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Aortic Aneurysmal Dilation in a 6-month-old Baby in Rajaie Cardiovascular, Medical and Research Center: A Case Report

Running Title: Aortic aneurysmal dilation in a six month child

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Abstract
Background: Aorta dilatation at the root surface of the aorta can have a variety of causes. Many congenital or acquired conditions can lead to weakness of the aortic wall. Ascending aortic dilatation is rare in childhood and is often associated with forms of connective tissue disease or predisposing cardiac malformations, especially bicuspid aortic valve disease.
Case Presentation: Herein, we present the case of a six-month-old boy with severe dilation of the ascending aorta. The patient was referred to the doctor by the parents because of persistent cough. After several examinations, the infant's heart disorder was noted, and he was referred to Shahid Rajaie Heart Center. After performing echocardiography, severe dilatation of the ascending aorta was diagnosed.He was transferred to the operation room. Total thymectomy was performed, and then resection and repair of the ascending aorta and arch of the aorta was done. After ventilating the cardiac cavities, the aortic clamp was removed, and the patient's heart found a rhythm as it warmed up. After hemostasis and placement of the chest tube drain and installation of a temporary pacemaker wire, the patient's sternum was closed, and he was transferred to the ICU. After 2 days, the patient was transferred from the ICU to the ward in good condition and then discharged from the hospital without any problems.
Conclusions: Aortic dilation is a rare disease in children. The sign and symptoms may be different in any patient, and complete diagnostic tests must be performed before performing the procedure.
Keywords: Aortic root, Aortic insufficiency, Aneurysm aorta,Dilation of aorta

Background

Aortic surgery in children with chronic aortic regurgitation is used in order to avoid the irreversible left ventricular (LV) dysfunction, which could increase a patient's life expectancy.
In children, the complications of ascending aortic dilatation and surgical indications are less clear.[1].

It should be noted that in children, due to the mismatch of aortic size with the patient's age, a larger ascending aortic diameter is acceptable[2]. This report is our experience of a six-month-old baby (weight: 6700 grams, height: 66 cm) who presented with severe dilatation of the ascending aorta and mild to moderate aortic insufficiency (AI).

Case Presentation

The patient was referred to the doctor by the parents because of persistent cough, and after several examinations, the infant's heart disorder was noted, and the infant was referred to Shahid Rajaie Heart Center, Tehran, Iran for further examination.

Aortic dilation was observed in chest X-ray (CXR) (figure 1). After performing echocardiography by an attending cardiologist and with the diagnosis of severe dilatation of the ascending aorta, surgery was scheduled.After obtaing the informed consent of the parents,he was transferd to the operation room.After entering the patient to the operating room and connecting the monitor, he had a blood pressure (BP) of70/35 with non-invasive blood pressure (NIBP), a heart rate of 140, and SpO2of 98%. After induction with 0.1 mg/kg midazolam,5 mc/kg fentanyl followed by atracurium 1 mg/kg. After the required time,an endotracheal tube (ETT) with a size of 4.5 was placed for the child.After anesthesia induction, a large bore peripheral IV line was inserted.

