Case 16870

Eurorad

Interrupted Aortic Arch Type A Subtype I: A Rare Experience

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Section: Paediatric radiology
Area of Interest: Arteries / Aorta Cardiovascular system
Paediatric
Procedure: Computer Applications-3D
Imaging Technique: CT-Angiography
Special Focus: Education and training Case Type:
Clinical Cases
Authors: Pradeep Raj Regmi1, Isha Amatya2
Patient: 8 years, male

Clinical History:

An 8-year-old child was brought with history of recurrent episodes of dyspnoea predominantly on exertion and an on-and-off fever for last 6 months. The child had no cyanosis. Chest X-ray appeared normal. Transthoracic echocardiography revealed suspicious congenital anomaly of aorta and small ventricular septal defect. Then, CT aortogram was planned for detail evaluation.

Imaging Findings:

Abrupt luminal discontinuity was seen distal to the origin of left subcalvian artery (figure 1a and 1b). PDA is seen originating from the pulmonary artery and measures 6.9mm in diameter in its narrowest part and seen as hourglass appearance in axial images (figure 2a). Descending Thoracic Aorta (DTA) was seen continuous with patent ductus arteriosus (PDA) (figure 2b and 2c). Dilated collaterals were noted reinforcing the descending thoracic aorta via posterior intercostal arteries (figure 1c). Patchy consolidation was seen in anterior segment of left upper lobe and superior segment of left lower lobe (figure 1b, 1c and 2a).

Discussion:

Interrupted Aortic Arch (IAA) is an uncommon congenital vascular anomaly *(2-3 per million live births)* where there is a separation between the ascending and descending aorta. It can either be complete or connected by a fibrous band. Exact pathogenesis of IAA is not known. However, significantly decreased flow during the embryological process within the aortic arch might play an important role. According to Celoria-Patton classification, IAA classified as [1-3];

1.Type A: Second most common distal to the subcalvian arterial origin

2.Type B: Most common (>50%), between the left CCA origin and subclavian arterial origin.

3.Type C: Rare.Interruption occurs proximal to left CCA origin

Each type divides into three subtypes;

Subtype1: Normal subcalvian artery

Subtype2: Aberrant subclavian artery

Subtype3: Isolated subcalvian artery from ductus arteriosus

Type A is the second most common anomaly and is thought to result from abnormal regression of the left fourth arch segment after the left subclavian artery has ascended to its normal position. Interrupted aortic arch is associated with intracardiac anomalies like ventricular septal defect (VSD) and patent ductus arteriosus (PDA). The most common anomaly is patent ductus arteriosus (PDA) which accounts for about 97% of the cases. About 50% of IAA is associated with a chromosome 22q11.2 deletion (associated with DiGeorge syndrome) which is more particularly with type B [2].

Prior to the surgical planning, imaging plays a crucial role to know the exact types of IAA and associated cardiac anomalies. Echocardiography plays the first and important role in screening diagnosis of aortic arch anomalies which is free of radiation, particularly advantageous for paediatric population. However, it has its limitations as it depends on the observer capabilities and to define the exact site of interruption might be challenging. Therefore, CT or MR angiography plays an important diagnostic role. Diagnostic assessment by CT or MRI can demonstrate the morphological features and potential complex associated findings because of its multiplanar capabilities. MR angiography is especially advantageous in children due to abscence of radiation but one major disadvantage is its long time for acquisition compared to CT angiography [4].

IAA is related to mortality rate of more than 90% if left untreated after 1 year of age. However, it can present late as in our case due to some unique collaterals which maintain the flow to the descending aorta or presenting with refractory hypertension in adolescent [5] The main treatment for IAA is maintaining the flow through PDA with prostaglandin and planning the reconstruction of aortic continuity to enable appropriate blood flow. It can be done in one stage or multistage sugery to repair the interruption and related cardiac septal defect [1]. In conclusion, CT or MR angiography plays an important role in diagnosis and guiding proper surgical management in cases of Interrupted Aortic Arch.

