‘Vascular ring’, the hidden congenital anomaly having symptoms that may mimic asthma: A case report

Safaa Wafy

Introduction This is a case report of a double aortic arch forming a complete vascular ring, presenting with extrinsic tracheoesophageal obstruction.

History A female patient spent a large part of her life being treated for bronchial asthma. Her complaint started early in childhood marked by repeated attacks of wheezy chest and difficulty in breathing up to stridor sometimes. The wheezes were marked and often associated with stridor. Multislice computed tomography of the chest with angiogram was performed for diagnosis.

Summary Vascular rings, due to the double aortic arch, are an important cause of tracheoesophageal compression. Its presentation may mimic asthma attacks and thus the clinical suspicion and diagnosis of vascular rings can lead to early surgical intervention. *Egypt J Broncho* 2016 10:69–72

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Department of Pulmonology, Assiut University, Assiut, Egypt

Correspondence to Safaa Wafy, MD, FCCP, Department of Chest Diseases, Assiut University, Assiut 71111, Egypt
e-mail: safaa_wafy@hotmail.com

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Introduction

Double aortic arch (DAA) is the most common of the complete vascular ring anomalies, causing tracheoesophageal compression (Fig. 1). This is a case report of a DAA forming complete vascular ring, presenting with extrinsic tracheobronchial obstruction.

History

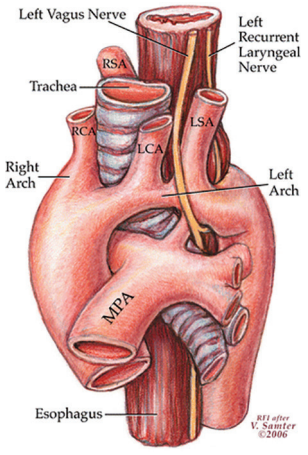
A 53-year-old housewife, having four offspring and with no special habits, spent a large part of her life being treated for bronchial asthma. Her complaint started early in childhood marked by repeated attacks of wheezy chest and difficulty in breathing up to stridor sometimes. She admitted several times to the emergency room, inpatient award, or ICU with the same complaint. She received at all times the regimen for acute severe asthma. All patients gave their formal consent. The protocol was approved by the ethical committee of the Assiut University.

Although she was on asthma medications, more or less regularly, her condition worsened over the course of time. The wheezes were marked and often associated with stridor. The severity of the stridor changed with the patient’s body position; it was worse when she was lying on her back rather than her side. It extended to the neck giving her the sensation of suffocation. Sometimes the stridor could be relieved by extending the neck.

Subsequently, she suffered from difficulties in swallowing, starting with liquids followed by solid food, with choking or regurgitating and increased respiratory obstruction during feeding.

Suddenly, the patient developed massive hemoptysis with severe attack of stridor and wheezy chest. She was admitted to the ICU for evaluation and management. On admission, she was markedly distressed, with inspiratory and expiratory wheezing and stridor. The blood pressure was 200/100 mmHg despite hemoptysis. Cough was marked with expectorations tinged with blood later on.

Fig. 1



A diagram illustrating double aortic arch forming the vascular ring. The trachea and esophagus are passed through and compressed. The right (posterior) arch gives the right subclavian artery and the right common carotid, whereas the left (anterior) arch gives the left subclavian artery and left common carotid.

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Chest examination revealed wheezes with some wet crepitations scattered basally.

Investigations

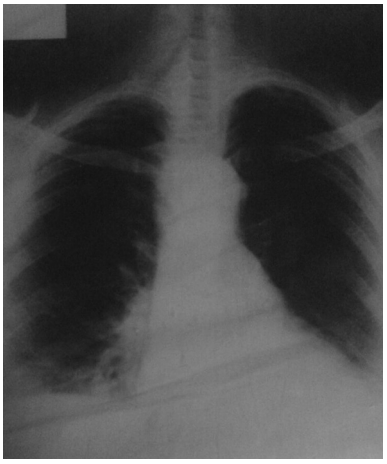
Apart from high blood glucose level and hypochromic microcytic anemia, all other blood investigations were normal.

Chest radiography revealed no abnormalities (Fig. 2). After controlling of hemoptysis, multislice computed tomography of the chest with angiogram was performed. It showed greater bilateral bronchiectasis on the left side with multiple arteriovenous malformations (Fig. 3). The aortic arch divided into two arches – the anterior and posterior – forming a vascular ring. It encircled the trachea and esophagus and compressed both (Fig. 4). The ascending and descending parts were markedly dilated (Fig. 5).