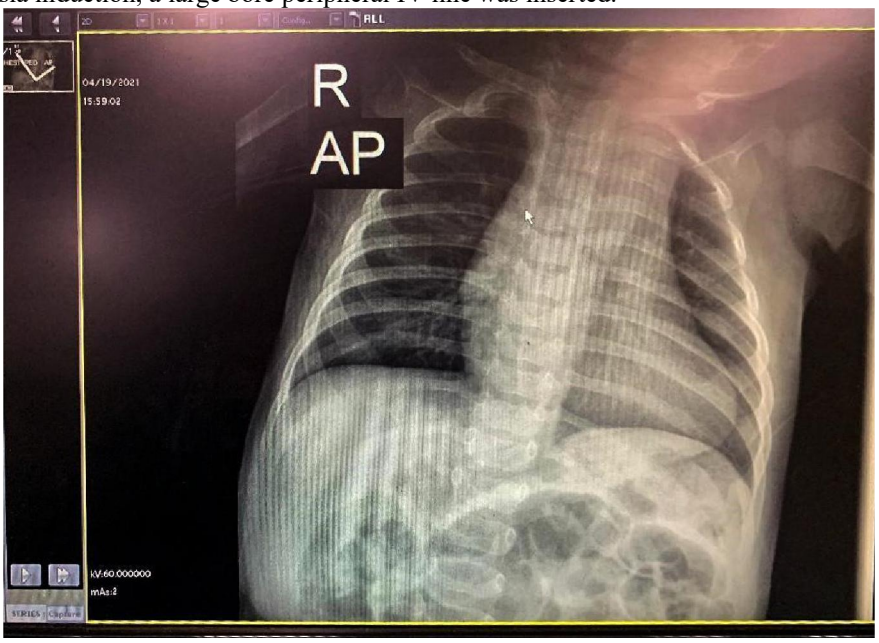

Figure 1: Chest X-ray patient: Dilation of aorta is present

The arterial line was taken from the right radial and left femoral arteries (BP: 85/55), and then arterial blood samples were sent to the laboratory for examination. The central venous line (CVL) was then implanted in the right internal jugular region, and the urinary catheter was also put in place (urine flow was maintained during surgery and there was a sufficient volume of urine). For cerebral monitoring, cerebral oximetry was done, and an esophageal temperature probe was placed. After proper positioning, the patient was covered, and the surgery began at 14:30. At 16:00, a partial circulatory arrest was performed for 23 minutes.

Total thymectomy was performed, and then resection and repair of the ascending aorta and arch of the aorta was done (figures2 and 3). After ventilating the cardiac cavities, the aortic clamp was removed, and the patient's heart found a rhythm as it warmed up.