(Written consent was taken from the parent of the child for publication)

Differential Diagnosis List: Interrupted Aortic Arch Type A Subtype I, Coarctation of aorta, Aortic arch hypoplasia

Final Diagnosis: Interrupted Aortic Arch Type A Subtype I

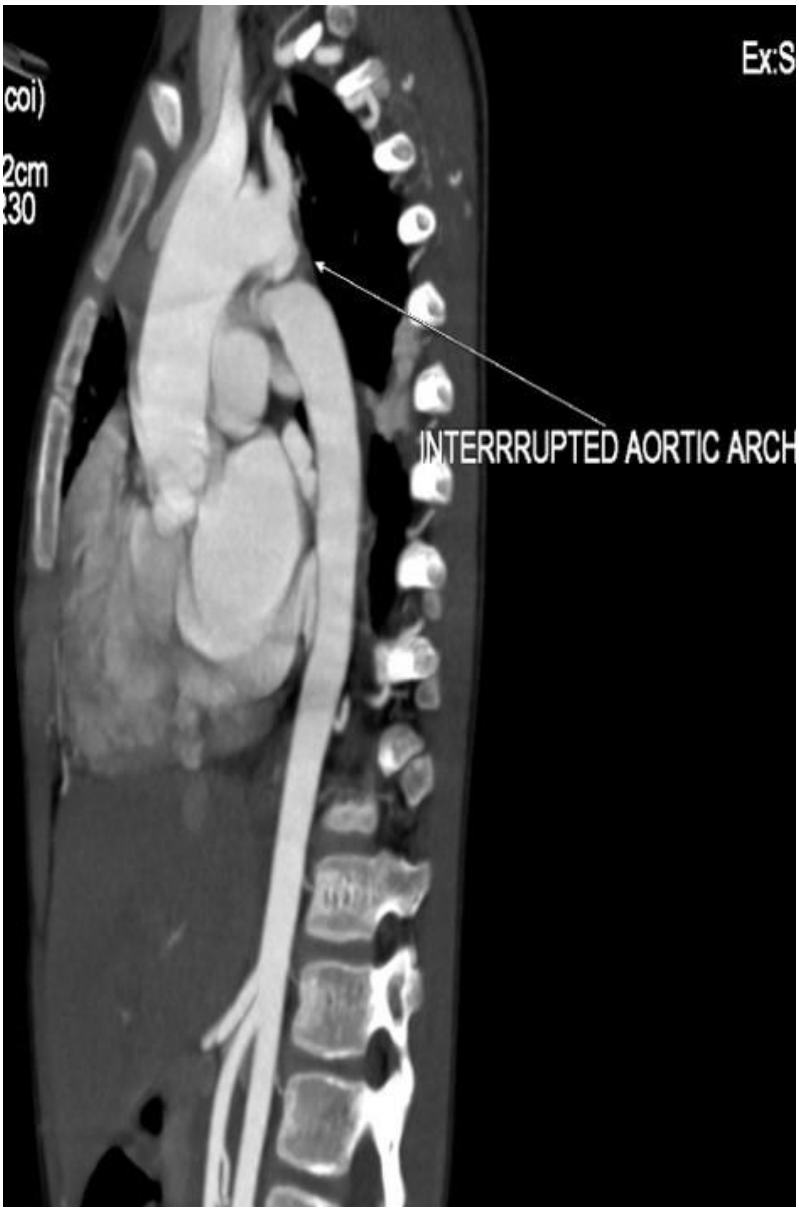
References:

Dillman JR, Yarram SG, D'Amico AR, Hernandez RJ. Interrupted aortic arch: spectrum of MRI findings. American Journal of Roentgenology. 2008 Jun;190(6):1467-74. Available from: https://www.ncbi.nlm.nih.gov/pubmed/18492893
Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. Radiographics. 2017 Jan;37(1):32-51. Available from: https://www.ncbi.nlm.nih.gov/pubmed/27860551
Celoria GC, Patton RB. Congenital absence of the aortic arch. American heart journal. 1959 Sep 1;58(3):407-13. Available from: https://www.ncbi.nlm.nih.gov/pubmed/13808756
Akdemir R, Ozhan H, Erbilen E, Yazici M, Gündüz H, Uyan C. Isolated interrupted aortic arch: a case report and review of the literature. The international journal of cardiovascular imaging. 2004 Oct 1;20(5):389-92. Available from: https://www.ncbi.nlm.nih.gov/pubmed/15765861
Murphy A, Yudi M, Calafiore P, Johns J, Matalanis G, Horrigan M. A RARE CASE OF INTERRUPTED AORTIC ARCH AS A CAUSE OF REFRACTORY HYPERTENSION IN AN 18-YEAR-OLD. Journal of the American College

of Cardiology. 2019 Mar 12;73(9 Supplement 1):2516. Available from: http://www.onlinejacc.org/content/73/9_Supplement_1/2516

