Chinese Medical Journal®

Interrupted aortic arch with multiple vascular malformations

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To the Editor: A 66-year-old male patient was referred to the First Hospital of Lanzhou University with complaints of sudden dizziness, with nausea and emesis. His medical history revealed heavy smoking for 40 years, and no other relevant information was found.

Computed tomography angiography (CTA) revealed an interrupted aortic arch (IAA) with complete discontinuity of the aortic lumen, distal to the origin of the left common carotid [Figure 1]. The density of the ascending aorta (AA) and descending aorta (DA) were different; both common carotid arteries (CCAs) and dilated tortuous left subclavian artery (SCA) originated from the distal AA and DA, respectively. The right SCA originated from the DA, posterior to the trachea and esophagus. Abundant systemic collaterals formed pathways among the cervical vessels. And a persistent left-sided superior vena cava (PLSVC) was observed. A cerebral CTA was performed as dizziness was suspected; it showed that a part of the dominant left vertebral artery was not in the intervertebral foramen, but originated from a collateral vessel of the left CCA. Both the internal carotid siphons were slender, and the fetal posterior cerebral artery was also present. Based on the available evidence, we concluded that the patient had a type B IAA associated with PLSVC and an aberrant right subclavian artery (ARSA). After a week of medication, the patient's condition improved. Although surgery, such as stent placement, is effective for IAA, the patient chose conservative management due to the surgical sequelae risk in elderly adults.

IAA is a congenital cardiovascular malformation with a high mortality rate pre-natally and during infancy, without surgical correction. It is rarely identified among adults.[1] IAA occurs in about $3/10^6$ live births per year and accounts for 1% of congenital heart disease cases. It is classified into three types according to the location of the interruption: type A IAA, the arch interruption is distal to the origin of the left SCA; type B IAA, the arch interruption is between the site of origin of the left CCA and SCA; and type C IAA, the arch interruption is between the site of origin of the brachiocephalic and left CCA.[2] This is a case report on type B IAA concurrent with PLSVC and ARSA. The factors that enabled our patient's survival drew our interest. We found that abundant collaterals played a pivotal role in blood circulation, which was significant for ensuring our patient's survival to adulthood. The collaterals may represent compensatory growth during the fetal stage, but further research on the mechanisms underlying the formation of such collaterals is warranted.

Dizziness experienced by our patient may correspond to cardiovascular malformations. Previous studies suggested that central nervous system diseases were the principal cause of dizziness in the elderly and cardiovascular diseases were secondary factors.[3] When episodes of dizziness are accompanied with an abnormal cardiac rhythm or heart rate, abnormal blood pressure, and changes in electrocardiograms or following chest distress and dyspnea, cardiovascular factors should be considered for dizziness. Our patient may have had transient ischemia due to vertebrobasilar artery malformation and vascular aging. Assessment of IAA mostly depends on the various imaging techniques used for detection. Echocardiographic examination is economical and efficient, always used for IAA diagnosis during the fetal stage, and also helps to identify hypoplasia like a high-risk group for 22q11 deletion and possible associated IAA and then post-natal intervention. Despite its limitations in evaluating the aortic arch and the DA, echocardiography can also be used for adults for detecting concomitant cardiac abnormalities.[4] Angiography, though invasive, is considered the gold standard for IAA diagnosis, and interruption of the aortic arch with a blind cul-de-sac of the proximal AA and a blind-ending at the superior margin of the DA are clearly observed. The shape and size of the aortic arch and the collateral vessel branches can also be determined. Furthermore, magnetic resonance angiography (MRA) and CTA are reliable non-invasive examinations with multi-planar capabilities and three-dimensional imaging. With these imaging modalities, the morphologic characteristics of IAA and associated

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Quick Response Code:

Website: www.cmj.org

DOI: 10.1097/CM9.0000000000000438

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Chinese Medical Journal 2019;132(19)

