

Double Aortic Arch: The Importance of Computed Tomography Diagnosis

Duplo Arco Aórtico: A Importância do Diagnóstico por Tomografia Computadorizada

Keywords: Aorta, Thoracic/abnormalities; Aorta, Thoracic/diagnostic imaging; Computed Tomography Angiography

Palavras-chave: Angiografia por Tomografia Computadorizada; Aorta Torácica/anomalias congénitas; Aorta Torácica/diagnóstico por imagem

Dear Editor,

The double aortic arch (DAA) is a congenital anomaly arising from abnormal embryogenesis, where the right segment of the fourth aortic arch fails to regress.¹ This results in two aortic arches, one on each side, often causing tracheal and esophageal compression.² Symptoms vary from mild to severe depending on the extent of obstruction. The right arch is typically dominant, but codominance is possible.³

A 77-year-old woman presented to the emergency department with severe dyspnea, thoracic discomfort, and postural instability. Pulmonary thromboembolism was initially suspected, but a computed tomography angiography (CTA) of the chest revealed DAA with codominant arches that slightly compressed the esophagus (Fig. 1); the trachea and bronchi remained patent. Other findings included cardiac enlargement, aortic and coronary atheromatosis, multiple bilateral ground glass opacities (pneumonia), small pleural effusions, and normal additional vascular structures in the mediastinum. She was treated with inhaled fenoterol and oral prednisone, improving her symptoms, and was started on levofloxacin. Follow-up with a pulmonologist and cardiologist was recommended.

Double aortic arch, the most common type of complete vascular ring, is usually diagnosed in childhood, often presenting with symptoms like stridor, vomiting, dysphagia, and breathing difficulties.⁴ Asymptomatic cases may go undetected, being found incidentally in adulthood.¹ Adults may develop symptoms later in life due to reduced vascular compliance from aging, atherosclerosis, and hypertension.¹

Adults with DAA symptoms, such as dyspnea and epigastric pain, face diagnostic delays, often receiving long-term symptomatic treatment instead.⁵ Thoracic aorta CTA is the preferred diagnostic tool, providing high quality and high-resolution images, that can be reconstructed in multiple 2D and 3D planes.¹ In patients with dysphagia, retrosternal pain and pronounced epigastric symptoms, chest CT with positive oral contrast may help to detect esophageal compression, replacing esophagram technique, while tracheal compression can be identified with chest X-ray or CT scan.² Yang et al suggest combining echocardiography and CT for optimal preoperative evaluation.⁴ Magnetic resonance imaging (MRI) is also useful for diagnosing vascular rings, as it is radiation-free and can detect associated congenital heart defects. But, in infants, its use is limited in symptomatic infants due to the need for anesthesia.⁴

Double aortic arch may coexist with other congenital anomalies, including DiGeorge syndrome, tetralogy of Fallot, ventricular septal defects, and truncus arteriosus.¹ Treatment depends on symptom severity. Patients with mild symptoms are managed conservatively,¹ as in the described case. However, severe cases with chronic wheezing, dyspnea, or dysphagia may require surgical intervention, which generally involves dividing the smaller arch.⁴ It remains uncertain whether intrauterine intervention could prevent DAA.⁴

AUTHOR CONTRIBUTIONS

All authors contributed equally to this manuscript and approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

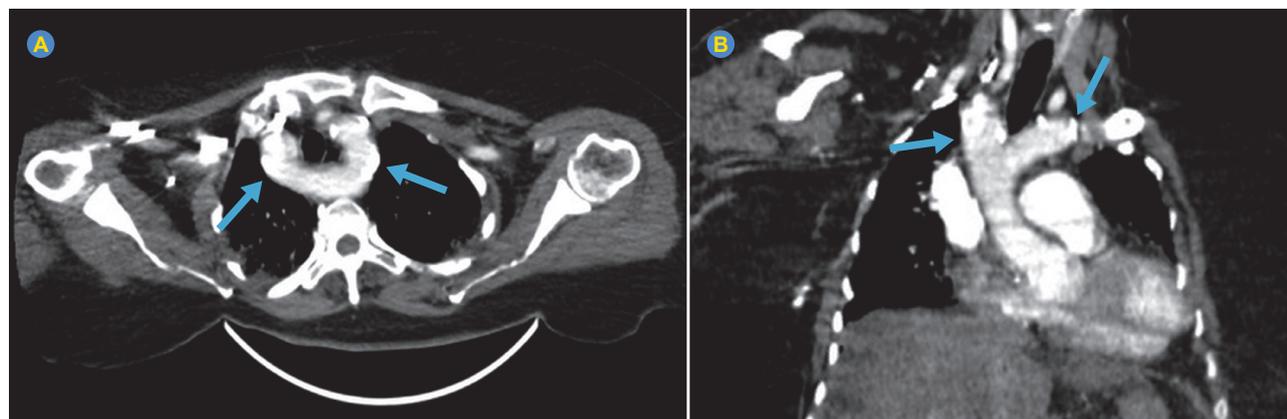


Figure 1 – 64-multislice CTA MIP images (7 mm) in axial (A) and coronal (B) demonstrating the double aortic arch (arrows) with a slight impression on the esophagus

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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