

Interrupted aortic arch: A case report

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Abstract

Interrupted aortic arch diagnosed in adult age is a rare entity, with only a few cases published in the literature. Most of them are classified as type A interrupted aortic arch and differential diagnosis is associated with severe chronic coarctation. We present a case of a 52-year-old woman accessed to the emergency department for chest and right upper limb pain that increased in the last days. She underwent a computed tomography angiogram showing interruption of the aortic arch, distal to left subclavian artery origin, large bilateral collateral vessels connecting subclavian arteries to descending aorta with multiple voluminous aneurysms, a bicuspid aortic valve, dilatated tubular segment of ascending thoracic aorta, and a suspected atrial septal defect. A nonsystematic literature review regarding these conditions has been performed.

Key words: Aneurysm; bicuspid aortic valve; coarctation of the aorta; computed tomography angiogram; interrupted aortic arch

Background

Interruption of the aortic arch (IAA) is a malformation characterized by the congenital absence of luminal continuity between ascending and descending thoracic aorta.^[1] Three different types of IAA have been classified based on the site of the interruption, namely, type A when the interruption is distal to the left subclavian artery origin; type B when the interruption is between the left subclavian artery and the left common carotid artery; and type C when the interruption is between the innominate and the left common carotid artery.^[2] Differential diagnosis is with coarctation of the aorta (CoA), defined as a focal eccentric narrowing of an aortic segment, usually distal to the left subclavian artery and close to the insertion of arterial duct.^[3]

Herein, we present a case of adult IAA, voluminous aneurysms of subclavian collateral arteries, bicuspid aortic valve, and enlargement of ascending thoracic aorta.

Procedure

A 52-year-old woman with a not better-specified history of thoracic great vessel malformation presented to the emergency department upon indication of her primary care physician. She referred to experiencing pain in the chest and right upper limb since the last 10 months, more intense in the last days.

Because of her history and the intensification of symptoms, she underwent an ECG-synchronized computed tomography angiogram (CTA) of the heart and thoracic aorta and a subsequent, not ECG-synchronized scan extended from brain to pelvis to exclude other vessels malformations.

The CTA study showed a complete interruption of aortic arch distally to subclavian artery origin [Figures 1 and 2A],

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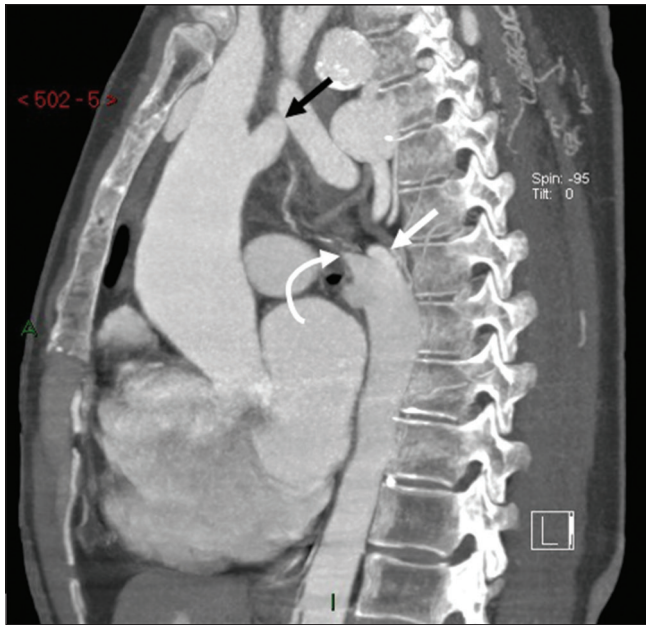