Received: 30-05-2019 **Edited by:** Ning-Ning Wang

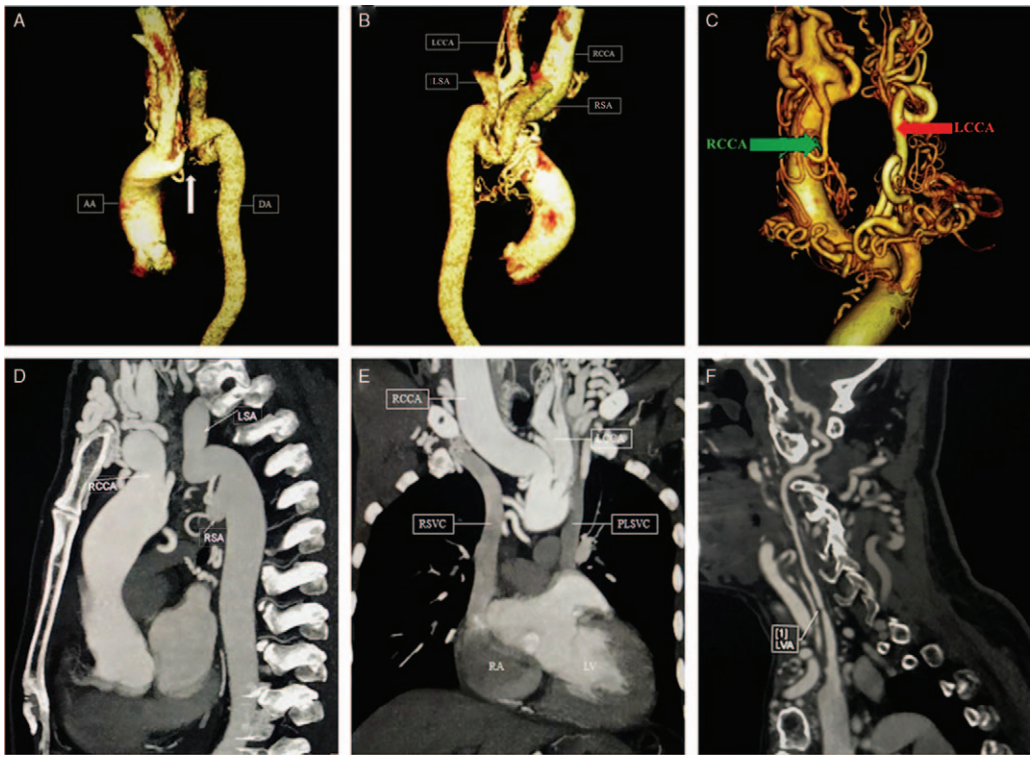

Figure 1: Three-dimensional reconstruction showed type B IAA (A–C). Maximum intensity projection illustrated IAA associated with persistent left-sided superior vena cava and aberrant right subclavian artery (D–F). AA: Ascending aorta; CCA: Common carotid artery; DA: Descending aorta; IAA: Interrupted aortic arch; LVA: Left vertebral artery; PLSVC: Persistent left-sided superior vena cava; RSVC: Right superior vena cava; SA: Subclavian artery.

abnormalities can be observed, and the airway and lungs can be evaluated simultaneously. MRA is superior due to the use of hypo-toxic contrast agent and non-ionizing radiation. In contrast, CTA is cheaper and less time consuming, with higher spatial resolution and less susceptibility to artifacts. In our case, the use of CTA provided more visual information and led to better identification of the anatomic relationships between IAA and sophisticated associated malformations, resulting in a deeper understanding and description of the patient's condition.

In conclusions, our patient with type B IAA associated with PLSVC and ARSA has survived to adulthood due to collateral vessels providing sufficient blood flow for the distal aorta. His dizziness may be caused by vertebrobasilar artery malformation and vascular aging. Notably, CTA plays a crucial role in the diagnosis and evaluation of cardiovascular malformations, and combined diagnostic imaging should be performed to aid in the identification of IAA and in surgical approach.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Funding

This work was supported by the grants from the Gansu Province Youth Science and Technology Foundation (No. 18JR3RA364), the Gansu Provincial Health Industry Research Projects Foundation (No. GSWSKY2016-34), and First Hospital of Lanzhou University Hospital Foundation (No. ldyyyn2015-06).

Conflicts of interest

None.

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How to cite this article: Xiang XR, Chen ZX, Zhang L, Lei JQ, Guo SL. Interrupted aortic arch with multiple vascular malformations. Chin Med J 2019;132:2386–2387. doi: 10.1097/CM9.0000000000000438