

Coexistence Of Interrupted Aortic Arch With Aortopulmonary Window Detected On MDCT In A 9 Year Old Male Child

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Citation

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Abstract

Interrupted aortic arch is a congenital malformation of the aortic arch that occurs in 3 per million live births¹. This anomaly is defined as a loss of luminal continuity between the ascending and descending portions of the aorta. Aortopulmonary window is another rare cardiac anomaly that results from incomplete division of the aortopulmonary septum between the ascending aorta and the main pulmonary artery. The incidence of this disorder is estimated at 0.1 to 0.2% of all patients with congenital heart disease².

With advent of Multidetector Computed tomography (MDCT), utility of CT in evaluation of cardiac pathology has gained importance with supplementary role in the evaluation of patients with congenital heart disease³.

We report a case of 9 years old child with cyanosis, radio-femoral delay and claudication. Contrast CT chest was performed on MDCT that showed co-existence of Type A Interrupted aortic arch with large Aortopulmonary window.

CASE REPORT

A 9 year old male child presented with generalized cyanosis, radio-femoral delay and lower limb claudication. On 2D Echocardiography the patient was diagnosed as coarctation of aorta with changes of pulmonary hypertension and pulmonary regurgitation. The child was referred to the CT scan to confirm the same and to look for any ancillary finding. Plain and contrast CT scan of chest was performed on Multidetector CT scanner [Siemens Somatom MDCT] by injecting 100 cc of nonionic contrast at the rate of 2.5 ml/sec with 1.25mm collimation, which were used to generate 1.25 mm axial CT scan slice. These axial slices were utilized for reconstructions in various planes.

The CT angiography revealed normal aortic root, normal origins of both the coronary arteries, with 2.5 cms of normal ascending aorta. A 3 cms sized large aorta pulmonary window was seen between the ascending aorta and main pulmonary artery, beyond which normal 1 cms of ascending aorta and proximal arch of aorta was seen. The three arch branches namely innominate artery, left common carotid and subclavian arteries originated normally from the arch, but in close proximity to each other. Beyond the origin of left subclavian artery the arch of aorta was interrupted. The

descending aorta was seen in higher CT scan sections arising or rather forming from the distal portion of main pulmonary artery (MPA). Thus a diagnosis of a large AP window associated with interruption of arch of aorta was made.

Figure 1

Figure 1: Chest CECT image reveals a large AP window (black arrow) with RPA origin from the dilated MPA.

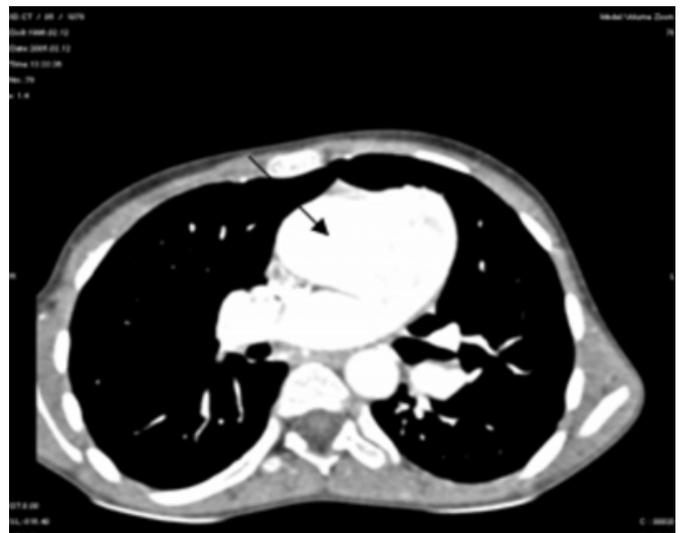


Figure 2

Figure 2: Chest CECT image reveals large AP window with origin of LPA.



Figure 4

Figure 4: Volume rendered 3D image Type A Interrupted aortic arch (white arrow) with AP window (black arrow).

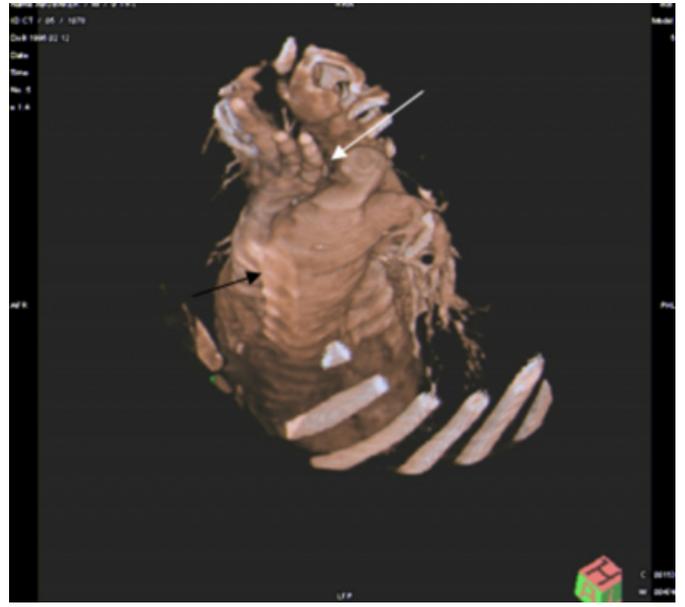


Figure 3

Figure 3: Sagittal reconstructed image shows Type A Interrupted aortic arch (white arrow) with AP window (black arrow).

