

Right Aortic Arch with A Complete Vascular Ring Causing Tracheoesophageal Compression — A Case Report

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Abstract

Right aortic arch (RAA) with a vascular ring, a congenital malformation of the great vessels of the thorax, is a rare disease in adulthood. Mostly, RAA is accompanied by congenital cardiovascular anomalies diagnosed during childhood, and cause symptoms from the compression of the trachea and/or esophagus.¹ We report a case of a 53-year-old female with a RAA who presented with dyspnea on exertion, dysphagia, and a characteristic images of contrast-enhanced computed tomography (CT) scan, and spirometry. There are a few reports in the literature about symptomatic RAA presented in adulthood, and it may mimic exercise-induced bronchospasm. So diagnosis of this condition may eliminate the need for corticosteroids and the symptoms may resolve completely by surgical intervention only. (J Intern Med Taiwan 2007; 18: 104-107)

Key Words : Right aortic arch, Vascular ring, Dyspnea, Dysphagia, Computed tomography, Spirometry

Introduction

In normal individuals, the left aortic arch passes in front and to the left of the trachea, and the ligamentum arteriosum connects the descending aorta to the pulmonary trunk. The left subclavian artery has a normal course at the left of the trachea. Congenital malformations of the aortic arch arise from disorders in the formation of the primitive double aortic arch². Anomalies of great vessels are frequently incidental findings, because they are usually asymptomatic.

A vascular ring is a rare abnormality of aortic arch that can cause complete or partial obstruction of the trachea and/or esophagus. A complete vascular ring is formed by a right aortic arch, left ligamentum arteriosum that fails to regress, the pulmonary trunk, and aberrant left subclavian artery³.

Case Report

A 53-year-old female, nonsmoker, visited our out-patient-department due to gradually worsening dyspnea on exertion and dysphagia of 1 year's duration. Physical examination was unremarkable. Her chest x-ray revealed a

right aortic knob with trachea deviated to left side (Fig. 1).

The contrast-enhanced CT scan of the chest demonstrated a RAA and the compression of the posterior wall of the lower part of the trachea and the esophagus, the latter from the retroesophageal aortic arch and Kommerell's diverticulum, caused at proximal part of the aberrant left subclavian artery (Fig. 2).

Spirometry showed a FVC of 1.81 L (72% of predicted), FEV1 of 1.16 L (54.7% of predicted), and FEV1/FVC ratio of 64.16%. Her flow-volume loop showed flattening of the inspiratory and expiratory segment (Fig. 3), and evidence of fixed large airway obstruction.

The patient's symptoms were attributed to the complete vascular ring malformation, and a surgical correction of the anomaly was recommended. The patient refused therapeutic intervention, because she can tolerate the symptoms caused by the vascular ring now.

Discussion

Vascular rings usually presents in infancy or childhood with symptoms related to congenital heart anomalies, such as tetralogy of Fallot, pulmonary atresia with ventricular septal defect, and truncus ar-



Fig.1.Chest X-ray revealed a right aortic knob with trachea deviated to left side.



Fig.2.Computed tomography scan of the chest, which shows the compression of the trachea by the right aortic arch with retroesophageal position and and the coma-like appearance of Kommerell's diverticulum as well as the compression of the trachea and esophagus by the vascular ring. The left subclavian artery arose from a diverticulum on the descending aorta.

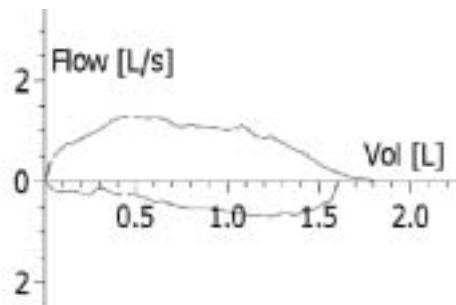


Fig.3.Flow-volume loop demonstrating flattening of the inspiratory and expiratory portion.

teriosus, or to compression of mediastinal structures such as the trachea or the esophagus. The incidence of vascular rings in adult is unknown, a right sided aortic arch occurs in adults with a frequency of 0.1 percent and is associated with a high likely hood of a complete vascular ring, but seldom associated with congenital cardiac abnormalities¹.

The symptoms are more often the result of early atherosclerotic changes of the anomalous vessels, dissection, aneurysmal dilatation with compression of surrounding structures and the vascular rings tend to become more constrictive as the trachea grows. Symptoms may occur transiently with exertion, supine positioning, and fluid administration that dilates the aorta¹.

The anomaly may cause dysphagia, dyspnea, stridor, wheezing, cough, recurrent respiratory track infections, obstructive emphysema or chest pain, and respiratory distress. When these appear in adulthood, they can mimic exercise

induced asthma or other obstructive airway diseases⁴.

