by types B and C with 16% and 3%, respectively.[8] The common characteristic among these survivors is the presence of extensive collateral vessels that connect distal blood flow. In our case, the descending thoracic aorta is supplied by the left subclavian artery, which is provided by the vertebrobasilar system and extensive collateral vessels from the right internal mammary artery, bilateral intercostal arteries, bilateral inferior phrenic artery, thoracoabdominal wall artery, and other arterial branches.

The main treatment for IAA is surgical intervention. As for neonates and infants, several surgical means, such as end-to-end or end-to-side anastomosis, exist. While older children and adults can undergo end-to-end anastomosis, the older population can undergo placement of interposition grafts. The main reason for this surgical option is the risk of neurological events, such as paraplegia. However, massive collateral circulation prevents this kind of complication in adult patients with very severe true interruptions of the aorta.[9] Therefore, the display of extensive collateralization for IAA in adult is beneficial for surgery.

Transthoracic echocardiography is the initial imaging modality, but it is mandatory to differentiate IAA from coarctation of the aorta. Therefore, other diagnostic techniques, such as computed tomography and magnetic resonance imaging of the heart, are preferred modalities for the diagnosis of IAA. To our knowledge, this case is among the oldest adults with isolated IAA to live to an age of 70 years without any previous intervention. Moreover, we are also first to clearly display the form of extensive collateral vessels on CTA.

4. Conclusion

It should be considered that IAA can present in adulthood with different clinical symptoms. Careful physical examination of the lower and upper peripheral pulses in adults with a chief complaint of hypertension is of outmost importance,[10] because this provides the first clue to the diagnosis of IAA. CTA is a useful noninvasive imaging modality for the morphologic evaluation of the aortic interruption position and collateral vessels.

Author contributions

Conceptualization: Guang-wen Chen, Hong Pu.
Data curation: Guang-wen Chen, Hang Li, Hong Pu.
Formal analysis: Guang-wen Chen, Hang Li, Hong Pu.
Funding acquisition: Guang-wen Chen, Hong Pu.
Investigation: Guang-wen Chen, Hong Pu.
Methodology: Guang-wen Chen, Hang Li, Hong Pu.
Project administration: Guang-wen Chen, Hong Pu.
Resources: Guang-wen Chen, Hang Li, Hong Pu.
Software: Guang-wen Chen, Hang Li, Hong Pu.
Supervision: Guang-wen Chen, Hang Li, Hong Pu.
Validation: Guang-wen Chen, Hang Li, Hong Pu.
Visualization: Guang-wen Chen, Hang Li, Hong Pu.
Writing – original draft: Guang-wen Chen, Hang Li, Hong Pu.
Writing – review & editing: Guang-wen Chen, Hang Li, Hong Pu.

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