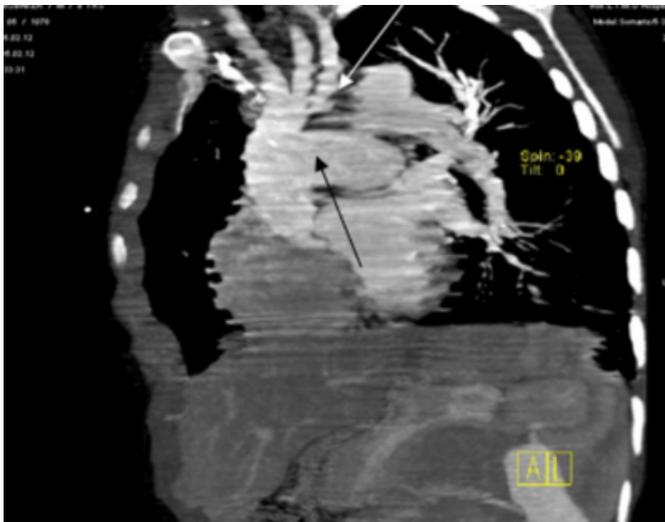
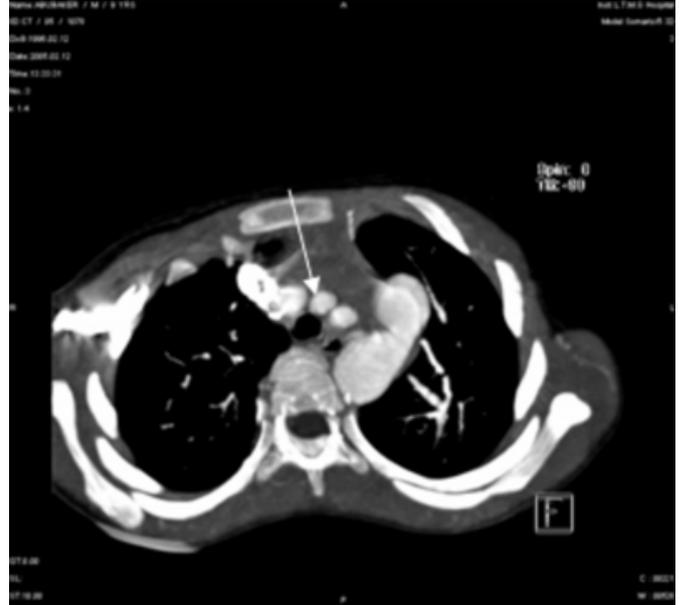


Figure 5

Figure 5: MIP images showing three arch vessels (white arrow); MPA and proximal descending aorta forming an arch like structure (arrowhead).



DISCUSSION

Interrupted aortic arch and AP window individually are rare congenital cardiac anomalies and we present an even rarer condition in which both these pathologies showed co-existence.

An aortic arch is interrupted when the artery does not allow a continuity of flow from ascending to descending aorta. Loss of continuity is usually complete; however, there may be a segment of tissue interposed between proximal and distal aspects. The ductus arteriosus provides the only direct communication between the heart and the lower half of the body. Aortic interruption occurs in 3 per million live births and represents 1.3% of children with known heart disease resulting in death, cardiac catheterization, or surgery in the first year of life.

Classification

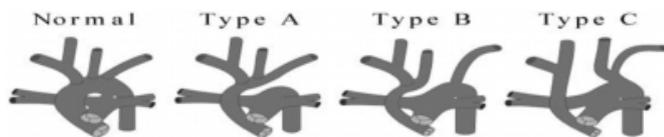
The classification of arch interruptions⁴ is based on the position of the interruption relative to the brachiocephalic vessels. It consists of three major types,

A: distal to the left subclavian artery.

B: distal to the left common carotid artery.

C: distal to the brachiocephalic artery.

