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Case report

ADULT PRESENTATION OF RIGHT-DOMINANT DOUBLE AORTIC ARCH WITH TRACHEOESOPHAGEAL COMPRESSION

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Abstract

Introduction: Double aortic arch (DAA) is a rare congenital vascular ring anomaly resulting from the persistence of both fourth embryonic aortic arches, creating a complete encirclement of the trachea and esophagus. It is most frequently diagnosed in infancy when symptoms of airway or esophageal compression are pronounced. Adult presentation is uncommon and may manifest as progressive dysphagia or respiratory symptoms.

Case Report: We report the case of a 62-year-old male with a 3-month history of progressive dysphagia to solids. Initial esophagography demonstrated smooth, well-circumscribed indentation of the right lateral esophageal wall with leftward displacement, suggestive of a right aortic arch. Subsequent computed tomography angiography (CTA) revealed a double aortic arch with a dominant right arch and hypoplastic left arch, forming a complete vascular ring. There was significant anterolateral tracheal compression with leftward deviation and anterior displacement of the esophagus.

Conclusion: This case underscores that DAA, although typically diagnosed in childhood, can remain asymptomatic until late adulthood. CTA with three-dimensional reconstruction is the diagnostic modality of choice, providing essential anatomical detail for diagnosis and surgical planning in symptomatic adult patients.

Keywords: double aortic arch (DAA), vascular ring, dysphagia lusoria, adult presentation, computed tomography angiography (CTA), 3D reconstruction; tracheoesophageal compression

Introduction

Vascular rings are a heterogeneous group of congenital anomalies of the aortic arch system that result from abnormal regression or persistence of embryonic aortic arches, leading to a complete or partial encirclement of the trachea and esophagus[1,9,10]. This abnormal vascular configuration can cause variable degrees of airway and esophageal compression, producing symptoms ranging from mild feeding difficulties to severe respiratory distress in infancy[9,10].

DAA is the most common type of complete vascular ring, accounting for approximately 50-60% of cases[2]. It results from the persistence of both right and left fourth embryonic aortic arches, which remain patent and form two separate arches that join posteriorly to become the descending aorta. Together, these arches create a complete vascular ring encircling the trachea and esophagus. In most cases, the right arch is dominant, with a larger caliber and a more posterior course, while the left arch is hypoplastic but still functional[4].

Clinically, DAA often manifests during infancy or early childhood due to the small caliber of the airway and esophagus relative to the encircling vascular structures. Typical

symptoms include biphasic stridor, recurrent respiratory infections, cough, and feeding difficulties. Dysphagia in this context is sometimes referred to as dysphagia lusoria, although this term is more classically associated with aberrant subclavian artery anomalies[2,9,10].

Adult presentations of DAA are rare[3]. When symptoms occur later in life, they are usually milder and dominated by progressive dysphagia, sometimes accompanied by exertional dyspnea or wheeze. The delayed onset is thought to be related to age-dependent changes, including decreased compliance of the vascular ring, stiffening of the aortic wall, and reduced elasticity of surrounding tissues. These factors may progressively exacerbate tracheoesophageal compression in a previously asymptomatic individual.

The diagnosis of DAA relies on imaging. Historically, barium esophagography was used to demonstrate characteristic extrinsic impressions on the esophageal wall [8]. However, CTA and magnetic resonance angiography (MRA) have now become the modalities of choice, as they provide high-resolution, multiplanar, and three-dimensional volume-rendered reconstructions, enabling detailed evaluation of the aortic arch anatomy, dominance pattern, vessel branching, and relationship to the trachea and esophagus[4-6]. These imaging capabilities are critical not only for diagnosis but also for preoperative surgical planning.

Case report

A 62-year-old male was referred to Internal Medicine Clinic with a 3-month history of progressive dysphagia to solids. No weight loss, odynophagia, or respiratory distress were reported. Past medical history for congenital heart disease was unremarkable.

Esophagography revealed a smooth, well-circumscribed indentation of the right lateral wall of the upper thoracic esophagus, with leftward displacement, suggestive of a right aortic arch[2] (Figure 1).

