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An Uncommon Cause for Hypertension in the Young; Case Study of A 28-Year-Old Lady and its Management

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Abstract: ***27 years old Female secondary hypertension presented with history of palpitation. On evaluation she was found to have Interrupted Aortic Arch Type - A. Aortogram also taken which showed extensive development of collaterals with Type A Interrupted aortic arch. She was managed conservatively due to well formed collaterals and absence of symptoms.***

Keywords: Interrupted Aortic arch, Celoria patton classification, Ductus dependent circulation

1. Introduction

Congenital heart disease has an incidence of 8 cases of every 1000 live birth worldwide [1]. Approximately 97% of babies born with a non - critical congenital heart disease have a life expectancy of one year of age, and approximately 95% are expected to live around 18 years of age. [2] [3] A rare type of congenital heart disease is an interrupted aortic arch (IAA), which affects approximately 1.5% of congenital heart disease patients. Interrupted aortic arch is an anomaly that can be considered the most severe form of aortic coarctation [4].

2. Case Presentation

27 years old Female known case of hypertension came with complains of palpitation. Her blood pressure was controlled despite not on medications at present. On examination, her Lower limb pulses were feeble. On examination, there was a systolic murmur in the inter scapular area with a visible collateral in interscapular area. Chest Xray was taken which showed Inferior Rib notching, with interruption of Aortic arch (Figure 1). Echocardiogram was done which showed Interruption of Aorta distal to Left subclavian artery. We differentiated it from coarctation by the absence of colourflow, and Doppler signals at the site of interruption. She was proceeded with CT aortogram which showed Type A Interruption with well-formed collaterals (Figure 2). There were multiple Paravertebral, Mediastinal, Scapular and intercoastal collaterals with Fully thrombosed Ductus aneurysm. Her Normal blood pressure probably must be due to well-formed collaterals that are decompressing. Angiogram taken also confirmed the above findings (Figure 3&figure 4). As she was asymptomatic, she is managed conservatively at present.

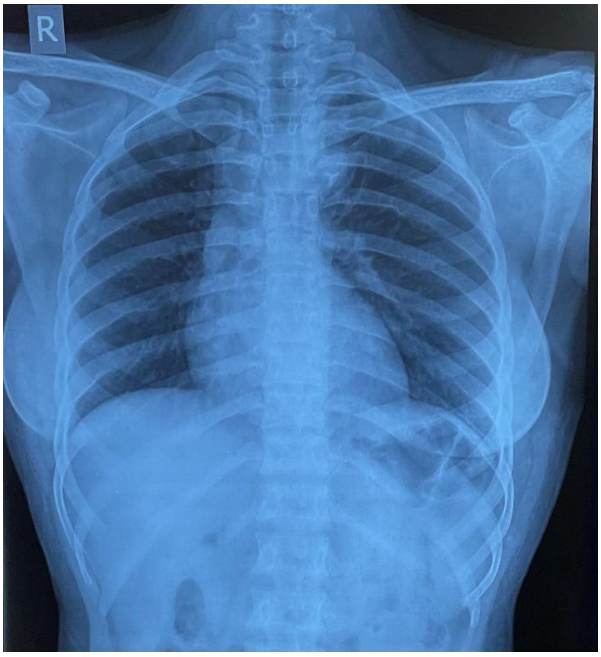

Figure 1: Interruption of Aortic arch, Dilated ascending aorta, Inferior rib notching