Figure 6



The associated anomalies seen with Interrupted aortic arch are DiGeorge syndrome (in Type B: 10-50%)⁵, associated abnormalities of the aortic arch (aberrant or isolated left or right subclavian arteries), intracardiac anomalies (96%)^{6,7} including VSD, patent ductus arteriosus, left ventricular outflow obstruction (subaortic stenosis or bicuspid aortic valve), truncus arteriosus, CHARGE syndrome (CHARGE syndrome is a disruption of the neural crest migration with consequent effect on tissues arising from the third and fourth branchial arches. It includes coloboma, heart disease, choanal atresia, retardation of postnatal growth, genital hypoplasia and ear abnormalities). The embryological fault is thought to be multifactorial but may be associated with aneuploidies and teratogenic exposure (Bis-dichloroacetyl diamene, isotretinoin).

Aortopulmonary window is a rare cardiac anomaly that results from incomplete division of the aortopulmonary septum between the ascending aorta and the main pulmonary artery. The incidence of this disorder is estimated at 0.1 to 0.2% of all patients with congenital heart disease. An aortopulmonary window allows the 'red' oxygenated blood

to pass from the aorta back into the pulmonary artery and mix with the unoxygenated or 'blue' blood.

There are three types of APW defects⁸:

Type I: a proximal defect in the wall between the aorta and the pulmonary artery usually located right above the sinus of valsalva.

Type II: a distal defect in the wall between the aorta and the pulmonary artery.

Type III: a total defect that usually extends the length of the wall between the aorta and the pulmonary artery

When an APW occurs, the aortic and the pulmonary valves are usually normally formed.

The clinical presentation in AP window is related to its haemodynamics of a significant left to right shunt. In early life, patients present with signs and symptoms of congestive heart failure, failure to thrive and propensity for frequent chest infections². The extent and severity depend on the amount of left to right flow, which is in turn is dependent on the size of the defect. Elevation of pulmonary pressure results in development of pulmonary hypertension. Physical findings may include a widened pulse pressure, praecordial heave, and a systolic or continuous heart murmur.

Whereas the Interrupted aortic arch infants do well right after birth but develop symptoms of congestive heart failure such as rapid breathing, clammy sweating, and poor feeding during the first week of life⁹. If the condition is left untreated, 90% of the affected infants succumb at a median age of 4 days¹⁰. Most babies with interrupted aortic arch are born at term and are of normal weight and length. On physical examination a heart murmur is usually heard. The heart rate and breathing rate may be increased and the liver may be enlarged. The left arm and/or leg pulses may be absent. In the few documented cases in adults, the presentation ranges from a lack of symptoms to limb swelling with differential blood pressures in all extremities. Substantial collateral circulation must be present to maintain flow and enable survival. However, collateral vessels are subject to atrophy and atherosclerosis, which can lead to other challenging problems.

INVESTIGATIVE PROCEDURES

Echocardiography- this is an excellent non-invasive means for the diagnosis. A combination of multiple,

echocardiographic views, including praecordial, subcostal and suprasternal cuts, can help to confirm the diagnosis. The use of colour flow and Doppler interrogation techniques are further adjuncts to the real-time imaging.

In this patient the 2D echo findings were possibly interpreted due to the MPA and descending aorta forming a narrow arch, thus simulating a coarctation.

Cardiac catheterization-The role of cardiac catheterization is now decreasing with the advent of sophisticated non-invasive imaging modalities mentioned above. However, it remains a definitive investigative tool in confirming the diagnosis, and in demonstrating or excluding the presence of important associated cardiac anomalies. It is also carried out to establish reversibility of the pulmonary vasculature before surgical repair, usually in the older patient. Measurement of the pulmonary and systemic pressures, and calculating shunts, vascular resistances are important data obtained from the catheter study

Prenatal diagnosis- Interrupted aortic arch can be diagnosed before birth by a fetal echocardiogram or heart ultrasound as early as 18 weeks into the pregnancy 9. This test is done when there is a family history of congenital heart disease or when a question is raised during a routine prenatal ultrasound.

Computed tomography (CT) and magnetic resonance (MR) imaging- have important roles in evaluation of congenital cardiac malformations. MDCT has advantages over echocardiography and MR imaging in evaluating cardiac pathologies. These advantages include short scanning time, high temporal and spatial resolution, and compatibility with ECG gating. MDCT can clearly delineate the cardiac chambers and the vessels in the mediastinum and chest wall. The technique is particularly useful in young children without a need for deep sedation. Thus MDCT has revolutionized cardiac evaluation. Its role in imaging of coronary arteries and in calcium scoring is well established. It also plays a highly promising role in diagnosing, if not all but in many cardiac congenital anomalies.

The rarity in our case lies in the co-existence of two rare congenital cardiac anomalies i.e. Type A Interrupted aortic arch and Aortopulmonary window with a delayed age of presentation i.e. 9 years.

CONCLUSION

CT is a useful imaging modality for the morphologic evaluation of CHD. Reformatted images from MDCT can accurately and systematically delineate the normal and pathologic morphologic features of the cardiovascular structures.

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