

Interrupted aortic arch, mixed aortic valve disease and intra ventricular haemorrhage in an adult

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Abstract

Interrupted aortic arch (IAA) is a scarce and generally lethal congenital malformation. Patients with complete IAA rarely reach adulthood without surgical intervention. In this report, we describe the case of a 50-year-old man who was found to have IAA and mixed aortic valve disease after examination for hypertension. Later in the hospital course, he developed intra ventricular haemorrhage. The exact cause of intra ventricular haemorrhage and its association with IAA could not be further evaluated as unfortunately patient died 2 days after the haemorrhage. To our knowledge, this is the first report of an adult patient with all 3 abnormalities.

Keywords: Interrupted aortic arch, Mixed aortic valve disease, Intra ventricular haemorrhage.

Introduction

Congenital aortic arch anomalies represent a diverse set of malformations with a common embryologic origin and a wide array of clinical manifestations. Aortic arch anomalies are noted in 1-10/1,000 live births and account for approximately 15-20% of all congenital heart disease.¹

Interrupted aortic arch (IAA) is a congenital malformation characterized by complete luminal dissociation between the ascending and descending aorta, accounting for less than 1% of all cases of congenital heart disease² with incidence rate of 3 per million live births per annum.³ Although primarily considered a diagnosis of infancy, with high mortality rate of 75% by 10 days and 90% at 12 months of life (without surgical correction), there have been upto 40 cases reported in the adult cardiac literature.^{3,4}

IAA associated with other cardiac anomalies have been reported in literature however, to our knowledge, IAA associated with mixed aortic valve disease and intra ventricular brain haemorrhage has not been reported earlier.

Case Report

Ethical approval was obtained from the Chairman of Department of Radiology, Aga Khan University Hospital, Karachi, for publishing this case report.

In February 2016, a 50 year-old man attended the Emergency Department of Aga Khan University Hospital, Karachi, Pakistan, with complaints of severe headache, pain in nape of neck radiating to the back, increased sweating and agitated behaviour for the last two days. He was a known hypertensive for last 10 years taking tablet losartan 50 mg/day with poor compliance. The patient was also a smoker since last 20 years with one pack/day. On presentation, his blood pressures were 160/80 mmHg in the right upper limb and 110/80 mmHg in the right lower limb. The lower-limb pulses were feeble and barely palpable, whereas the upper-

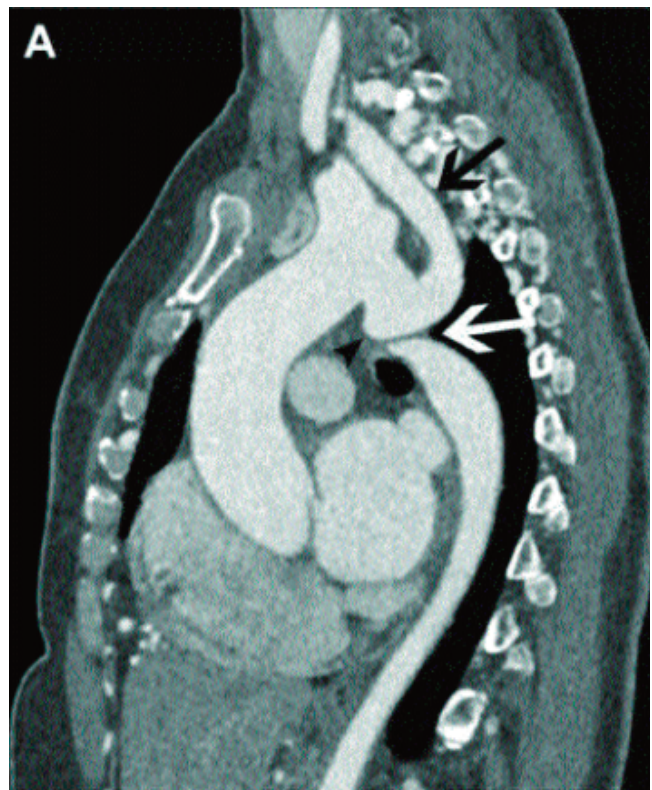


Figure-1: Type A Interrupted Aortic Arch.

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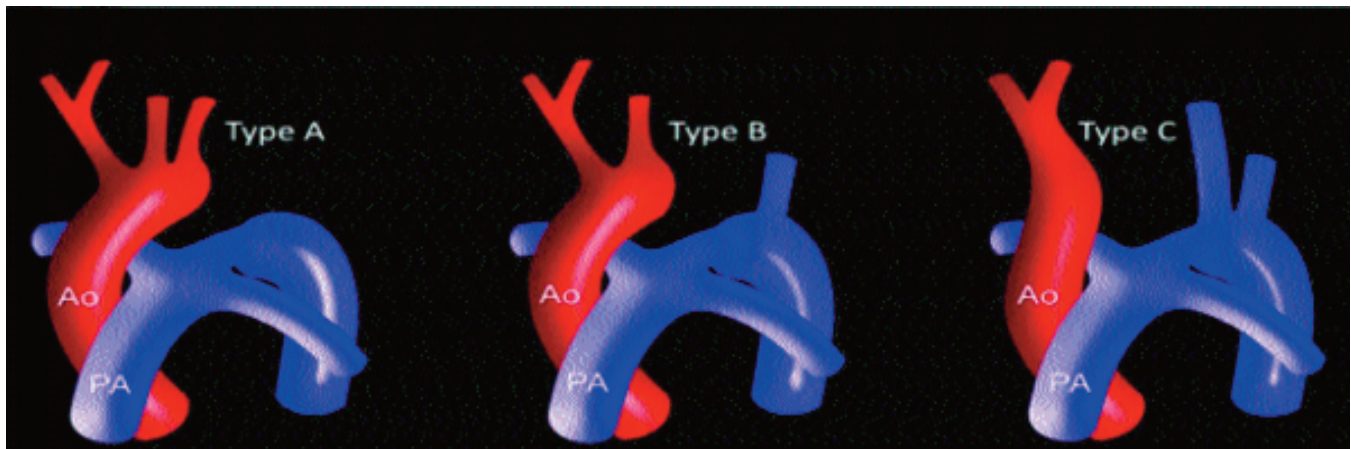


