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Case Report

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CERVICAL AORTIC ARCH ANOMALY WITH BOVINE BRANCHING PATTERN – UNUSUAL CAUSE OF PULSATILE NECK SWELLING AND DYSPHAGIA.

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ABSTRACT

Many diseases of thoracic aorta and aortic arch disorders have been noted in individuals irrespective of gender without any predilection to specific age group. However, aortic arch anomalies are conditions with an atypical combination of respiratory, oesophageal and cervical regional involvement resulting in unusual presentation. It is essential to diagnose anatomical vascular deviations such as the cervical aortic arch and bovine branching pattern to suggest further diagnostic and treatment options whenever required as per the severity of compression of adjacent anatomical structures.

KEYWORDS: Aortic Arch (MeSH unique ID: D001013), Angiography (MeSH unique ID: D000792), Brachiocephalic Trunk (MeSH unique ID: D016122), Dysphagia (MeSH unique ID: D003680).

INTRODUCTION

Aortic arch anomalies can be classified as that of abnormal position or branching pattern.[1] These anomalies can be found in isolated form or in association with each other and cardiac anomalies like atrial septal defect, ventricular septal defect, double outlet right ventricle and tricuspid atresia. Suspicion due to the clinical presentation of pulsatile neck swelling, recurrent respiratory tract infections along with dysphagia is an indicator for Doppler and computed tomographic analysis.[2]

CASE REPORT

A 54 year old female presented with pulsatile neck swelling, pain on the right side of neck and dysphagia for liquids. Ultrasonography of the neck revealed the unconventional tortuous course of common carotid arteries visualized as tubular intermediate thickness walled structures showing pulsatility with wall to wall color flow and arterial waveform. However, no focal lesion was demonstrated.

CT neck angiography was performed which revealed an aortic arch at the level of superior surface of clavicles suggestive of cervical aortic arch (high riding aortic arch) (Figure 1).

There was common origin of the right brachiocephalic artery and the left common carotid artery suggestive of Bovine arch configuration (Figure 2).

Right brachiocephalic and right subclavian arteries showed tortuous course in the neck. Both common carotid arteries showed tortuous course and were seen traversing in close proximity to the oesophagus, abutting the lateral oesophageal wall on both sides (Figure 3).

Rest of the arteries, including left subclavian, both internal and external carotid, vertebral arteries were unremarkable.

a.

b.

Figure 1: Contrast CT neck angiography showing high riding cervical aortic arch.

Figure 2

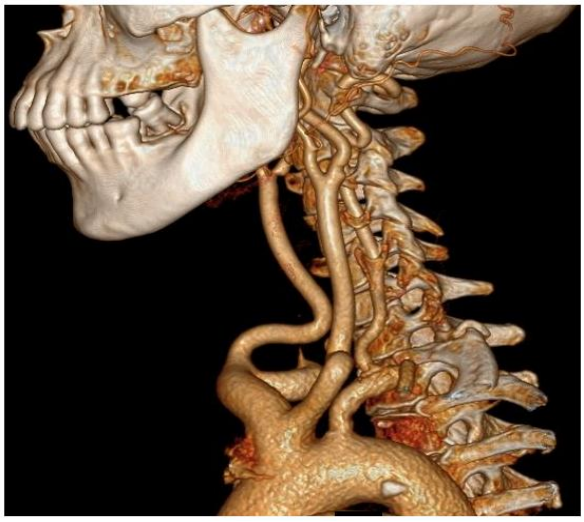

Figure 2: Three dimensional volume rendered image showing common origin of the right brachiocephalic artery and the left common carotid artery.

a.

b.

