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Double aortic arch masquerading as bronchial asthma for five decades

Ghulam Nabi Lone¹, Shyam Singh Rathore¹, Javed Ahmad Malik²,
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Abstract

A 60-year-old man treated for chronic bronchial asthma for more than 5 decades, presented in a debilitated condition. Chest radiography showed mediastinal widening, electrocardiography revealed left anterior hemiblock, magnetic resonance imaging and angiocardiology confirmed double aortic arch. He underwent excision of the anterior arch and release of the trachea and esophagus. Double aortic arch should be considered in the differential diagnosis of chronic asthma when a patient fails to respond to medical treatment.

Keywords

Airway obstruction, aorta, thoracic, asthma, tracheomalacia

Introduction

Double aortic arch most commonly manifests in infants and children. Presentation after the 5th decade is an extremely rare entity that mimics chronic asthma.

Case report

A 60-year-old man, non-smoker, presented with paroxysmal atrial tachycardia that required cardioversion with intravenous diltiazem. After stabilization, his baseline electrocardiogram showed left anterior hemiblock. He had been treated for bronchial asthma for more than 55 years without any relief. He had recently developed hoarseness and dysphagia. His chest examination revealed bilateral rhonchi. Lung function tests showed forced vital capacity (FVC) of $1.6 \text{ L} \cdot \text{min}^{-1}$, forced expiration in 1st sec (FEV_1) of $1.4 \text{ L} \cdot \text{min}^{-1}$, and FEV_1/FVC of 87.5%. Arterial blood gas analysis results were: pH 7.27, PaCO_2 37.8 mm Hg, PaO_2 87 mm Hg, HCO_3^- $17.3 \text{ mmol} \cdot \text{L}^{-1}$, base excess $8.7 \text{ mmol} \cdot \text{L}^{-1}$, and oxygen saturation 90.6%. Chest radiography demonstrated a right-sided aortic arch and very prominent bronchovascular markings. Barium swallow showed 2 posterior indentations in the esophagus (Figure 1(a)). Computed tomography-angiography revealed double aortic arch (Figure 1(b)). Double aortic arch was seen on echocardiography, and both components were

demonstrated. Cardiac catheterization and angiocardiology confirmed double aortic arch, however, the flow across the anterior arch was not seen clearly, and the descending aorta arched towards the right side (Figure 2(a)). Exploration through a left posterolateral thoracotomy revealed that the left common carotid and left subclavian arteries arose from the anterior arch which was seen communicating with the posterior arch on the right side and the descending aorta on the left side after the origin of subclavian artery. The left common carotid artery was smaller than the subclavian artery, with a very short distance up to a hugely dilated Kommerell's diverticulum. The trachea and esophagus were severely compressed between the anterior and posterior arches. After proper dissection and vascular control with cardiopulmonary bypass on standby, the anterior arch was transected between the left common carotid and subclavian arteries after applying 2 clamps, and the stumps were closed with

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Figure 1. (a) Barium swallow revealing 2 posterior indentations caused by the retroesophageal component of the double aortic arch. (b) Dynamic computed tomogram of the thorax revealing double aortic arch with tracheal compression.

4/0 polypropylene (Figure 2(b)). The left subclavian artery was to the left of the descending aorta, due to its short origin from the anterior arch near the Kommerell's diverticulum, and the left common carotid artery was shifted to the anterior arch. The small patent ductus arteriosus was transected, and the ends were closed. The lateral stump of the anterior arch was hitched to the chest wall, and the medial stump was retracted and fixed to retrosternal tissue. The esophagus was separated from the posterior arch, trachea, and