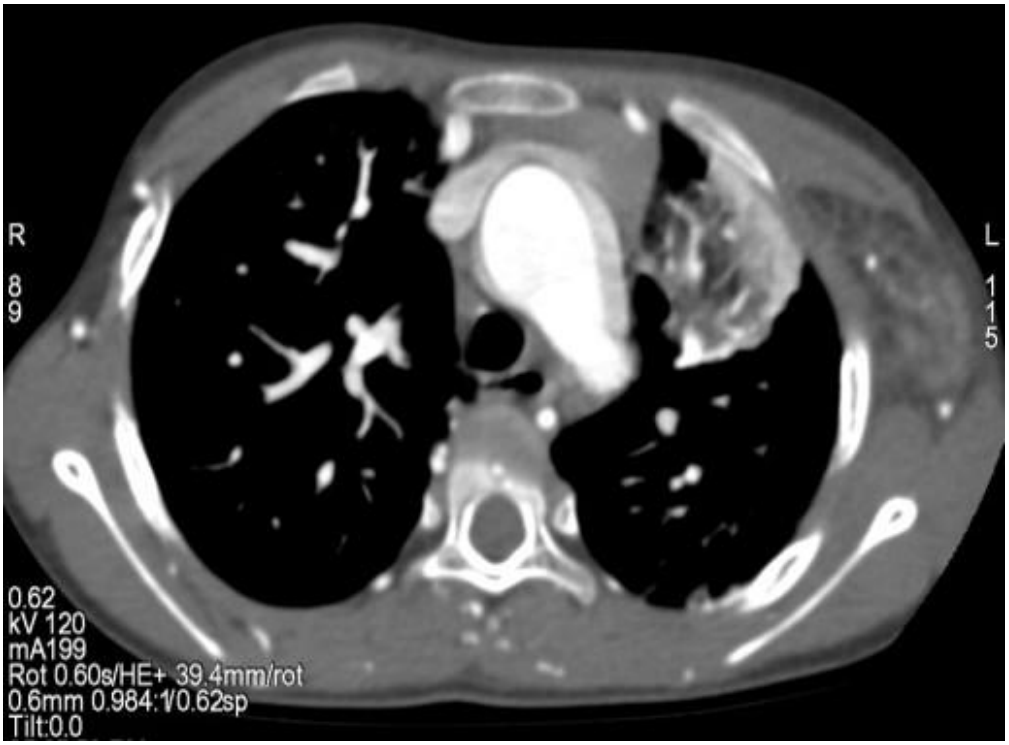
Figure 1

a



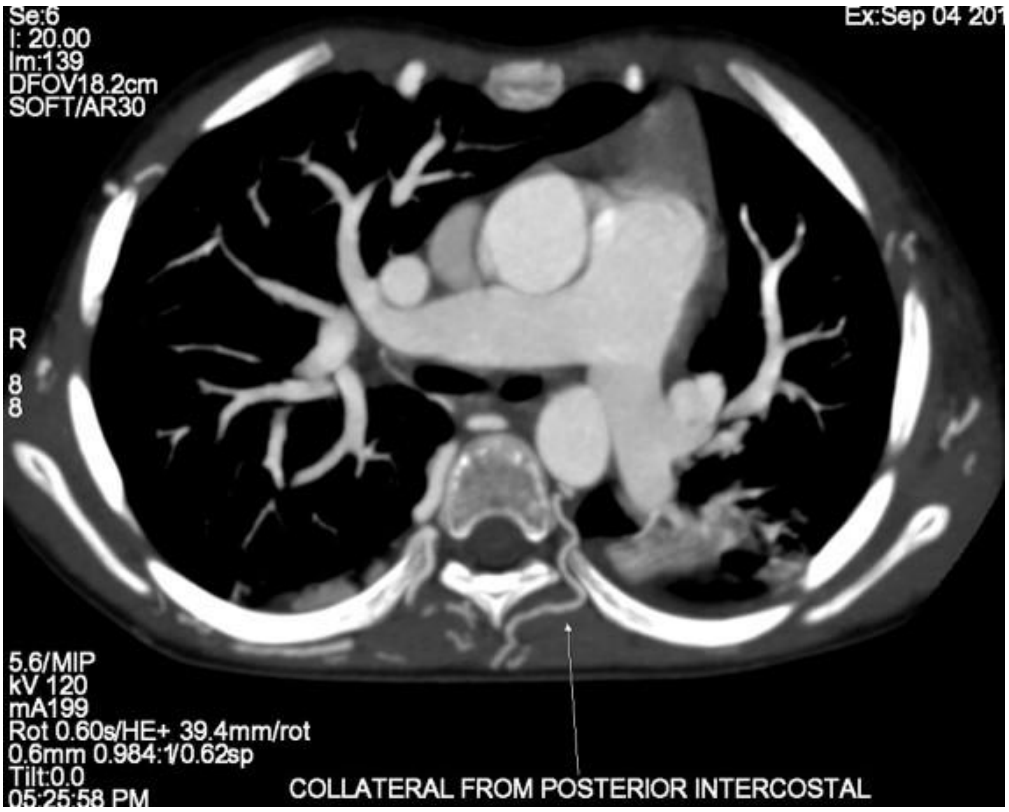
Description: Sagittal CT Aortogram shows abrupt luminal discontinuity in the aortic arch just distal to the origin of left subclavian artery **Origin:** © Hospital for Advanced Medicine and Surgery (HAMS), 2019

b



Description: Axial CT shows abrupt termination of left sided aortic arch. Patchy