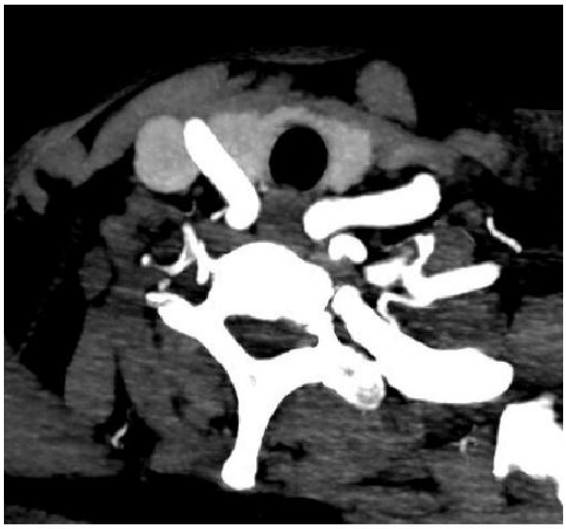

Figure 3: Three dimensional volume rendered image and axial contrast CT neck angiography showing tortuous course of both common carotid arteries which are also seen abutting the lateral oesophageal wall on either side.

DISCUSSION

Aortic arch anomalies include anomalies of abnormal position (of aortic arch) and abnormal pattern of branching aortic arch vessels (pattern of origin of major branches from the arch of aorta). Both types of abnormalities have been found in association with each other. Anatomically, aortic arch anomalies can be divided into 5 types consisting of a right sided aortic arch with normal branching (mirror image), double aortic arch, left aortic arch with abnormal branching, right aortic arch with abnormal branching, interrupted aortic arch and cervical aortic arch.[1] The cervical aortic arch is an infrequent congenital anomaly and shows prevalence of less than 0.01 %. It is characterized by the presence of aortic arch above the clavicle. It may occur as an isolated anomaly or in association with other cardiac aortic anomalies. Aneurysm has been seen in association with left sided cervical aortic arch in 20 % cases.[3]

Embryology: During the phase of embryogenesis, 5 or 6 pairs of aortic arches are present connecting the truncus arteriosus with paired dorsal aortae. Consequently, formation of normal aortic arch occurs by progressive involution of segments of the dorsal aorta, 3rd and 4th arches, with inferior migration of arch from cervical to the thoracic region. The possible etiological process of cervical aortic arch consists of persistent 2nd or 3rd branchial arch with resorption of 4th arch; faulty descent of normal 4th arch into the neck; fusion of 3rd and 4th arches with inability to migrate inferiorly into the neck.[2]

Commonest branching pattern of aortic arch includes 3 great vessels arising separately from the arch (from right to left) namely innominate artery, left common carotid artery, and left subclavian artery (**Figure 4**). A variation in this branching pattern occurs where in there is common origin of the innominate artery and left

common carotid artery. Origin of left common carotid artery is found to be on the right side in these cases. This configuration/ branching pattern is known as bovine aortic arch (**Figure 5**). This type of branching pattern is seen in approximately 13% population.[4]

Figure 4

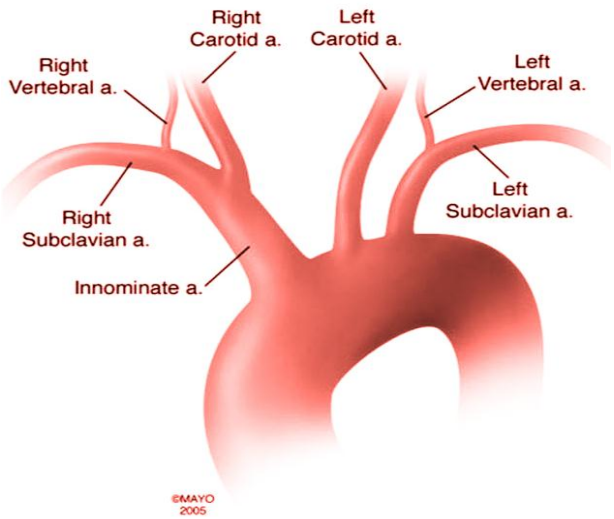

Figure 4: The most common aortic arch branching pattern showing separate origins for the innominate, left common carotid, and left subclavian arteries. Picture courtesy: Layton KF, Kallmes DF, Cloft HJ, Lindell EP, Cox VS. Bovine aortic arch variant in humans: clarification of a common misnomer. AJNR Am J Neuroradiol. 2006 Aug; 27(7):1541-2.PMID: 16908576.

