CASE REPORT
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Double aortic arch due to a persistent left arch remnant: a tomographic finding in a patient with tetralogy of Fallot

Doble arco aórtico por remanente de arco izquierdo: hallazgo tomográfico en un paciente con tetralogía de Fallot

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ABSTRACT

The term vascular ring refers to a group of aortic arch anomalies that compress the trachea, esophagus, or both. These malformations represent approximately 1% of all congenital heart defects and are characterized by the formation of a complete ring around the trachea and esophagus, composed of structures derived from the primitive aortic arches. Within this group is the double aortic arch, a rare condition that can be associated with various congenital heart defects. We present the case of a double aortic arch secondary to a persistent remnant of the left aortic arch, identified incidentally on computed tomography in a patient with tetralogy of Fallot.

Keywords: aortic arch anomalies, computed tomography, congenital heart disease, double aortic arch, tetralogy of Fallot, vascular ring.

RESUMEN

El término anillo vascular se refiere a un grupo de anomalías del arco aórtico que producen compresión de la tráquea, el esófago o ambos. Estas malformaciones representan aproximadamente 1% de todas las cardiopatías congénitas y se caracterizan por la formación de un anillo completo alrededor de la tráquea y el esófago, constituido por estructuras derivadas de los arcos aórticos primitivos. Dentro de este grupo se encuentra el doble arco aórtico, una entidad poco frecuente que puede asociarse a diversas cardiopatías congénitas. Presentamos el caso de un doble arco aórtico secundario a un remanente persistente del arco izquierdo, identificado como hallazgo tomográfico en un paciente con tetralogía de Fallot.

Palabras clave: *anomalías del arco aórtico, tomografía computarizada, cardiopatías congénitas, doble arco aórtico, tetralogía de Fallot, anillo vascular.*

Abbreviations:
CT = Computed Tomography
DAA = Double Aortic Arch

Double Aortic Arch (DAA) results from the persistence of both primitive fourth aortic arches. In this condition, the subclavian and common carotid arteries typically arise independently from each arch. The larger arch is termed dominant, with the right arch being predominant in 75-81% of cases.[1]

As a vascular ring, DAA belongs to a group of cardiovascular and ligamentous anomalies originating from abnormal embryonic development, frequently causing

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symptomatic compression of the trachea or esophagus.[2] While DAA often occurs as an isolated lesion, it is associated with other congenital heart diseases in approximately 22% of cases.[3] Among these associations, ventricular septal defect and tetralogy of Fallot are the most prevalent, followed by transposition of the great arteries and truncus arteriosus.[4,5]

This report presents a case of double aortic arch identified as an incidental tomographic finding in a patient with tetralogy of Fallot.

CASE DESCRIPTION

A 7-year-old male presented with a heart murmur and dyspnea on moderate exertion. Echocardiography revealed Tetralogy of Fallot; however, the anatomy of the aortic arch could not be definitively visualized. Consequently, cardiac Computed Tomography (CT) angiography was performed, demonstrating a right aortic arch with mirror-image branching of the supra-aortic trunks and a remnant of the left aortic arch *(Fig. 1)*, consistent with a truncated double aortic arch. The left arch terminated caudally in a blind *cul-de-sac*, originating from the left brachiocephalic trunk *(Fig. 2) (Fig. 3)*.

Given the absence of obstructive symptoms characteristic of a vascular ring and no detectable pressure gradient on echocardiography, the medical team proceeded with the complete surgical repair of the tetralogy of Fallot while opting for conservative monitoring of the double aortic arch. To date, the patient's postoperative progress has been satisfactory.

COMMENTARY

Vascular rings are embryonic developmental anomalies of the aortic arch and great vessels resulting from the persistence of the fourth primitive aortic arches. These structures partially or completely encircle the trachea and esophagus, potentially causing compression and subsequent respiratory or gastrointestinal symptoms.[3,4] The term encompasses both complete rings and partial configurations, with or without fibrous ligamentous bands, that exert mass effect on the aerodigestive tract.[3] While the DAA is the most common complete vascular ring, the case presented here describes an incomplete or truncated variant.