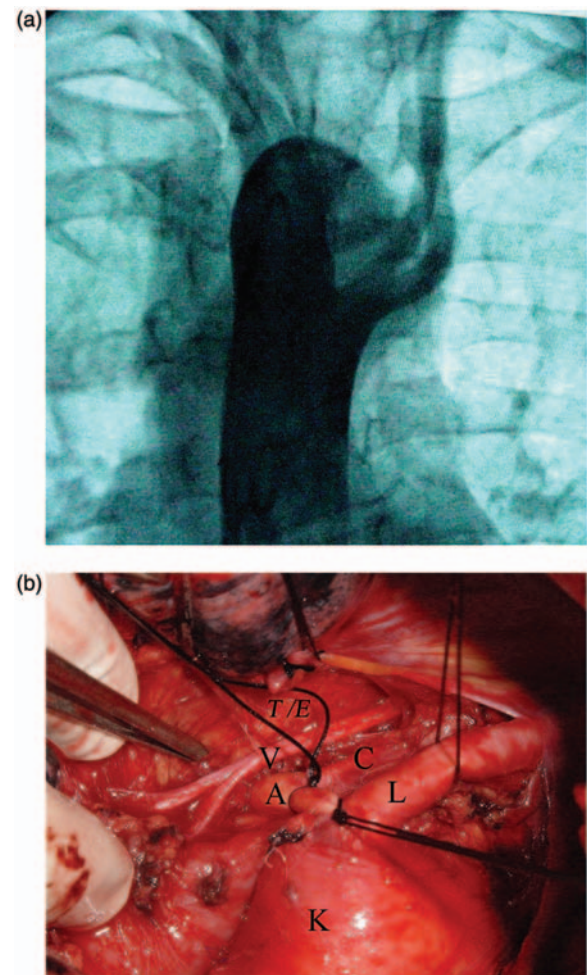


Figure 2. (a) Angiocardiogram depicting the ascending aorta dividing into anterior and posterior arches, giving rise to the arch vessels. (b) Perioperative photograph clearly indicating the anterior arch (A), left subclavian artery (L), Kommerell's diverticulum (K), small left common carotid artery (C), and left vagus nerve (V). However, the esophagus and trachea are not well delineated, being severely compressed by the arch (T/E).

vagus. The thoracotomy wound was closed after inserting an intercostal tube. The patient was electively ventilated for 36 h to avoid any respiratory distress. During the early postoperative period, he developed coagulopathy which resolved after treatment. Dyspnea, dysphagia, and hoarseness improved gradually. The latest pulmonary function results were: vital capacity $2.85 \text{ L}\cdot\text{min}^{-1}$, FVC $2.89 \text{ L}\cdot\text{min}^{-1}$, FEV₁ $2.76 \text{ L}\cdot\text{min}^{-1}$, and FEV₁/FVC 96.0%.

Discussion

Vascular ring is a rare congenital anomaly of the great arteries, which comprises only 1%–2% of congenital heart diseases. Gross¹ coined the term vascular ring after performing repair of double aortic arch.

Most commonly, vascular rings present in infants and children, with either respiratory or gastrointestinal symptoms in approximately 93% and 41% of cases, respectively.² However, dysphagia is the presenting symptom of a vascular ring in adults.³ A right-sided aortic arch in adults is frequently associated with a complete vascular ring that may lead to symptomatic compression of the trachea and esophagus.¹ Delayed presentation is rare. Stoica and colleagues⁴ described a patient with double aortic arch masquerading as bronchial asthma for 30 years. Their patient had an excellent outcome after surgery. Ours is the second-oldest patient reported in the English literature. The exact mechanism whereby such asymptomatic vascular rings present later in life is poorly understood.

A vascular ring results from either abnormal or incomplete regression of one of the 6 embryonic branchial arches. In patients with double aortic arch, the ascending aorta arises normally, but as it leaves the pericardium, it divides into 2 branches, left (anterior) and right (posterior) arches, that join posteriorly and on the left side to form the descending aorta. The right arch, after giving rise to the right common carotid and right subclavian arteries, passes towards the right, posterior to the esophagus, to join the left-sided descending aorta, while the left arch moves anteriorly and in front of the trachea, giving rise to the left common carotid and subclavian arteries. It is joined leftward by the ductus arteriosus or ligamentum arteriosum at the site of the descending aorta, thus completing the vascular ring. Distally, the anterior arch may remain patent or be replaced by a fibrous band that joins the descending aorta at the diverticulum. The morphology of double aortic arch in our case was similar to this type of complete vascular ring. However, the anterior arch was patent throughout its course, and the left subclavian artery originated close to the site of the diverticulum.

Symptoms result from aortic compression of the trachea, compression by aortic dilatation, fluid administration, and exercise, during which the diameter of the ascending aorta increases by approximately 1 mm or more for every 15 mm Hg increase in systolic or diastolic pressure, with consequent circumferential airway narrowing by 3–6 mm.² Other mechanisms include enlargement of the aortic diameter and tortuosity due to hypertension and atherosclerosis, with further worsening of tracheal compression; aneurysmal dilatation of the anomalous vessel and age-related

changes of the thorax limit the dimensions of the mediastinum.² The vascular ring can be approached through a left thoracotomy or median sternotomy when other cardiac anomalies coexist. However, a right-sided approach may also be used. Tracheomalacia, if significant, poses a difficult problem for the surgeon. Tracheomalacia in adults diminishes or disappears altogether after surgical relief of the external compression, although no definitive time frame exists.² Symptoms usually resolve completely after surgery.⁵ If tracheal symptoms persist, then tracheotomy, aortopexy, tracheal resection, or placement of an intraluminal stent are potential therapeutic options.³ Extrinsic compression by a vascular ring, although extremely rare in the elderly, should be considered when a patient presents with respiratory distress or dysphagia refractory to medical treatment. Surgery, even if offered late, bears excellent results.

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Conflicts of interest statement

None declared.

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