In our case, the aortic arch was cited posteriorly and on the right of the trachea. The aortic arch had a retroesophageal course, passing between the esophagus and the thoracic vertebrae, and an aberrant origin of left subclavian artery, called Kommerell's diverticulum, from left-side descending aorta (Fig. 4). So the patient had a complete vascular ring around the trachea and esophagus, which caused dysphagia and dyspnea on exertion. The symptoms may gradually worsening because the atherosclerotic changes of the anomalous vessels is progressing by aging.

The diagnosis of vascular abnormalities of the chest that cause airway obstruction can be made by using barium esophagography, bronchoscopy, CT scan, angiography, and magnetic resonance imaging (MRI)⁵. CT scanning with contrast has replaced barium swallow as the diagnostic procedure of choice⁶. Spirometry findings may be normal or may show flattening of the expiratory and /or inspiratory portion of the flow-volume loop, suggesting a fixed or variable large airway obstruction. The diffusion capacity is typically normal⁴.

In our patient, the presence of RAA on the chest radiograph suggested that his symptoms were likely

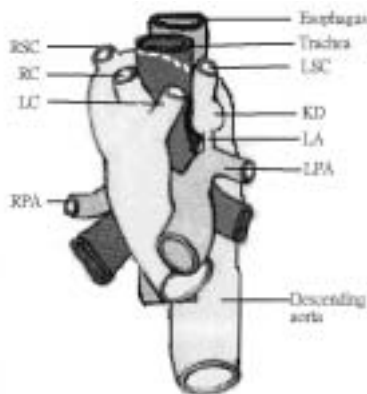


Fig.4. Right ascending aorta, retro-esophageal segment of the aortic arch, left ligamentum arteriosum and pulmonary artery causing a congenital complete vascular ring about the trachea and esophagus. RC: right carotid artery LC: left carotid artery RSC: right subclavian artery LSC: left subclavian artery RPA: right pulmonary artery LPA: left pulmonary artery LA: ligamentum arteriosum KD: Kommerell's diverticulum

to be the result of this anomaly. The vascular ring was confirmed by CT with intravenous contrast enhancement, 3D reconstructed tomography (Fig. 5) and spirometry study. The image of CT and spirometry may replace the bronchoscopy and define the location, degree of the bronchial compression and the respiratory function. The method may prevent the patient to suffer the risk of hypoxia.

Most patients with symptomatic congenital vascular rings will require surgical repair⁸. The right aortic arch with ligamentum arteriosum form of vascular ring causes between 25% and 30% of all cases of vascular rings, and it is the second most common type of vascular ring requiring surgery¹. In adults, the surgical division of the ligamentum arteriosum with dissection of mediastinal structures allows the vessels to assume a less constricting pattern. Division of the retroesophageal artery is often unnecessary, as it was in this case⁸. Patients with mild symptoms may be satisfactorily managed by dietary modifications alone⁹.

After surgery, secondary tracheomalacia is mostly present after the prolonged period of airway compression, and symptoms may not resolve completely. In those who continue to have persistent symptoms despite surgery, tracheal resection or intraluminal stenting may be necessary^{10,5}. A preoperative or intraoperative bronchoscopy is necessary for patients who need operation to rule out associated intratracheal pathology and provide an assessment of bronchomalacia-tracheomalacia¹⁰.

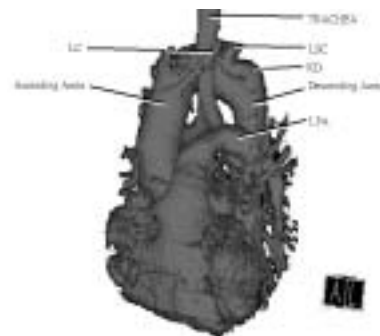


Fig.5. 3D reconstruction computed tomography revealed the relationship between trachea and vascular ring. LC: left carotid artery LSC: left subclavian artery LPA: left pulmonary artery KD: Kommerell's diverticulum

Summary

Congenital vascular ring is rare in adult and a high degree of clinical suspicion is required when all adult patients with dyspnea and/or dysphagia accompanied by an aortic arch anomaly. The image of CT and spirometry can define the disease and evaluate the condition for surgical intervention. The disease may mimic exercise-induced bronchospasm or other obstructive disease, so diagnosis of this condition in patients with chronic respiratory complaints may eliminate the need for corticosteroids and other respiratory medications.

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右主動脈弓合併完整血管環異常導致 氣管食道壓迫——病例報告

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摘 要

右主動脈弓合併血管環異常是一種胸腔內大血管的先天性異常，在成人是非常少見的。因為常常會合併先天性心臟血管異常，而且會因為結構壓迫到氣管與食道而產生症狀，所以絕大部分是在兒童時期就會被發現。我們報告一位53歲女性因右主動脈弓異常合併食道後異位與Kommerell憩室，導致氣管食道壓迫產生運動時呼吸困難與吞嚥困難。病患的胸部電腦斷層與肺功能檢查有典型的變化，可作為診斷的依據。因右主動脈弓異常在成人期才產生症狀的病例報告非常的少，而且和運動性氣喘的臨床症狀相似。因此正確診斷可以避免病人使用類固醇，而且唯有開刀才能完全緩解病人的症狀。