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Case Report

The Uncommon Vascular Ring - Not that Uncommon

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Abstract

A cervical aortic arch is a rare vascular finding, which can cause symptomatic compression of the trachea and the esophagus by a vascular ring. We present 3 cases of right cervical arch with varied presentation. Cardiac MRI and CT are consideredthe gold standards for a definitive diagnosis by providing a precise determination of arch and aberrant vessel anatomyand evidence of compressionin order to guide surgical management. We demonstrated the use of cardiac CT scan/ MRI in evaluating all 3 cases of right cervical arch.Based on these patients, important to evaluate the arch to rule out ring compression and coarctation in these patients.

Keywords: *Right aortic arch (AA); Cervical arch (CAA); Coarctation of aorta (COA); Aberrant left subclavian artery (ALSCA); Vascular ring*

Abbreviations: CAA: Cervical arch; CCA: Common carotid artery; COA: Coarctation of aorta; RAA: Right aortic arch; SCA: Subclavian artery; VSD: Ventricular septal defect

Case Report

Case 1

A 9-year-old girl with Di-George syndrome and hyperparathyroidismpresented to our outpatient cardiology clinic with a right sided pulsatile neck mass. The neck mass was noticed for a few months. She had no complaints of dysphagia, vomiting or shortness of breath. Her vital signs were normal for her age and her physical exam revealed a grade 2/6 vibratory systolic murmur in the right upper sternal edge. Her pulsatile neck mass in the right supraclavicular area, associated with a bruit. Peripheral pulses in both upper and lower extremities were equal. She underwent a cardiac MRI and MRA of the neck to further evaluate the pulsatile neck mass, which revealed some interesting findings. She had a high right cervical arch, descending abruptly to the left indicating that it is a circumflex aortic arch, which is most likely the substrate for the vascular ring in her case (Figure 1). The branching pattern wasthat the left carotid artery arose low in the

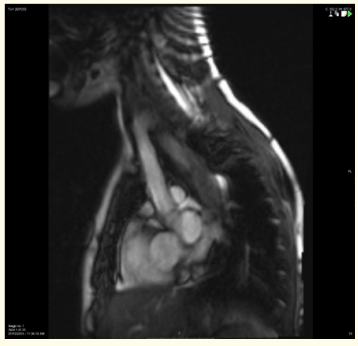

Figure 1: *A 3 chamber view of Cardiac MRI showing a circumflex right aortic arch.*

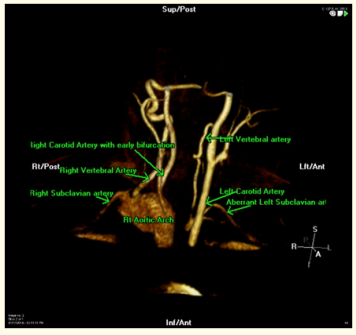

Figure 2: *3D reconstruction showing the abnormal arch vessel branching.*

ascending aorta, a right subclavian artery, separate origin of the right vertebral artery from the aortic arch and the left subclavian artery arising aberrantly from the descending aorta (Figure2). There was no evidence of coarctation of the aorta (COA) or obvious compression of the trachea and esophagus.She is currently being followed clinically as she is asymptomatic.

Case 2

A 3-week old infant with a known history of VSD, diagnosed at birth at an outside hospital, was admitted to our pediatric ICU for RSV bronchiolitis and respiratory distress. She had been experiencing coughing, wheezing and difficulty breathing for 1-2 days prior to the admission. Her initial vital signs showed a heart rate of 137 bpm, respiratory rate of 60/ min, BP of 86/59 and oxygen saturation of 99% on 40% high flow nasal cannula oxygen. Her physical exam revealed subtle dysmorphic features including up- slanting palpebral fissures, a depressed nasal bridge, an anteverted lip and prominent ear lobes. She had mild subcostal retractions, deceased air entry in the lung bases and diffuse wheezing. Her cardiac exam revealed grade 3/6 holosystolicmurmur heard best in the left lower sternal edge, normal upper and lower limb pulses and hepatomegaly measuring 1 cm.

