Case Report

320-slice CT angiography of an interrupted aortic arch patient relying on collaterals: An addition to classification criteria

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\begin{abstract}
A 13-year-old female patient presented with chest pain had no history of heart disease or trauma. She was finally diagnosed with the interrupted aortic arch by 320-slice CT angiography, without intracardial malformations or patent ductus arteriosus. Her descending aortic blood was supplied by plentiful collateral circulation on the chest. According to literature reports, patients with complete interruption of the aortic arch rarely live 10 years without surgical intervention. More particularly, this case does not fit the current classification systems.

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Introduction

Interrupted aortic arch (IAA) is a rare congenital aortic arch anomaly defined as a luminal and anatomical interruption between the ascending and descending aortas. It is usually complicated with complex intracardiac malformations, such as patent ductus arteriosus, ventricular septal defect, or bicuspid aortic valve. It accounts for fewer than 1% of infant congenital heart disease with an incidence of 19 per 1 million live births [1–4]. Absence of surgical intervention for the affected infants at an early stage would lead to a 90% mortality rate in the first year of life [5]. Few cases demonstrate that IAA is the isolated finding without another associated cardiac defect have been reported on CT angiography (CTA). This case without patent ductus arteriosus, therefore, does not belong to the classic Celoria anatomical classification standards.

Case report

A 13-year-old female came to a clinical visit with chest pain after exercise but had no history of heart disease or trauma. She had slight dizziness sometimes in the previous 12 months. Physical examination: heartbeat 83/min, upper limb blood pressure 148/90 mm Hg, lower limb blood pressure 128/78 mm Hg. Saturation of Hemoglobin with Oxygen SpO\textsubscript{2}...
aorta, many distorted feeder vessels from the neck and the upper thoracic involved to set up a vascular network which was found connected to the descending aorta. Two-sided internal thoracic artery, intercostal artery, costocervical trunk, and 2-sided external thoracic artery all had enlarged and involved (Fig. 2).

She was an outpatient from a poor family. We later learned that her parents did not consider surgery and did not have hospitalization. The rarity of this case is that no special treatment was received in 13-year survival before diagnosed. The outpatient doctor gave her oxygen therapy to relieve her symptoms and told her: (1) to avoid high-intensity exercise and pay attention to rest. (2) Regularly review echocardiography and angiography. (3) Outpatient follow-up. If she does not have timely surgery, the chance of complications increases over time, including aneurysm, dissecting aneurysm, heart failure, syncope, cerebrovascular accident, ischemic peripheral neuropathy, and myocardial infarction.

Discussion

The CT angiography is one advantage technology of the 320-slice CT than other MSCT. Its faster scan speed can reduce artifacts caused by cardiovascular motion, even more, its powerful software special optimization for vascular image reconstruction, so we can obtain high-quality images in one-stop. Interrupted aortic arch (IAA) can be caused by the dysplasia of fourth and sixth pairs of arterial arches during embryonic

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**Fig. 1** – The patient’s chest radiograph did not show a complete aortic arch structure (thick arrow); an obvious intercostal artery indentation was seen at the lower edge of the rib (thin arrow).

**Fig. 2** – Continuity of the aorta interrupted at the initial distal part of the left subclavian artery without other cardiovascular abnormalities (thick arrows). Bilateral internal thoracic artery, intercostal artery (thin arrows), costocervical trunk, and bilateral external thoracic artery all had enlarged; multiple distorted arteries appeared in the back and upper thoracic, and collateral circulation network formed.
development; the related teratogenic factors and mechanisms remain unclear [6]. General classification standards for IAA were put forward by Celoria in 1959 [7], who summarized data from 28 cases with IAA, and classified the ailment into 3 types according to the interrupted positions. Type A (55%): aortic interruption occurs distally to the origin of the left subclavian artery. Type B (40%): aortic interruption occurs between the left common carotid artery and the origin of the left subclavian artery. Type C (5%): aortic interruption occurs between the innominate artery and the origin of the left common carotid artery. This case did not belong to any of the mentioned types. Indeed, an IAA patient survived only by collateral circulation that is not included in the 28 cases used to work out classification standards. According to the developmental condition of the right subclavian artery, Oppenheimer-Dekker A divided the Type A IAA into 3 subtypes (A, A1, or A2) based on Celoria classification standards [8]. However, the IAA discussed in this article was still not included.

This rare case, without intracardial malformations or patent ductus arteriosus, does not fit mentioned classifications. We propose an additional type to be added to the existing classification to account for this case and others like it. Our modified classification is "two categories and five types." The 2 categories should include cyanosis and noncyanosis groups. The cyanosis group encompassing types A, B, and C of Celoria. The noncyanosis group is divided into types D and E (Fig. 3). These adjusted classification standards conform to the routine classification of congenital heart diseases, and more comprehensive than Celoria classification. This case might be described as "type E, noncyanosis category."

**Conclusion**

The CTA could visually show intracardiac malformations, interrupted position of IAA, the collateral circulation, and measurements of relevant arteries which are useful for surgical planning. Therefore, CTA before treatment for patients with IAA may be necessary. Developments in medical imaging technology could help us to observe diseases more accurately and make an addition to theories simultaneously. Our case is rare, but it reveals the deficiency of IAA classification that should hardly be ignored.

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