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Case Report

A rare case of right aortic arch with mirror-image branching and vascular ring in an adult patient☆,☆☆

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ABSTRACT

A right-sided aortic arch with mirror-image branching is a rare vascular anomaly, often associated with congenital heart defects. However, its occurrence with a vascular ring in the absence of cardiac abnormalities is extremely rare. We report the case of a 62-year-old male presenting with acute dyspnea, in whom CT angiography revealed a right-sided aortic arch with a vascular ring formed by an aortic diverticulum and the left brachiocephalic artery, causing mild tracheal and esophageal compression. Echocardiography confirmed the absence of cardiac anomalies. This case highlights a rare presentation of a right-sided aortic arch with a vascular ring in an adult patient. While often asymptomatic, such anomalies may contribute to airway or esophageal compression, warranting careful imaging evaluation and tailored management.

Introduction

A right aortic arch is a rare congenital vascular anomaly, with an estimated prevalence of 0.1% [1]. When accompanied by mirror-image branching, this condition is frequently associated with congenital heart defects, including tetralogy of Fallot [2]. In some rare cases, this vascular anomaly can lead to the formation of a vascular ring, where the aorta, along with other vessels encircles and potentially compresses the trachea and esophagus. This compression can cause symptoms such as dysphagia, dyspnea [3]. In this report, we present a very rare case of a right aortic arch with mirror-image branching and a vascular ring observed in adulthood, without any associated cardiac anomalies.

Case report

A 62-year-old male patient with a history of occasional dysphagia to solids presented to the emergency department with sudden-onset dyspnea. He denied chest pain, palpitations, or recent trauma. His vital signs were within normal limits,

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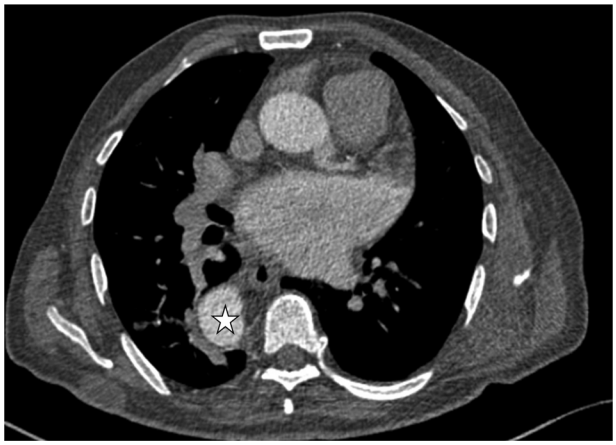

Fig. 1 – Axial contrast-enhanced CT image showing right-sided descending aorta (asterisk).

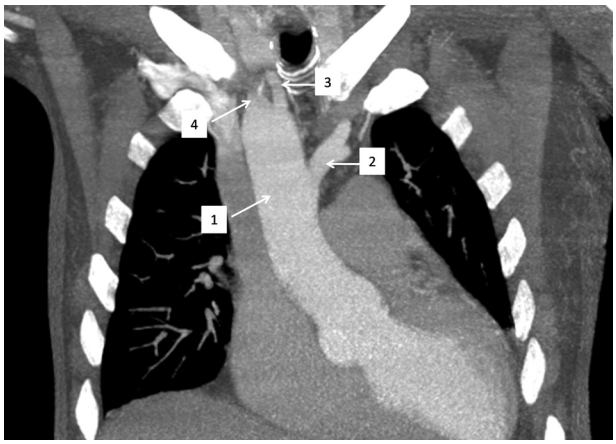

Fig. 2 – Coronal MIP reconstruction of contrast-enhanced CT image showing a right aortic arch (1) giving rise to the left brachiocephalic trunk (2), the right common carotid (3) and right subclavian arteries (4), consistent with a right-sided aortic arch with mirror-image branching.

and there were no signs of respiratory distress on examination. Initial investigations, including complete blood count, D-dimer levels, renal function tests, and electrocardiogram, were all unremarkable. Given the acute presentation, pulmonary embolism was suspected, and a thoracic CT angiography was performed. It shows good opacification of the pulmonary arteries, effectively ruling out embolism.

However, an incidental vascular anomaly was identified. The descending thoracic aorta was located on the right (Fig. 1), and the aortic arch was positioned to the right of the trachea, giving rise to the left brachiocephalic trunk first, followed by the right common carotid and right subclavian arteries, consistent with a right-sided aortic arch with mirror-image branching (Figs. 2 and 4). Additionally, a large aortic diverticulum was observed at the level of the aortic arch, forming a vascular ring in conjunction with the right-sided aortic arch and the left brachiocephalic artery (Figs. 3 and 5). This vascular ring caused mild extrinsic compression of both the trachea and esophagus, leading to a reduction in their calibers.