consolidation is seen in the anterior segment of left upper lobe. **Origin:** © Hospital for Advanced Medicine and Surgery (HAMS), 2019

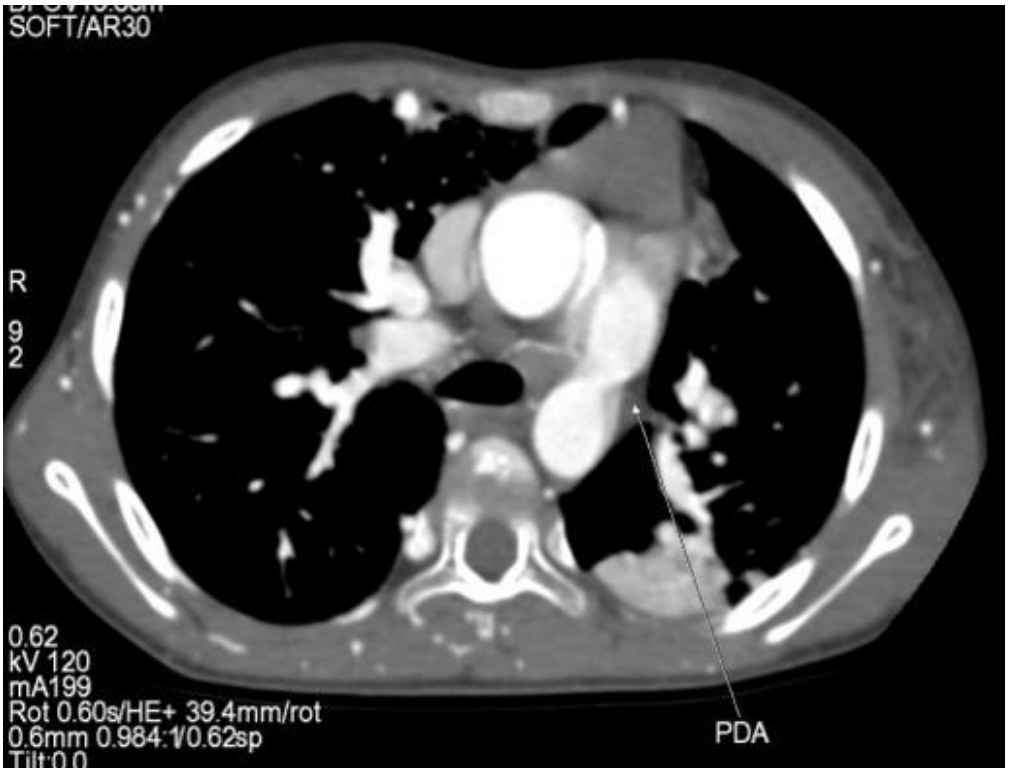
c



Description: Axial MIP thick slice CT image shows collateral from the posterior intercostal artery is seen reinforcing the descending thoracic aorta. **Origin:** © Hospital for Advanced Medicine and Surgery (HAMS), 2019