Figure-2: Types of interrupted aortic arch.

limb pulses were bounding. The initial electrocardiogram revealed sinus rhythm, with no signs of ischaemia, infarction, or left ventricular hypertrophy. Transthoracic echocardiography was then done which showed mild aortic stenosis and trace aortic regurgitation, mildly dilated left atrium with Grade IA left ventricular diastolic dysfunction. Mild asymmetrical septal hypertrophy was also noted. EEG was also done to rule out seizure disorder and it revealed mild encephalopathy. Imaging was done in the Department of Radiology, Aga Khan University Hospital. Chest radiographs were normal. Computed tomography angiography revealed interruption of the aorta at the junction of the aortic arch and the descending aorta after the origin of left subclavian artery (Type A) (Figure-1). Filling of the descending aorta was entirely from the extensive collaterals from the neck, upper and lower extremity and intercostal vessels. Bilateral external iliacs are also filled up by the collaterals arising from lower extremity arteries. No evidence of PDA or any communication of the aorta with the pulmonary arteries was present. Suggestion of bicuspid aortic valve was raised. Left heart catheterization was then done to obtain coronary and aortic angiogram that showed diffuse plaque in proximal 30% of left anterior descending artery. It was done initially through right femoral artery but the catheter tip could not be advanced beyond the descending thoracic aorta so, it was then done via right radial artery.

On fifth day of hospital admission, there was sudden drop in patients GCS.

MRI Brain was performed when showed signal drops out on SWI in the fourth ventricle and bilateral parietal sulci representing haemorrhages. Global involutional changes,

bilateral old lacunar infarcts and acute infarcts at splenium of corpus callosum and deep white matter along the occipital horn of the right lateral ventricle were identified. Patient was put on ventilator. The exact cause of intra ventricular haemorrhage and its association with IAA could not be further evaluated as unfortunately, on 7th day of admission patient became pulseless. Resuscitation was done but the patient could not be revived. The cause of death was obstructive hydrocephalus with brain stem compression secondary to intraventricular haemorrhage.

Discussion

First classified by Celoria and Patton, interrupted aortic arch (IAA) has been divided into 3 types based on the location of the interruption (Figure-2): Type A is distal to the subclavian artery, type B occurs between the second carotid and the ipsilateral subclavian arteries, and type C occurs between two carotid arteries.⁵ Each of these three types is further subdivided as follows: Subtype 1, normal subclavian artery; subtype 2, aberrant subclavian artery and subtype 3, an isolated subclavian artery that arises from the ductus arteriosus.^{6,7}

In neonates, type B is most common representing about 53% of the cases, followed by type A (43%) and type C (4%).^{5,8} The presentation of IAA in adulthood differs markedly. Type A is the most common form, likely due to an involution of the aorta at the site of an aortic coarctation (CoA), resulting in a functionally IAA,⁹ representing about 79% of the cases, followed by type B (16%) and type C (3%).⁵

Type B IAA is strongly associated with DiGeorge syndrome and chromosome 22q11.2 deletion.¹⁰ A right-sided descending aorta with aortic interruption is almost

always associated with DiGeorge syndrome.^{5,7}

It may occur as a simple or complex anomaly. In simple IAA, only ventricular septal defect (VSD) and patent ductus arteriosus (PDA) are seen. The complex form is associated with truncus arteriosus, transposition of the great arteries, double-outlet right ventricle, aorto pulmonary window or functional single ventricle.

Newborns with IAA usually present soon after birth with congestive heart failure and respiratory distress when the ductus arteriosus is closing.

In a reported study, the average age of diagnosis for the adults with IAA was 39.4 years (range, 18-72 years) and most (74%) patients were men.⁵

In adult patients, the usual presenting symptom is hypertension refractory to medical management and seen in 70% of the patients. Other symptoms include claudication (13%), aortic insufficiency (10%), and congestive heart failure (6%).⁵

However, occasionally, asymptomatic cases could also be observed; these patients are often diagnosed either by coincidence or when a hypertension etiology is being searched. Extreme complications can occur sometime such as subarachnoid haemorrhage leading to sudden onset headache or loss of consciousness.

Conclusion

IAA is rare in adults and it can be associated with other cardiovascular abnormalities. Accurate diagnosis of congenital IAA in the adult may be challenging, as it is difficult to distinguish from a progressively narrowed coarctation resulting in an acquired IAA.

MDCT/CTA has emerged as a fundamental tool for the diagnosis and pre-surgical work-up of IAA due to its high spatial resolution, large area of coverage, and short scan time.

Conventional angiography is usually necessary for definitive anatomical evaluation in patients with IAA.

Disclaimer: Consent of the guardian was taken prior to the writing of the manuscript. The abstract has neither been presented or published in a conference, nor the manuscript was part of a research, PhD or thesis project.

Conflict of Interest: None to declare.

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