Although the patient's symptoms were initially attributed to pulmonary pathology, the presence of a vascular ring raised the possibility of airway compression syndrome caused by vascular abnormality as a contributing factor. Echocardiography confirmed the absence of additional cardiac anomalies. Given the mild nature of the compression and the absence of severe respiratory or digestive symptoms, a conservative management strategy was adopted. This consisted of regular outpatient clinical follow-up, including periodic physical examinations, assessment of respiratory and gastrointestinal symptoms, and imaging surveillance if clinically indicated. No surgical intervention was deemed necessary at this stage.

Discussion

A right-sided aortic arch is a rare vascular variant occurring in approximately 0.1% of the population (range 0.05%-0.2%), where the aortic arch courses to the right of the trachea [1]. It can be divided into at least 3 types: a mirror image branching, an aberrant left subclavian artery, and an isolated left subclavian artery [4].

A right-sided aortic arch with mirror-image branching is a vascular variant accounting for up to 59% of right-sided aortic arches, in which the aortic arch courses to the right of the trachea and gives rise to the left brachiocephalic trunk first, followed by the right common carotid and right subclavian arteries [5]. This variant is often observed without a vascular ring and is commonly associated with cyanotic congenital heart diseases, including tetralogy of Fallot, truncus arteriosus, tricuspid atresia, and transposition of the great arteries [4,6]. Our case is unique as it presents a right aortic arch with mirror-image branching and a vascular ring in an adult patient without any associated cardiac abnormalities.

The understanding of this anomaly is based on Edward's hypothesis regarding the double arch system during embryonic development [7]. In the formation of the double aortic arch, the normal left aortic arch develops when the portion of the right arch between the right subclavian artery and the descending aorta regresses completely. Conversely, when regression occurs in the same region of the left arch, a right aortic arch with mirror-image branching develops. In most of cases, this regression happens between the left ductus arteriosus and the descending aorta, resulting in mirror-image branching without a complete vascular ring (Edwards Type I). In extremely rare cases, the regression occurs between the left subclavian artery and the left ductus arteriosus. This leads to the left ductus extending from the descending aorta diverticulum to the left pulmonary artery, thus forming a vascular ring. As the ductus cannot be imaged directly, the imaging appearance of this arch anomaly resembles that of Edwards Type I right aortic arch, except for the presence of a left-sided retroesophageal aortic diverticulum and a posterior impression on the esophagus [1,8,9].

Anomalies of great vessels are incidental findings as they are usually asymptomatic. However, a right aortic arch with a vascular ring can cause partial or complete obstruction of

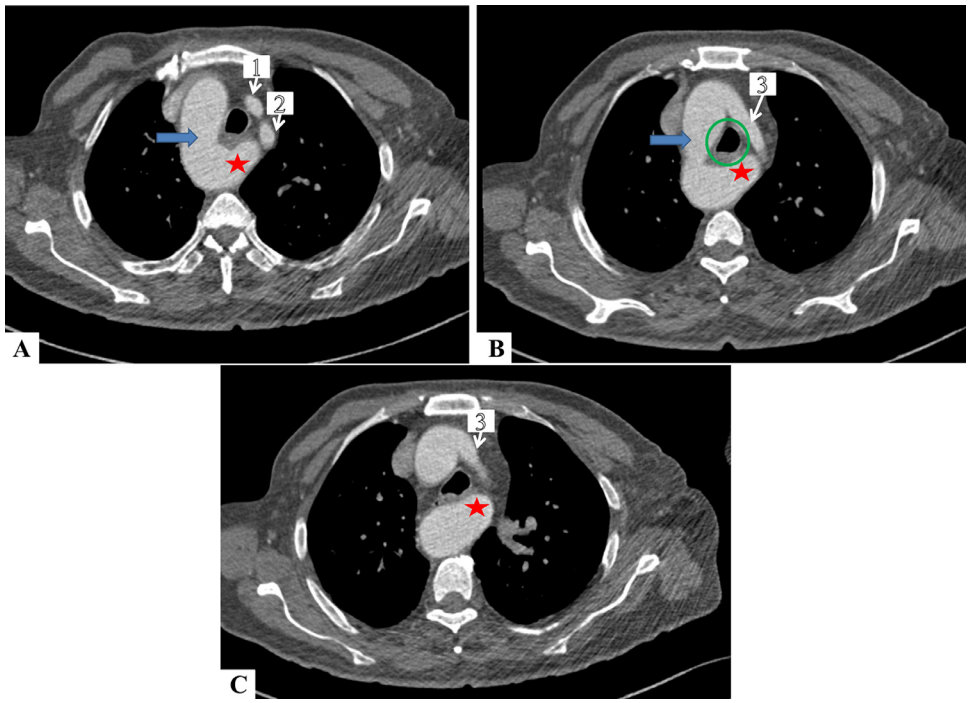

Fig. 3 – Axial contrast-enhanced CT images (A-C) showing a large aortic diverticulum (red asterisk) along with the right-sided aortic arch (blue arrow) forming a vascular ring (green circle) with the left brachiocephalic artery, and causing mild extrinsic compression of the trachea and esophagus. (1): left common carotid artery; (2): left subclavian artery; (3): left brachiocephalic trunk.