Figure 1: Maximum intensity projection of the reformatted sagittal image of the thoracic aorta shows interruption of the aortic arch just distal to the subclavian artery origin (black arrow), interruption of descending aorta (white arrow) and fibrotic and stretched arterial duct (curved white arrow)

reported as type A IAA.^[2] Distance between the interrupted aortic arch and the descending segment of the thoracic aorta was more than 4.5 cm. Arterial duct appeared fibrotic and partially calcified [Figure 1 and 2A]. The thoracic descending aorta was bilaterally refilled, distally to interruption, by collaterals of subclavian arteries arisen after vertebral arteries origin [Figure 2B]. Both collateral arteries presented proximal voluminous aneurysms, the biggest on the right side with endoluminal dissection flap [Figures 2B and 3A] and at least three on the left side [Figure 2A]. Collateral circulation was evident from the internal mammary artery, in particular, the left one [Figure 2A and B]. Aortic root showed a bicuspid aortic valve morphology [Figure 3C] with thick and calcified valvular flaps and an enlargement of the tubular segment of the ascending thoracic aorta [Figure 3B] was reported. No other aneurysm was detected in the rest of the body.

Conclusion

We reported a case of type A IAA with the complete anatomic discontinuity of aortic arch immediately distal to subclavian artery origin and with a conspicuous distance between the interrupted aortic arch and the descending segment of the thoracic aorta. Severe cases of CoA may have a complete loss of luminal continuity and progression into the complete obstruction as well.^[4] As previously reported, aorta morphology may help in distinguishing between IAA and CoA.^[4] In CoA, the distal aortic arch usually extends beyond the origin of the left subclavian artery over several centimeters, differently from our case where it discontinues

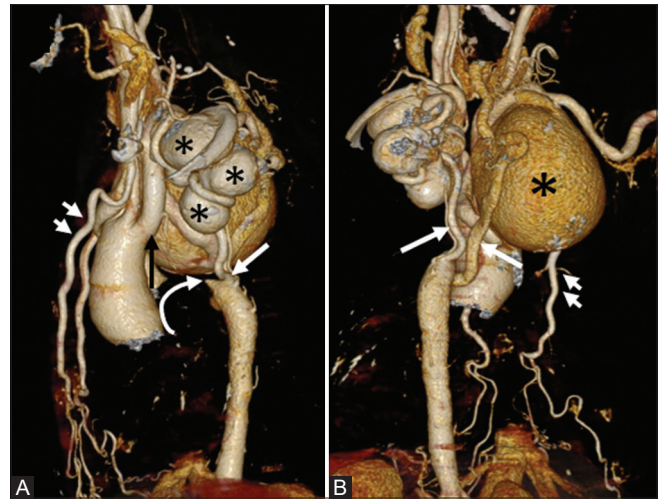


Figure 2 (A and B): (A) Sagittal volume-rendered-image shows cranial (black arrow) and caudal (white arrow) sites of interruption, the fibrotic and stretched arterial duct (curved arrow), enlarged internal mammary arteries (arrowheads) and aneurysms of collateral vessels of left subclavian artery (asterisks). (B) Posterior coronal volume-rendered-image shows right aneurysm (asterisk) proper of a subclavian artery collateral vessel, large bilateral collateral vessels connecting subclavian arteries with descending segment of the thoracic aorta (white arrows) and enlarged internal mammary arteries (arrowheads)

at the level of the left subclavian artery. Furthermore, in chronic CoA, both extremities usually present elongated, assuming a rat-tail appearance. However, in our case, both appeared morphologically round at the level of the interruption. In IAA, usually, ascending aorta presents a small caliber and a straight course to its branches; however, in our case, normal curvature and enlargement of ascending aorta were described. This may due to different embryological origin of the three types of IAA that determine different morphological aspect of thoracic aorta in the ascending segment; the result of type A IAA from abnormal regression of the left fourth arch segment late in development, after the left subclavian artery has ascended to its normal position,^[1] may contribute in maintaining the normal ascending aorta curvature. Furthermore, hypoplasia of the aortic arch and isthmus are described in most CoA patients too.^[1] CoA is more frequently diagnosed in the adult population compared to IAA. However, type A IAA is the commonest, among the three types of IAA, in adults' patients (79%). The average age of IAA diagnosis in adults is 39.4 years old and most of them are men (74%).^[5] CoA is reported more commonly in males than females (51%).^[6] Around 80% of CoA occurs as an isolated entity but it can be associated with other congenital cardiovascular lesions.^[1,3] Patency of the arterial duct (98%) and a ventricular septal defect (90%) are the most common congenital cardiovascular lesions associated with IAA.^[4,7] In our case, the arterial duct appeared fibrotic and partially calcified and ventricular septal defect was absent. Unlike type B IAA, the conal outlet septum is usually not deviated or misaligned, so subaortic stenosis is not present, and association with a ventricular