Her initial chest X-Ray showed hyper inflated lungs and diffuse perihilar peribronchial thickening with streaky areas of sub-segmental atelectasis. Because of the heart murmur and a known history of VSD, an echocardiogram was performed, which revealed a large cono-ventricular VSD, moderate pulmonary valve stenosis, right cervical arch (Figure 3) and an aberrant left SCA. A chromosomal microarray analysis was normal as a part of genetic work up.

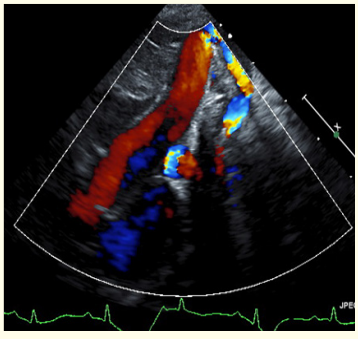

Figure 3: *Suprasternal notch view in echocardiogram showing cervical aortic arch.*

The infant's respiratory distress worsened requiring intubation and ventilatory support for a prolonged duration. A decision was made to obtain a cardiac CT to accurately define the anatomy of the aortic arch and branching and to rule out compression from a possible vascular ring as a right aortic arch with an aberrant left SCA is often a substrate for vascular rings. Cardiac CT revealed some unusual

findings of the right cervical aortic arch which descends to the right and a severe discrete coarctation of the proximal descending thoracic aorta (Figure4). The arch vessel branching pattern was abnormal, with the left CCA as the first branch, followed by theright CCA, which was diminutive throughout its entire length and right the SCA was the third branch (Figure 4). There was no evidence of the left SCA origin from the arch, and was occluded at its origin and the proximal portion. There was a collateral vessel arising at the mid descending thoracic aorta, turning posteriorly in the mediastinum and coursing superiorly to connect to the mid portion of the left SCA. There was no evidence of compression of the trachea or the esophagus.The infant under went elective surgical repair.

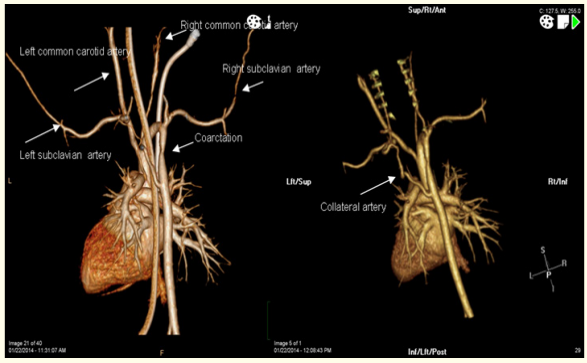

Figure 4: *Cardiac CT showing right cervical aortic arch, which descends to the right and a severe discrete coarctation of the proximal descending aorta and abnormal arch vessel branching.*

Case 3

A 46-year-old Hispanic female with a past history of bronchiectasis, obstructive sleep apnea, recurrent pneumonia and sinusitis, presented with worsening dyspnea on exertion, hoarseness and palpitations. She did not have any complaints of syncope, dysphagia or chest pain. She underwent an extensive work-up for immunodeficiency, cystic fibrosis and genetic testing for 22q11 deletion, which were all negative. Her physical exam revealed grade 1/6 ejection systolic murmur in the left lower sternal edge, rhonchi and wheezing in bilateral lung fields. The electrocardiogram showed sinus bradycardia. Her echocardiogram demonstrated good biventricular systolic function, a dilated left atrium and mild left lower pulmonary vein stenosis. She underwent cardiac MRI and MRA of the neck, whichrevealed a right circumflex aortic arch with mild to moderate compression of the trachea and right main stem bronchus(Figure 5) and mild left lower pulmonary vein stenosis.