Aortic reconstruction after subtraction of the lungs, bone, and all surrounding soft tissues was performed (Figs. 6 and 7). It showed DAA with right dominant arch. The ascending aorta arises normally from the left ventricle but then divides into two arches, a left and a right aortic arch, which joins posteriorly to become the descending aorta. The smaller left arch passes anteriorly and to the left and then joins with the right arch to form the descending aorta. The left arch gives first origin to the left common carotid artery and then the left subclavian artery. The right aortic arch completes the vascular ring by passing to the right and then behind the esophagus and trachea. The first vessel coming off the right arch is the right common carotid artery, followed by the right subclavian artery.

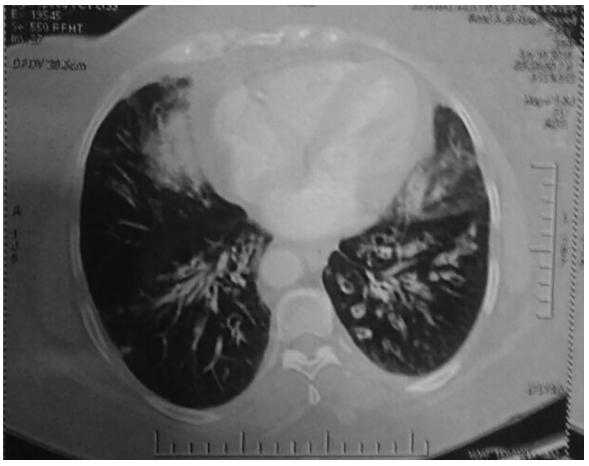
Other investigations were arranged (for academic interest), but postponed until the general condition

Fig. 2



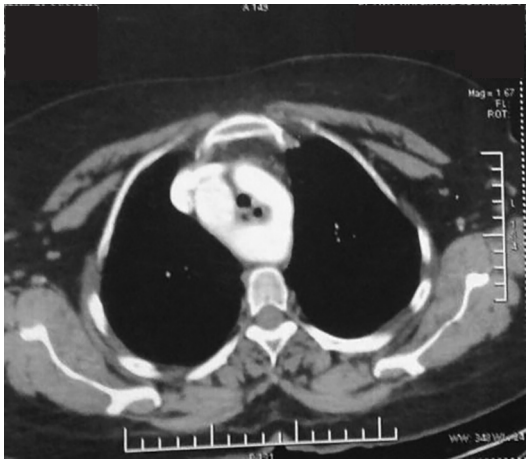
Radiography of the chest shows no abnormality can be detected.

Fig. 3



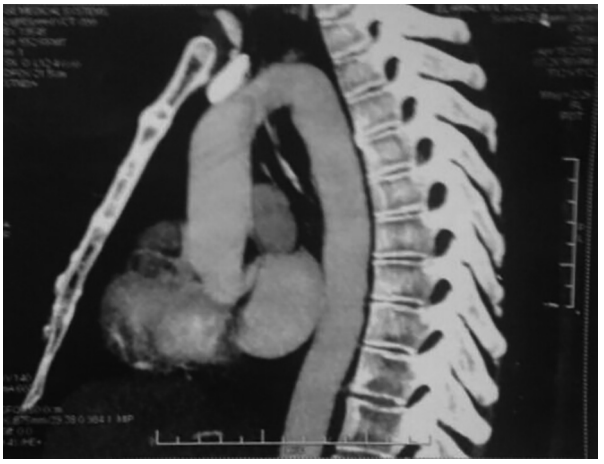
Computed tomography of the chest shows greater bilateral bronchectatic changes at the left side with multiple arteriovenous malformations.

Fig. 4



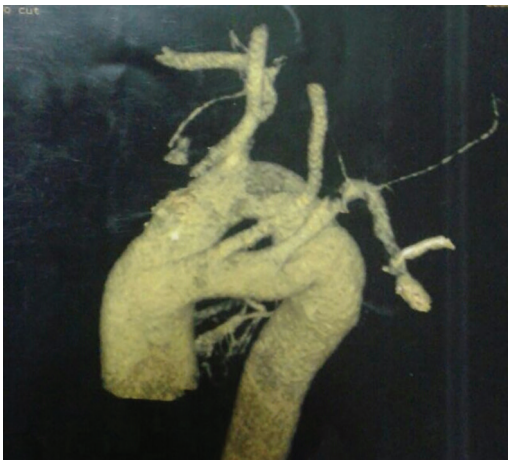
Aortic vascular ring encircles the trachea and esophagus, both are compressed.

Fig. 5



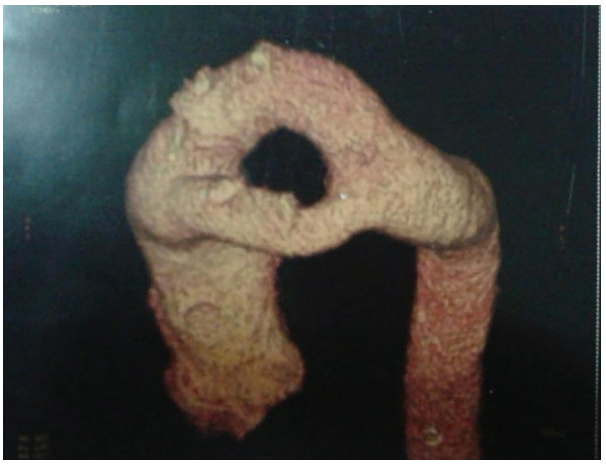
The ascending and descending aorta are markedly dilated.