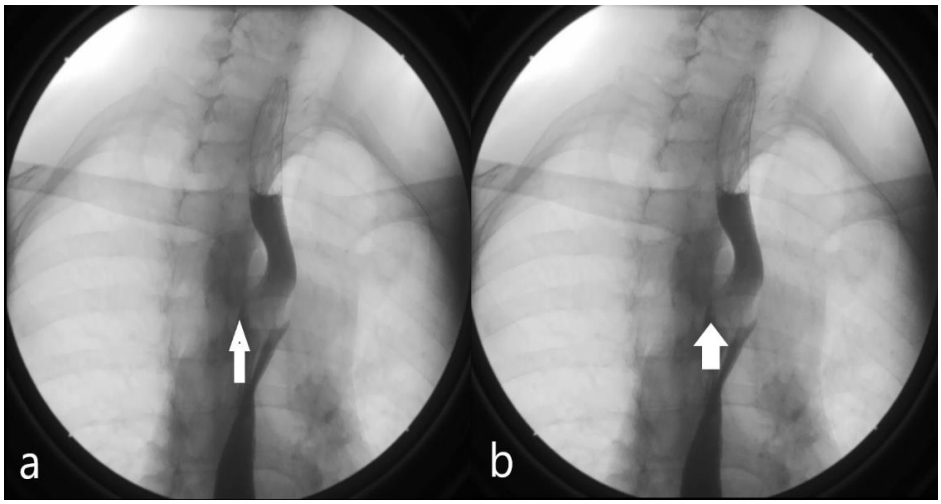

Fig.1. Esophagograms demonstrating smooth right lateral esophageal indentation with leftward displacement suggestive of vascular Ring, white arrows, a) and b).

CT angiography findings:

Double aortic arch anomaly with dominant right arch (diameter up to 22 mm) and hypoplastic left arch (17 mm).

The two arches formed a complete vascular ring encircling the trachea and esophagus.

Marked anterolateral tracheal compression with significant leftward deviation; anterior esophageal compression and displacement.

Ascending aorta diameter: 34 mm; descending aorta: 24 mm.

Supra-aortic vessels arose normally from respective arches.

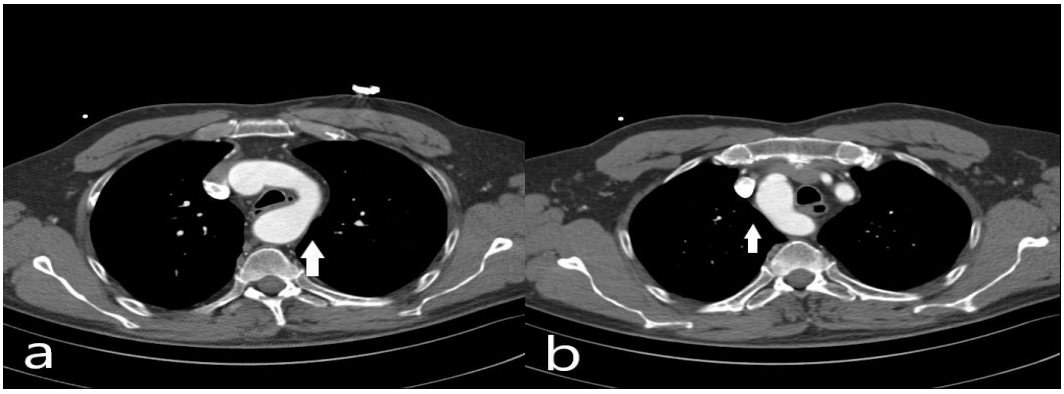

Fig.2. Axial CTA demonstrating double aortic arch with dominant right arch encircling and compressing the trachea and esophagus, white arrows, a) and b)

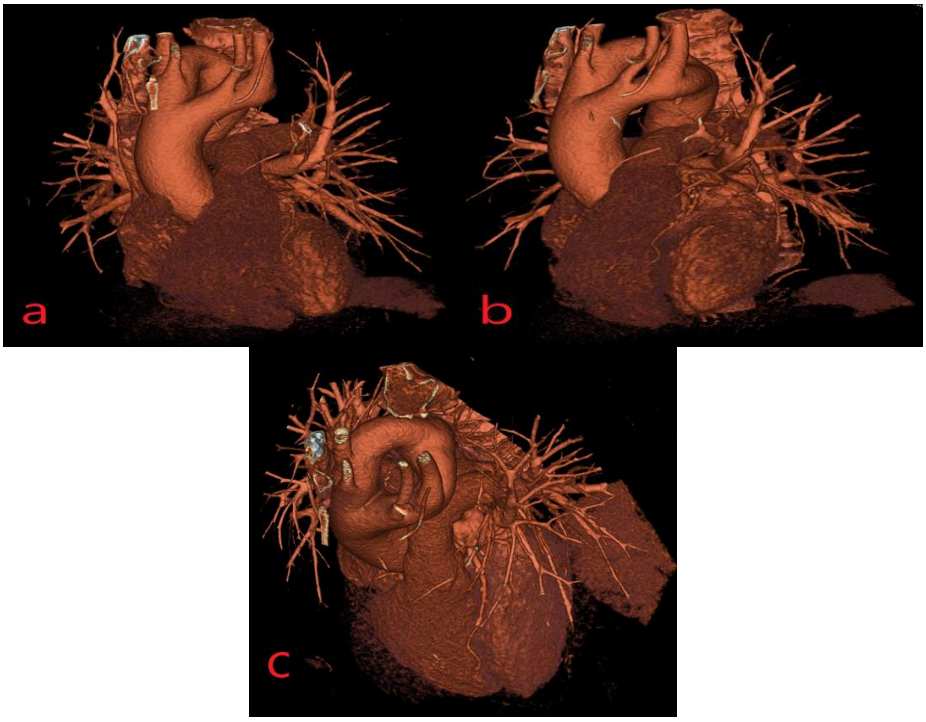

Fig.3. Three-dimensional volume-rendered CTA depicting right-dominant double aortic arch forming a complete vascular ring, a), b) and c).