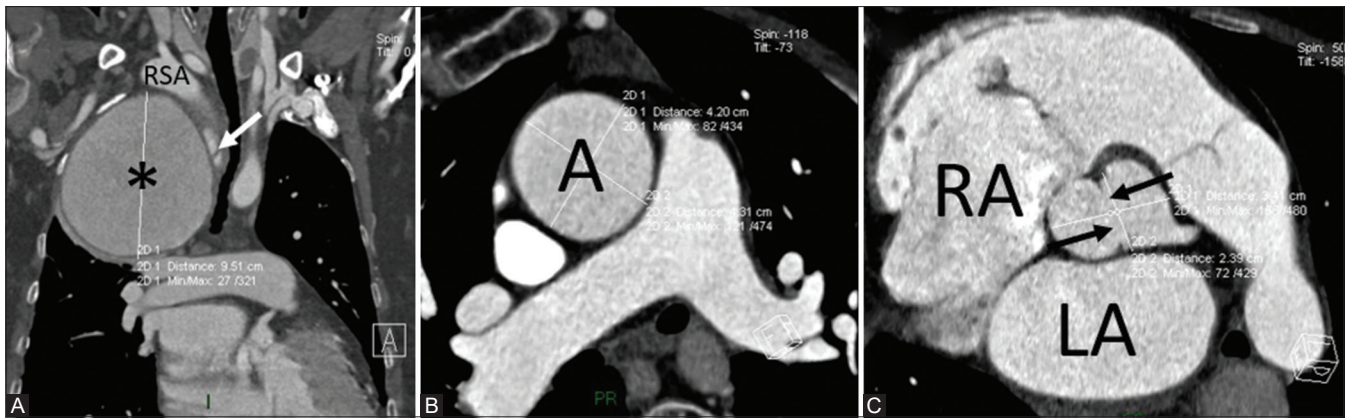


Figure 3 (A-C): (A) Maximum intensity projection of the reformatted coronal image of maximum craniocaudal extension of the right aneurysm (asterisk), the right subclavian artery (RSA) and the dissection flap of the collateral artery proximal to the aneurysm (white arrow). (B) Reformatted axial image shows enlarged ascending aorta (marked a) measured at the level of pulmonary artery trunk. (C) Reformatted view through the plane of the aortic valve shows bicuspid aortic valve morphology (black arrows). LA, left atrium; RA, right atrium

septal defect or an anomalous origin of the subclavian artery is unusual.^[1] However, the bicuspid aortic valve was present which is common in all aortic arch anomalies and occurs in 33% to 50% of IAA^[7] and in 27% to 85% of CoA.^[8]

Mainly caused by the increased velocity of blood flow in the site of aortic narrowing, CoA may be responsible for aneurysm formation, especially post-coarctation.^[9] In our case, aneurysms were described in ascending thoracic aorta and collateral vessels of the subclavian artery but not in the descending segment of the aorta.

In our case report, the age at the time of diagnosis, the absence of a patent arterial duct and a ventricular septal defect, the presence of aneurysms could suggest a chronic evolution of a severe CoA more than an IAA.

On the other side, the conspicuous distance between the aortic arch and descendent aortic segment, the round morphology of the interrupted segment of the thoracic aorta, the absence of a post-coarctation enlargement, the lack of an extension of interrupted aortic arch beyond the origin of the left subclavian artery make the diagnosis of type A IAA more confident than CoA. Nevertheless, differential diagnosis between type A IAA and severe CoA maybe not always feasible with imaging. Therefore, we recommend extensive imaging to find out other cardiac or extra-cardiac vascular abnormalities.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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