Fig. 6



The aortic arch is divided into two parts – the anterior and posterior arches – before it reconnects again forming the descending aorta.

Fig. 7



From this aortic vascular ring the trachea and esophagus are passed through (upper view).

of the patient improved. The barium swallow test was carried out to show the esophageal left-sided and right-sided indentations from the vascular compression. Bronchoscopy facilitates the visualization of the sites and severity of pulsatile tracheal compression. Echocardiography is considered to be sensitive and specific in making the diagnosis of DAA. MRI provides excellent images of the trachea and surrounding vascular structures. Cardiac catherization/aortography was performed to evaluate the hemodynamics and anatomy of associated congenital cardiac defects.

Treatment

Surgical correction was indicated in this patient with obstructive symptoms (stridor, wheezing, pulmonary infections, and poor feeding with choking). However, unfortunately, such major, costly operation was not available and thus a conservative approach with regular follow-up was recommended. Controlling the blood pressure is the main line of treatment to avoid recurrence of hemoptysis and to decrease the pressure effect of the aorta on the trachea and esophagus. Moreover, rabid treatment of any chest infection with management of wheezes is very important.

Discussion

A complete vascular ring (DAA) is a relatively rare congenital cardiovascular malformation representing about 0.5–1% of all congenital cardiovascular malformations [1]. The first post-mortem description of DAA was reported in 1737 by Hommell [2]. With the use of barium esophagography it became possible to diagnose aortic arch anomalies during life in the 1930s. The first open surgical correction by means of thoracotomy was performed by Gross [3] at Children's Hospital Boston in 1945.

Little is known as regards the exact causes of aortic arch anomalies. However, the association with chromosome 22q11 deletion (CATCH 22) implies that a genetic component is likely in certain cases [4].

Embryologically, the ventral and dorsal aortas are connected by aortic arches, which persist or involute to give rise to the normal aortic arch, its branches and minor arteries of the head. The right fourth aortic arch normally involutes at about 36–38 days in the 16 mm embryo and the left fourth aortic arch persists to give rise to the normal left aortic arch [5]. A schematic depiction with DAAs and double ductus arteriosus was described by Edwards to explain the various aortic arch anomalies due to the abnormal persistence or regression of various segments in this hypothetical DAA model [6]. The persistence of both the right and left fourth aortic arches leads to a DAA.

Anatomically, the ascending aorta arises normally and, as it exits the pericardium, it divides into two – the right and left aortic arches, which encircle the trachea and the esophagus and reunite posteriorly to form the descending aorta. Hypoplasia of one of the aortic arches is common, with one arch, more commonly the right aortic arch, being dominant. Atresia can be uncommonly present in any of the segments of either of the aortic arches, resulting in various subtypes of DAA with atresia [7].

Patients with DAA can be asymptomatic or present with symptoms ranging from nonspecific complaints to life-threatening respiratory distress. The symptoms of stridorous breathing, dysphagia, a 'barky' chronic cough, susceptibility to bronchopneumonia, head retraction, malnutrition, onset during early infancy,

and an increase in respiratory distress during feeding were described by Wolman [8].

Symptoms are caused by vascular compression of the airway, esophagus, or both. Presentation is often within the first month (neonatal period) and usually within the first 6 months of life. Starting at birth, an inspiratory and expiratory stridor may be present often in combination with an expiratory wheeze. Occasionally, patients with DAAs present late (during later childhood or adulthood). Symptoms may mimic asthma as in the case of this female patient.

Conventional chest radiography may show indentation of tracheal shadow, retrotracheal opacity, and anterior tracheal bowing. Specific radiological signs have been described for barium esophagography [9]. These include bilateral persistent extrinsic compressions of esophagus in anteroposterior view, with the dominant arch causing a deeper and superior indentation and a deep posterior indentation in lateral and oblique views. The barium swallow test is diagnostic in the majority of cases. However, multidetector computed tomography and MRI have become increasingly utilized in the diagnosis and evaluation of aortic arch anomalies, including DAA [10]. Echocardiography is recommended to rule out associated congenital cardiac defects [10,11].

In summary, vascular rings, due to the DAA, are an important cause of tracheoesophageal compression. Its presentation may mimic asthma attacks and thus the clinical suspicion and diagnosis of vascular rings can lead to early surgical intervention if available. This may relieve or avoid the immediate or long-term respiratory complications.

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Conflicts of interest
There are no conflicts of interest.

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