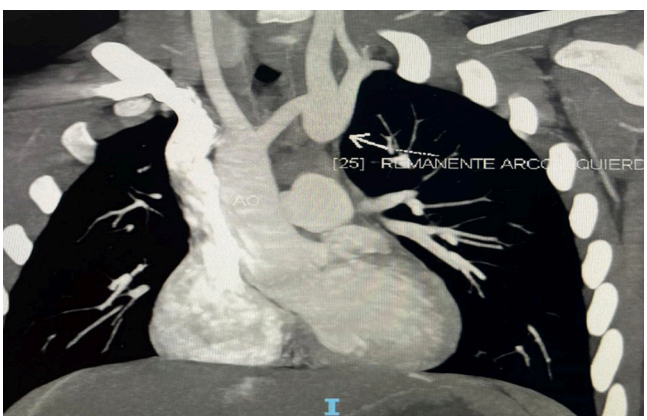

Figure 1: Sagittal section tomography of the heart showing the right aortic arch with a remnant of the left arch.

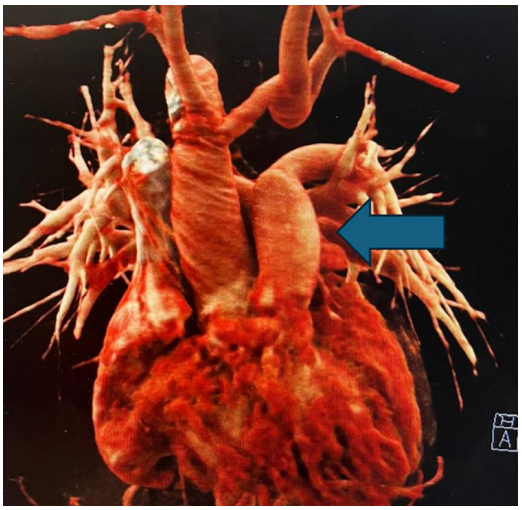

Figure 2: Reconstruction of angiotomography of the heart and large vessels where the arrow point to the remnant of the left arch.

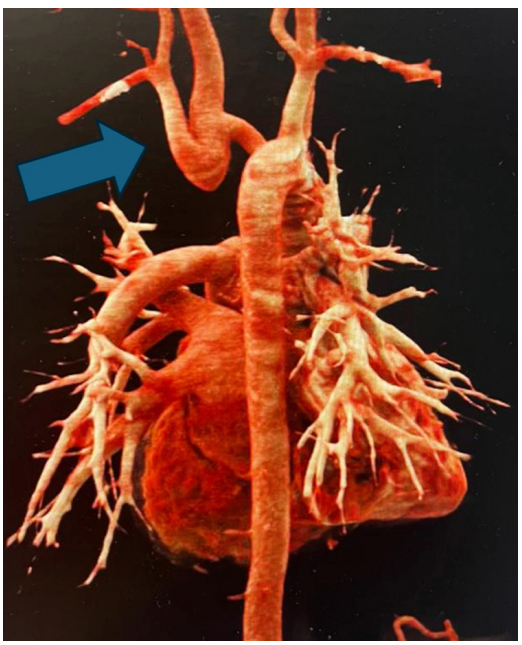

Figure 3: Reconstruction of angiotomography of the heart and large vessels with a posterior view where the remnant of the left arch can be seen ending in a blind *cul-de-sac* caudally.

During cardiac morphogenesis, five pairs of aortic arches connect the ventral and dorsal aortas. The persistence of both fourth aortic arches and both dorsal aortas give rise to a DAA.[3] Typically, these arches are asymmetrical, with right-sided dominance being the most frequent presentation,[5] as observed in our patient. This condition shows a slight male predominance and may occur in isolation; however, approximately 22% of cases are associated with other congenital heart defects, most notably ventricular septal defects and tetralogy of Fallot.[6] This aligns with our case of a male patient diagnosed with Tetralogy of Fallot.

Although symptoms from tracheal or esophageal compression are common, some cases remain asymptomatic and are discovered as incidental findings.[7] Among the various diagnostic modalities, non-invasive techniques such as CT angiography and magnetic resonance imaging are currently preferred. These tools provide detailed three-dimensional anatomical visualization, which is essential for accurate diagnosis and surgical planning.[8]

CONCLUSIONS

Vascular rings represent a rare group of pathologies that warrant exclusion in patients presenting with conotruncal heart defects. Non-invasive diagnostic modalities, particularly computed tomography angiography and magnetic resonance imaging, offer precise anatomical delineation, facilitating accurate diagnosis and management planning.

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