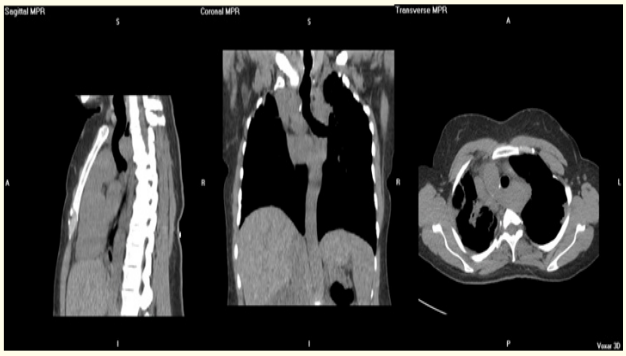

Figure 5: *Cardiac MRI demonstrating right circumflex aortic arch with compression of the trachea and right main stem bronchus.*

It's important to note from that patients with cervical arch present at different age group, present with various symptomatology. A high index of suspicion is required when encountered with symptoms described in the case series and eventually an appropriate modality of investigation like a cardiac MRI should be performed to confirm and carefully evaluate the arch, its branches, course and rule out any obstruction in the course.

Discussion

Cervical aortic arch is a rare anatomical finding, which is defined as the arch coursing above the thoracic inlet. Reid [1] in 1914 was the first to describe this condition. The cervical arch derives from the third branchial arch [2,3] with regression of the fourth branchial arch. In comparison, the origin of normal aorta is from the fourth branchial arch. An alternative explanation for a cervical arch is the failure of descent of the normal arch in the embryo.

A right aortic arch has been found in approximately 0.1% of cervical aortic arches [4-7].

There are five cervical arches [8] during the development that are categorized according to the configuration of the aorta, the brachiocephalic branching sequence and embryogenesis. Type A has separate external and internal carotid artery branches from the aortic arch. Type B has dual common carotid arteries. Type C is a left cervical arch with a right-sided descending aorta and carotid trunk. Type D is described as a left cervical arch with a normal branching pattern, there is a redundant transverse aorta and left-sided (often hypoplastic) descending aorta. Type E has a right cervical aortic arch with a right descending aorta and an aberrant left subclavian artery.

When the cervical arch is identified early in life, the presentation is often related to stridor and dysphagia. The circumflex aortic arch it self may lead to compression. It is important to note that the right aortic arch itself is not a clinical concern. We know that the persistence of the right aortic arch may be favored by flow-related factors in the developing embryo. The reason for persistence of left sided arch is due to the flow characteristicsin the out flow region of the developing heart favoring the laminar flow of right ventricular output through the left sided ductus arteriosus and to the left aortic arch, which in turn encourages the predominance of the left arch [9]. The CAA may lead to symptoms in one of these ways. A CAA can form a complete vascular ring with the right cervical arch, an aberrant left subclavian artery and a left ligamentum arteriosum or with a circumflex arch. Also during evaluation, it is important to perform genetic studies as CAA is linked to 22q11 microdeletion [10] although in these cases x did not have the micro deletion.

While some patients are asymptomatic early in life, they may present later on in life with a pulsatile neck mass as in Case 1. The cervical arch has been mistaken for an aneurysm. A murmur is heard or palpable thrill felt in the same location of tracheo-esophageal compression (stridor, dyspnea, recurrent bronchitis, and dysphagia). Now with cMRI/ CT the anatomical diagnosis can be defined accurately as in our case series. The presence of coarctation should be carefully evaluated [4,11]. As the normal position of the cervicalarch is higher than usual, the coarctation in the descending position may be easily missed if not carefully evaluated in relation to the arch.

A cervical aortic arch, although a rare finding, can present with various clinical findings and at different ages. cMRI/CT scanning have been used successfully as demonstrated in our case series to define the anatomy of the right cervical arch and also explain the potential mechanism causing the symptoms. Based on the findings, the decision can be made to either observe or recommend surgical correction.

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