Figure 5

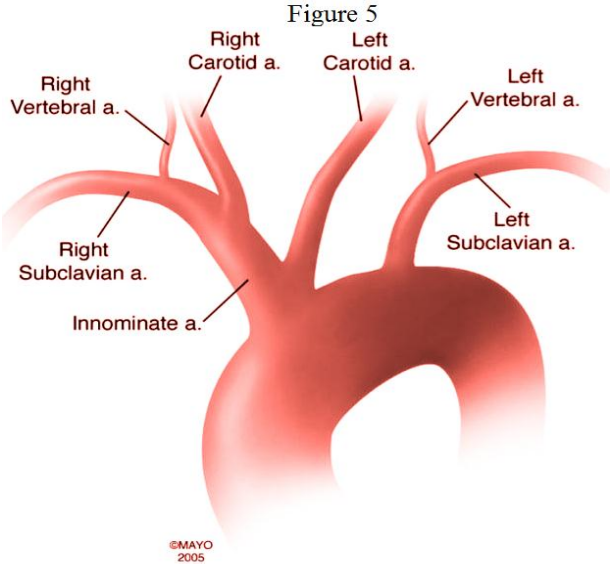

Figure 5: Bovine aortic arch branching pattern showing common origin for the innominate and left common carotid arteries. Picture courtesy: Layton KF, Kallmes DF, Cloft HJ, Lindell EP, Cox VS. Bovine aortic arch variant in humans: clarification of a common misnomer. AJNR Am J Neuroradiol. 2006 Aug; 27(7):1541-2.PMID: 16908576.

Aortic arch anomalies cause a variety of structural abnormalities like oesophageal and tracheobronchial compression due to compression by abnormal pattern of vessels encircling oesophagus and trachea. Other physiological abnormality caused by aortic arch anomalies is abnormal blood flow pattern.[1] These patients present with complaints of pulsatile neck swelling, recurrent respiratory tract infections, respiratory distress, stridor, apnea, brassy cough or dysphagia.[1, 2] Aortic arch anomalies are also found in association with Turner syndrome.[2]

Chest x-ray in these patients may reveal superior mediastinal mass, tracheal deviation towards contralateral side.

Contrast enhanced computed tomography is of great aid in detecting enhancing tubular shaped structure which shows the continuation with ascending and descending aorta. Apex of the enhancing tubular structure is found to be above the medial (sternal) ends of the clavicles. Apart from assessing thoracic vascular anatomy and associated cardiac anomalies, computed tomography gives details regarding the relation of the anomalous aortic arch and its branches with oesophagus and trachea. Degree of tracheo-oesophageal compression can also be assessed.[5]

Computed tomography with or without ECG gated method is precise and fast technique for characterization of aortic pathologies, as well as detection of complications and progression of the disease process.[6]

Multi-detector computed tomography angiography offers several advantages like non-invasive nature, rapid image acquisition, a smaller amount radiation exposure of the patient than transcatheter digital subtraction angiography. It also features advantageous post imaging processing with high spatial resolution and high image quality, thereby making it an observer independent imaging modality. A prominent disadvantage of MDCT angiography is sub-optimal assessment of calcified arteries.[7]

Management depends on the severity of symptoms and trachea-oesophageal compression. Intra-operative digital subtraction angiography provides on the spot useful vascular anatomical details. Open surgical repair consists of thoracotomy and cardiopulmonary bypass. Endovascular repair of the cervical aortic arch appears safe and suitable, despite rare cases of large aneurysm.[8]

CONCLUSION

Aortic arch anomalies can be associated with varied atypical presentation. Precise imaging is essential to reveal the diverse, intricate vascular anatomy in suspected cases. Information regarding specific anatomical relationship of aortic arch and its branching vessels with adjacent aerodigestive tract is of paramount importance as it is critical for surgical planning.

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