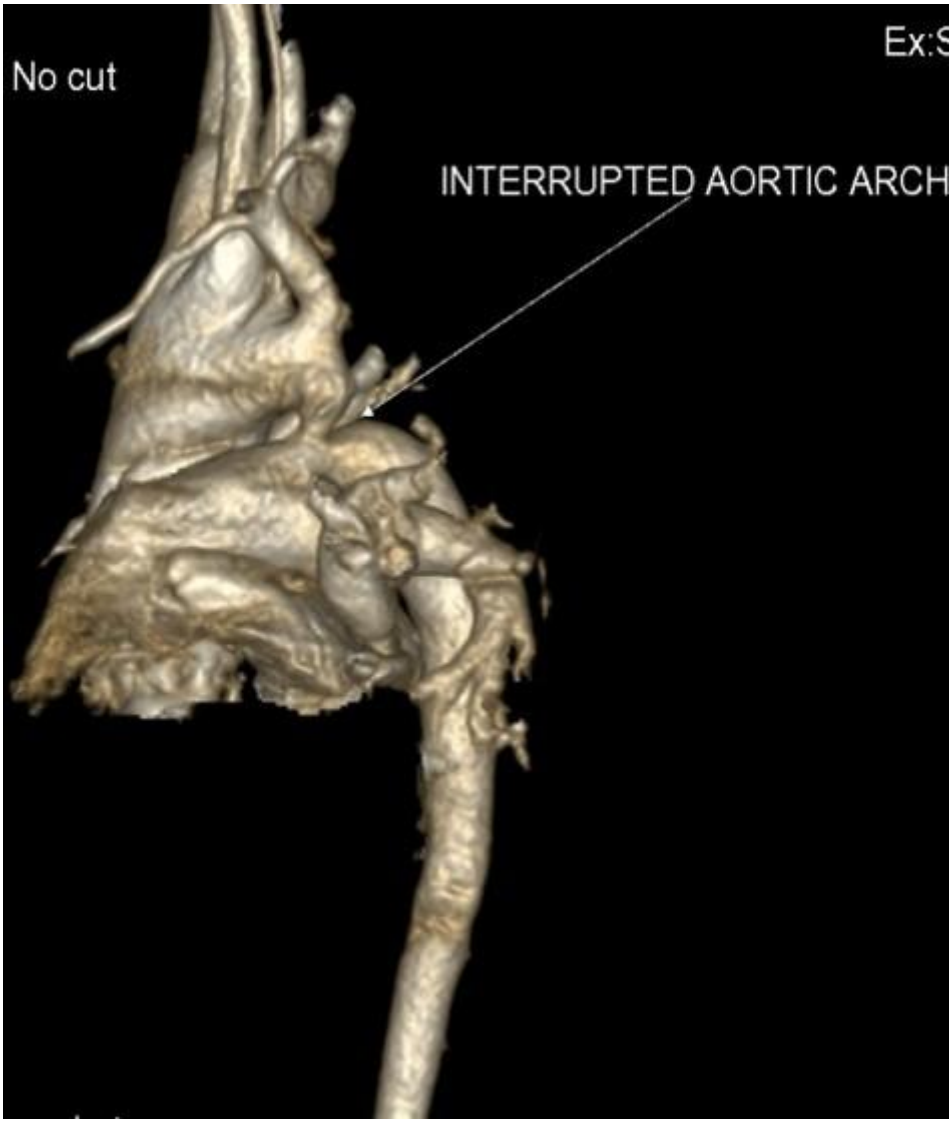
Figure 2

a



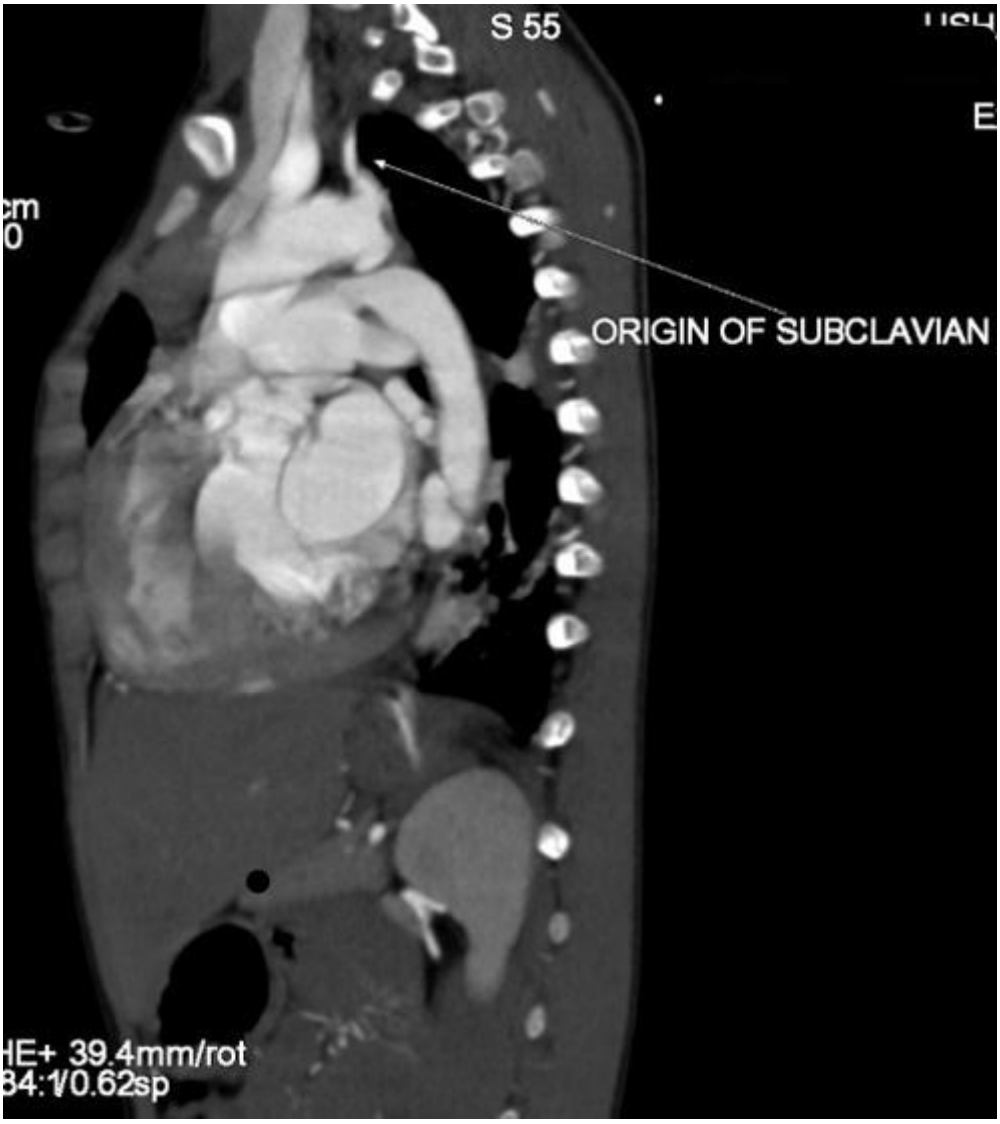
Description: Axial CT shows Patent ductus arteriosus (PDA) is continuous with descending thoracic artery. It is seen as hour glass appearance. Patchy consolidation is also seen in the peripheral aspect of superior segment of left lower lobe. **Origin:** © Hospital for Advanced Medicine and Surgery (HAMS), 2019

b



Description: 3D VRT shows interruption of aortic arch and patent ductus arteriosus is continuous as descending thoracic aorta. **Origin:** © Hospital for Advanced Medicine and Surgery (HAMS), 2019

c



Description: Sagittal CT image shows interruption of aortic arch is distal to the origin of left subclavian artery. Descending thoracic aorta is seen continuous from the patent ductus arteriosus. **Origin:**