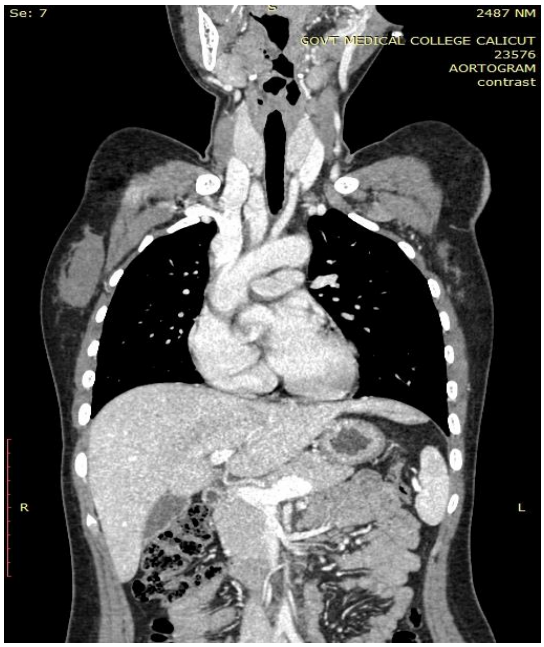

Figure 2: Interruption noted distal to Left subclavian artery

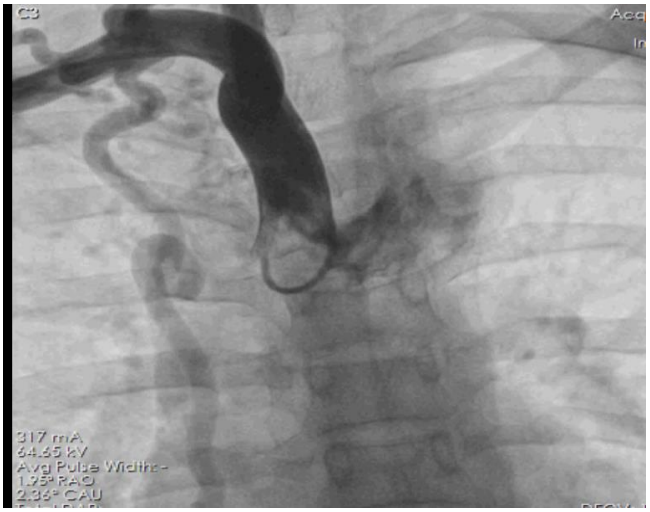

Figure 3: CT Aortogram showing Prominent Right internal mammary artery

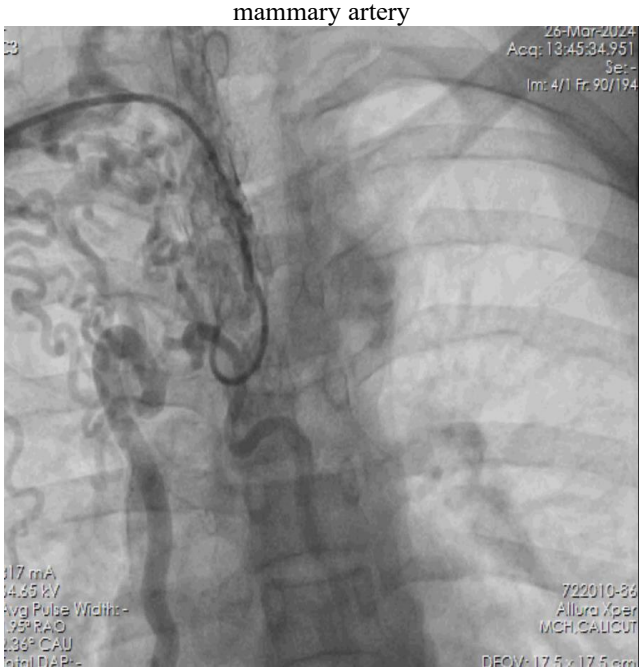

Figure 4: Showed Extensive collaterals

3. Discussion

Interrupted aortic arch results from an anatomical and luminal disruption between ascending and descending aorta. Systemic circulation depends on ductal flow and collaterals. The most common associated anomaly is Ventricular septal defect where there is posterior malalignment of conal septum leading to Left ventricular outflow obstruction. The other associated anomalies include Transposition of great vessels, Truncus arteriosus, Aorto - pulmonary window, Single ventricle, Aortic valve atresia, Right sided ductus and double outlet right ventricle. [5] Around one half of the patients with Interrupted aortic arch have 22q11.2 deletion – Di George syndrome. Approximately 50% of Patients with Interrupted aortic arch have Digeorge syndrome, that is usually TypeB [6]. Other syndrome associated with Interrupted aortic arch in CHARGE syndrome. [7]Mutation in charge syndrome is CHD7 on chromosome 8q12.1. CHARGE syndrome include Coloboma, Heart disease, Atresia choanae, Retarded growth and development and/or CNS anomalies, Genital hypoplasia and ear anomalies and/or Deafness syndrome. [8]

During fetal circulation, Ductus arteriosus provides blood to the distal extremities of the Fetus and the upper part of the body is supplied from the left ventricle to the aorta. After birth when the pulmonary vascular resistance falls, there is closure of ductus arteriosus leading to cyanosis and shock followed by death if the necessary measure are not instated in time.

Celoria and Patton classification divides Interruped aortic arch into 3 types based on site of disruption. Type A denote disruption distal to left subclavian artery. Type B denote disruption between left carotid artery and left subclavian artery. This anomaly is most common. Type C means disruption is between innominate artery and left carotid artery. This is the rarest of the types. [9] [10]

The physical exam will reveal absent pulses with a difference in Blood pressure between the right arm and lower extremities. The baby may be asymptomatic until the ductus arteriosus closes and the patient develops tachypnea, feeding difficulties, respiratory distress, cyanosis, and anuria which, ultimately, can lead to shock and death. Sometimes, there may be an oxygen saturation discrepancy between the left and right side of the body. [11]

The physical exam will reveal absent pulses with a difference in blood pressure between the right arm and lower extremities. When the diagnosis is suspected, it is necessary to perform Chest xray, ECG and Echocardiogram. The Chest x - ray shows Cardiomegaly with increased pulmonary blood flow. Echo will define the site of interruption. Cardiac angiography, Computed tomography and Magnetic resonance angiography gives a more comprehensive understanding of the lesion and anatomy before surgical repair. [12]

Treatment should start with initiating Prostaglandin E1 to avoid sudden cardiac collapse and death. The role of Prostaglandins is to maintain patency of the ductus arteriosus and maintain perfusion. Once diagnosed the treatment is immediate surgery. The objective of surgery is to form unobstructed continuity between ascending and descending aorta and to repair the coexisting defect, which is most commonly Ventricular septal defect. The repair of Interruption is done by using a native arterial tissue, a homograft or autograft vascular patch.

The role of the prostaglandins is to maintain the patency of the ductus arteriosus, thus guaranteeing the perfusion of the lower part of the body until surgical correction is done. In the presence of shock, the patient should be managed with inotrope support, and treatment should be adjusted depending on the clinical response of the patient. [13]

Once diagnosed, the treatment is immediate surgery. The objective of the surgery is to form unobstructed continuity between the ascending and descending aorta and to repair associated defects with the most common atrial and/or ventricular septum defect. The repair is done using either native arterial tissue, a homograph, or autograph vascular patch. For ventricular septal defect, repairs are closed with a synthetic patch. This synthetic patch is made up of polyester or polytetrafluoroethylene. In cases of significant outflow tract obstruction, it may be necessary to perform a complex combination of the Norwood and Rastelli procedures [14] [15].

4. Conclusion

Meticulous clinical examination helps to identify and diagnose even rarest of diseases. Most of the cases needs early surgery; In our case, we opted conservative management as her blood pressure was well controlled and peripheral perfusion was adequate with well established collaterals. Currently she is on close follow up.

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