The findings were consistent with double aortic arch, right dominant type, correlating with the patient's dysphagia (Figures 2 and 3). He was referred to cardiothoracic surgery for further management.

Discussion

DAA represents approximately 1-2% of congenital aortic arch anomalies[2,9], making it the most common form of complete vascular ring. The anomaly results from the persistence of both right and left fourth embryonic aortic arches, which remain patent and encircle the trachea and esophagus. Based on the relative caliber of the arches, DAA is classified into three subtypes: right-dominant (approximately 75% of cases), left-dominant (20%), and balanced (5%)[2,4,9]. In the right-dominant form, as demonstrated in our patient, the right arch is typically larger and passes posterior to the esophagus, while the smaller left arch passes anteriorly[4].

Although DAA is usually diagnosed in infancy or early childhood, presentation in adulthood is rare. Delayed symptomatic onset is attributed to a combination of factors,

including increased stiffness of the aortic walls, progressive calcification, and age-related reduction in esophageal and tracheal compliance, which together can exacerbate previously compensated compression[3]. In some adults, symptoms may be precipitated or worsened by concomitant comorbidities affecting swallowing or breathing. Dekeyzer *et al.* described an elderly patient with an incidentally detected, asymptomatic DAA[3], whereas Yahya *et al*. reported a young adult with a long history of uninvestigated dysphagia[2]. Our case is unusual because of the late onset of clinically significant symptoms and the clear correlation between imaging findings and clinical presentation.

From an imaging standpoint, barium esophagography remains a valuable initial tool, demonstrating characteristic smooth extrinsic impressions on the esophageal wall and displacement patterns that can suggest the side of the dominant arch[2]. However, this modality cannot fully characterize the vascular anatomy. CTA has become the gold standard for diagnosis in both pediatric and adult patients, as it allows rapid acquisition, excellent spatial resolution, and detailed multiplanar and 3D reconstruction[2,4,6]. CTA precisely depicts the course and caliber of each arch, the pattern of supra-aortic branching, and the extent of tracheoesophageal compression. These details are critical for preoperative planning and for avoiding intraoperative surprises. MRA remains an alternative in younger patients or those requiring radiation avoidance, and it has been shown to produce high-quality reconstructions, as demonstrated in the seminal MR-based work by Brockmeier *et al.*[5]. In addition to arterial vascular rings, congenital venous anomalies may also present with delayed or atypical symptoms in adulthood.

Partial anomalous pulmonary venous return (PAPVR) represents such an entity, where abnormal pulmonary venous drainage can remain clinically silent until incidentally detected or associated with comorbid conditions. Our recent institutional experience has highlighted that advanced CTA provides unparalleled detail for the assessment of PAPVR variants, enabling precise anatomical delineation and guiding therapeutic planning. Considered together with our present case of DAA, these reports emphasize that cross-sectional angiographic imaging is indispensable not only for diagnosing rare arterial rings but also for accurately characterizing venous anomalies within the thoracic circulation. This reinforces the concept of CTA as a comprehensive diagnostic modality across the spectrum of congenital vascular anomalies[11].

Management depends on symptom severity and functional impairment. In symptomatic patients, the standard surgical approach is division of the smaller arch, which effectively relieves the vascular ring and decompresses the trachea and esophagus[2,7]. Surgical outcomes are generally excellent, with most patients experiencing symptom resolution or significant improvement[4,7]. In adults, however, surgical decision-making must account for comorbidities, chronic changes in airway or esophageal wall compliance, and the potential risks of thoracic surgery. Conservative management may be appropriate in mildly symptomatic or high-risk patients, but delayed intervention carries the risk of persistent or progressive symptoms.

Our case underscores the importance of considering congenital vascular anomalies in the differential diagnosis of unexplained dysphagia in adults. A high index of suspicion, coupled with an appropriate imaging pathway - starting with esophagography and proceeding to CTA - can ensure timely diagnosis. Furthermore, 3D volume-rendered CTA not only confirms the diagnosis but also facilitates clear communication with the surgical team, helping to plan the optimal intervention strategy.

Conclusion

Double aortic arch with a dominant right arch, although classically diagnosed in infancy, can remain clinically silent for decades and present in adulthood with progressive dysphagia or other compressive symptoms. In such cases, computed tomography angiography with three-dimensional reconstruction is the imaging modality of choice, as it enables precise

delineation of arch anatomy, dominance pattern, and the degree of tracheoesophageal compression, all of which are critical for surgical planning. Awareness of this rare congenital anomaly in the adult population is essential to avoid diagnostic delays, ensure accurate differentiation from other causes of dysphagia, and facilitate timely, effective management.

Conflict of interest statement. None declared.
Written informed consent for publication was obtained from the patient.

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