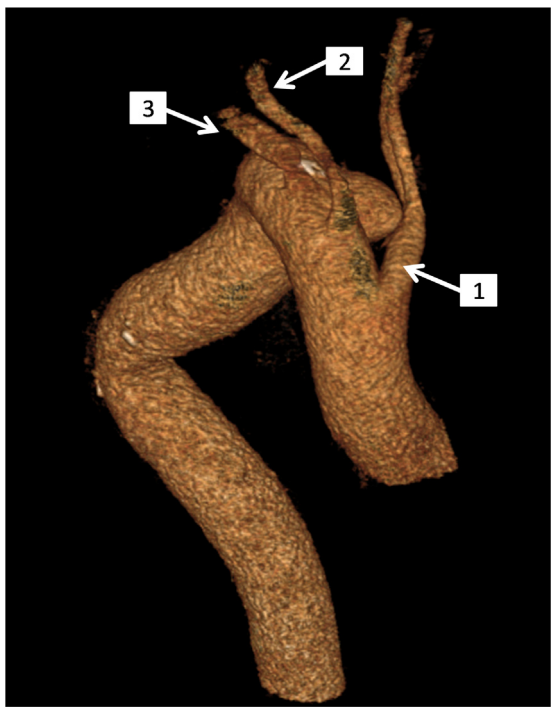

Fig. 4 – 3D Volume rendering image showing a right aortic arch with the left brachiocephalic trunk (1), the right common carotid artery (2), and the right subclavian artery (3).

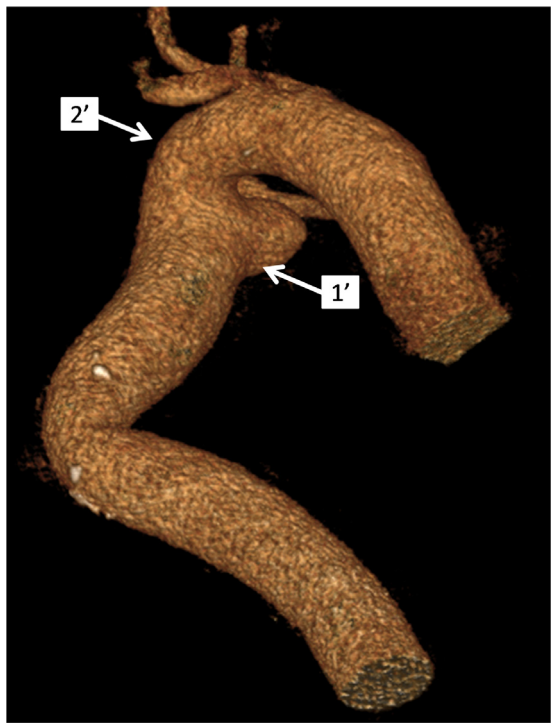

Fig. 5 – 3D Volume rendering image showing a large aortic diverticulum (1′) at the level of the right aortic arch (2′).

the trachea and/or esophagus, leading to symptoms such as wheezing, stridor, dyspnea, dysphagia, cough, chest pain, recurrent respiratory infections, and respiratory distress [2,8]. In adulthood, symptoms are often attributed to atherosclerotic changes in the anomalous vessels, dissection, or aneurysmal dilatation, which may compress surrounding structures [10].

Imaging plays a crucial role in diagnosing aortic arch malformations. The initial approach typically involves a chest X-ray, which can reveal signs of tracheal or esophageal compression. However, contrast-enhanced CT angiography or MRI are the gold standard for detailed imaging, providing a comprehensive view of the aortic arch anatomy and any associated anomalies [11]. These techniques allow for the accurate assessment of the aortic arch structure, the degree of compression on surrounding structures, and the identification of any aneurysms or vascular abnormalities. Echocardiography is also important for evaluating associated cardiovascular abnormalities, particularly when congenital heart defects are suspected [11].

Conclusion

This case demonstrates a rare occurrence of a right-sided aortic arch with mirror-image branching and a vascular ring in an adult patient without congenital heart disease. Despite the mild nature of the compression, the finding highlights the importance of considering vascular anomalies in the differential diagnosis of dyspnea and related symptoms. The case serves as a reminder that even uncommon anatomical variations can have clinical significance, requiring careful imaging and follow-up.

Author contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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