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DOUBLE AORTIC ARCH: A RARE ENTITY PRESENTING IN AN ADULT WITH DYSPNOEA AND DYSPHAGIA.

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ABSTRACT

We present a rare case of a congenital aortic arch anomaly with its clinical and imaging findings with multidetector computed tomography (MDCT) in an adult female who presented with dyspnea and dysphagia. MDCT made an accurate diagnosis of this life-threatening but correctable, anomaly in an adult with repeated respiratory infections, dyspnea and dysphagia.

KEYWORDS: Double aortic arch(DAA), multidetector computed tomography(MDCT), vascular ring, RSCA (right subclavian artery),LSCA (left subclavian artery),RCCA (right common carotid artery),LCCA (left common carotid artery),RAA(right aortic arch),LAA(left aortic arch).

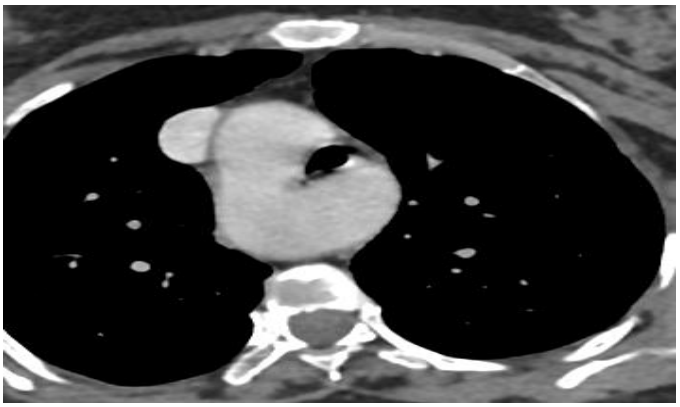
INTRODUCTION

Congenital anomalies though more commonly presenting in pediatric age group can often have a late presentation in adults wherein it can present as a masqueraderer of other diseases such as asthma. Although double aortic arch is a rare congenital anomaly and much rarer to present in adult age group; nevertheless is an important cause of long standing respiratory symptoms which may remain undiagnosed as was the scenario in our case. Multidetector computerized tomography has proved to be an important diagnostic modality which completelydelineates the anatomy of double aortic arch and aids in surgical repair.

CASE REPORT

A 46 year-old female presented with complaints of dyspnea on exertion, dry cough and dysphagia of chronic onset Thesymptoms remained undiagnosed before and the patient was started on bronchodilators.

Echocardiography did not show any evidence of double aortic arch because of suboptimum suprasternal acoustic window. However, it helped to rule out any associated intracardiac anomaly.



MDCT showed partial double aortic archforming an incomplete vascular ring, encircling and compressing the trachea and esophagus [Figure 1a and 1b]. The right aortic arch was the dominant arch with right subclavian, right common carotid artery and left common carotid artery also seen arising from the right arch and left subclavian arteries arising from the left arch.

Lung parenchyma was normal.

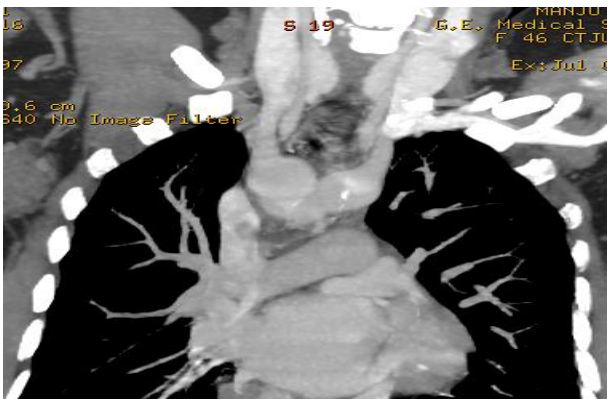


Figure 1: Partial DAA in a 46 year old female presenting with dysphagia and dyspnea. MDCT Axial (A)and Sagittal (B) multiplanar reformatted images show presence of incomplete vascular ring (white arrow) causing esophageal and tracheal compression

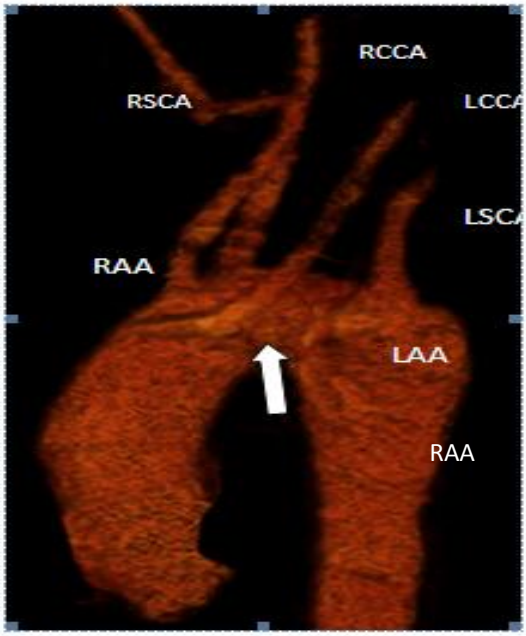


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Figure 2 A and B: Lateral (A) volume rendered images show partial double aortic arch with left common carotid artery (white arrows) seen arising from right aortic arch.

DISCUSSION

Double aortic arch, although rare, is an important cause of persistent respiratory and swallowing difficulties. There is failure of the normal regression of one or more segments of the six pairs of the aortic arches that arise from the truncus arteriosus leads to the formation of multiple anomalies of the aortic arch, like the vascular ring. Therefore, aortic arch anomalies could include the combination of a DAA with an equal right and left component, a smaller or compressive right or left component, and a left- or right-sided descending thoracic aorta. In the present case, the patient had a partial DAA with a right-sided aortic arch, compressive left component and right-sided descending thoracic aorta.

The clinical symptoms of vascular rings result from the compressive effects of the adjacent airway or part of the oesophagus. Respiratory symptoms are common in the case of vascular rings in infancy or early childhood.[6] On the other hand, adult patients complain of difficulties in swallowing rather than respiratory difficulties because of tracheal development. A DAA causing tracheal and esophageal compression is not unusual in children. There have been some references to DAA in adults, but symptomatic and operative cases in adults have been encountered much less frequently. The diagnosis of a DAA using chest radiography or transthoracic echocardiography is often difficult. Besides CT, magnetic resonance imaging is

an important diagnostic tool for identifying anomalies of the aortic arch and its branches and it can be considered the imaging technique of choice when planning surgical management.

Usually, double aortic arch occurs without associated cardiovascular anomalies. However, anomalies like ventricular septal defect, tetralogy of Fallot, truncus arteriosus, transposition of great arteries, pulmonary atresia and complex univentricular defects are seen, with a reported incidence of 17%. Diagnosis of double aortic arch can be made by echocardiography; however, it can be easily missed especially in those where the suprasternal images are not adequate.

Surgical intervention involving division of the minor arch and ligamentum arteriosum is indicated for adult patients who are symptomatic. Thoracotomy should be carried out on the side of the compressive aortic segment and the ligamentum arteriosum because in the presence of a DAA, esophageal compression is relieved by ligation and division of both the compressive aortic arch and ligamentum arteriosum.

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CONFLICTS OF INTEREST

None declared.

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