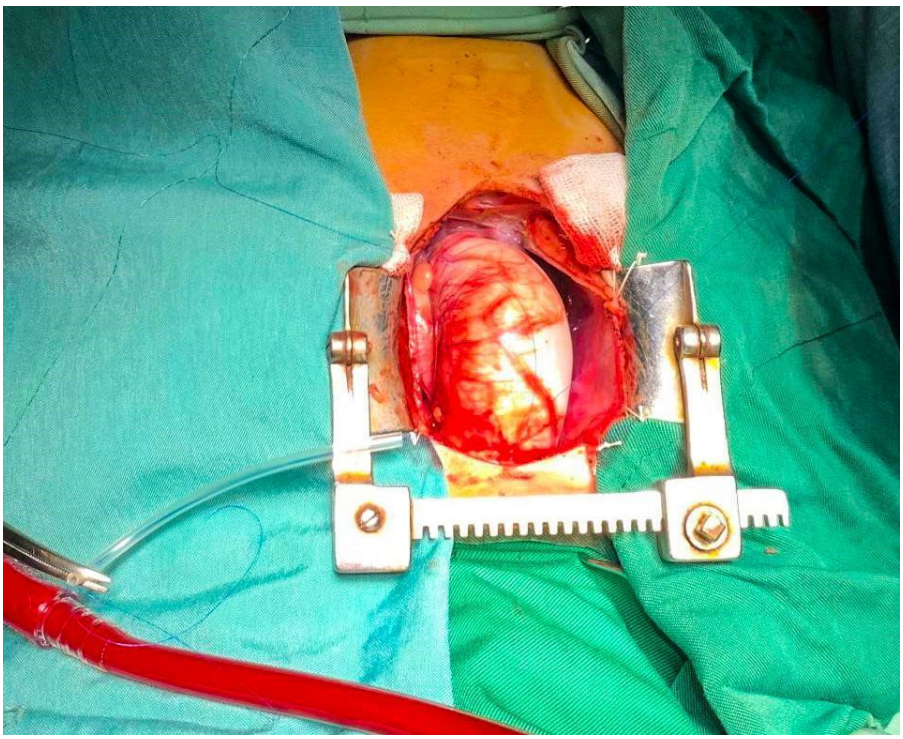

Figure 2: Ascending aorta in fild of surgery

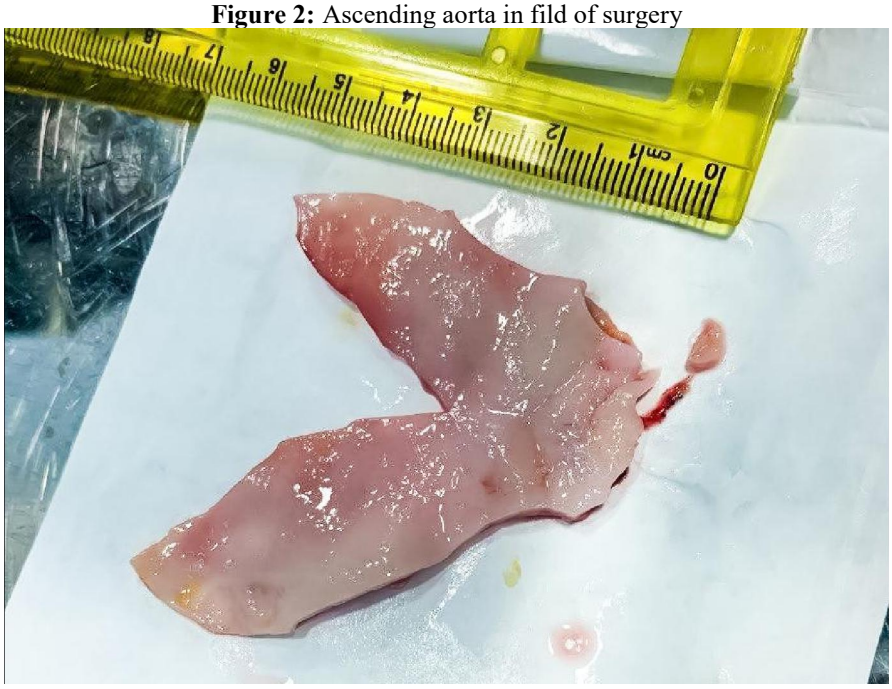

Figure 3: Aortic fragment after aortoplasthy

The patient's vital signs became stable after being put off-pump with 0.05 µg/kg/min Epinephrine and 0.5 µg/kg/min Milrinone. After hemostasis and placement of the chest tube drain and installation of a temporary pacemaker wire, the sternum of the patient was closed, and after 5 hours the child was admitted to the intensive care unit (ICU). In none of the stages of cerebral oximetry did the patient reach less than 80% of the normal level. After 2 days, the patient was transferred from the ICU to the ward in good condition and then discharged from the hospital without any problems. Inpatient pathology, the aortic wall with elastic fibers fragmentation, and cystic medial degeneration were reported.

Discussion

Aortic aneurysm is a common finding in the elderly that occurs as a result of aging, hypertension (HTN), or atherosclerosis but is rare in childhood. Aortic aneurysm is a permanent dilatation of the aorta that has at least a 50% increase in diameter and involves all layers of blood vessels. The location of the aneurysm and change of size determine the surgical strategy and perioperative complications.Despite being rare, the aortic root dilation are well-known cardiovascular manifestation,which is usually seen in people with underlying connective tissue disease[3].

Aneurysm of the aortic root and/or ascending aorta iscommonly associated with bicuspid aortic valve (BAV) disease, which could be found in about 1.5 percent of adults as a common congenital heart abnormalities. It has also been reported that BAV can be a risk factor for dilatation and dissection of the proximal aorta. [5]. Thoracic aortic aneurysm is often asymptomatic and is discovered accidentally. Common symptoms in a thoracic aortic aneurysm are as follows: chest and back pain because of dissection of the aneurysm,rupture or erosion because of a large thoracic aortic aneurysm causing a mass effect which can compress local elements in that area such as putting pressure on the laryngeal recurrent nerve and hoarseness,tracheal compression and primary bronchitis and development of superior vena cava syndrome dyspnea because of central venous HTN, or acute pain with or without hypotention (HOTN). Although ascending aortic aneurysm rupture may be associated with cardiac tamponade, a descending thoracic aortic rupture may be related to hemothorax, bronchial aortic fistula, or pharyngeal aortic fistula. Indications for auricular surgery include being symptomatic (despite the size and signs of rupture), ascending aortic diameter of <5.5 m, or descending aortic aneurysm <7 cm. The type of surgical repair depends on the function of the valve at the auricle and its expansion. Preventing HTN results in forwarding flow through the aortic valve and reduces the risk of aortic rupture.

The choice of right or left radial artery depends on the surgical approach to repair the aortic arch. Sometimes the catheter of both radial arteries can simultaneously providethe monitoring of cerebral and systemic perfusion. If right axillary, subclavian or brachiocephalic artery cannulation is to be performed, monitoring of nasopharyngeal, tympanic, and bladder temperatures is important for cardiopulmonary bypass (CPB) or antegrade cerebral perfusion (ACP). To assess the central and brain temperature for deep hypothermic circulatory arrest (DHCA) and to deal with it, monitoring of the jugular bulb oxygen and EEG could be helpful to determine cerebral metabolic activities.. Intraoperative monitoring with transesophageal echocardiography(TEE) is essential for surgical interventions. In patients with AR, the TEE can act as a guide for CPB cannulation, such as a retrograde cardiopulmonary cannula and LV volume monitoring, and ensure LV drainage to make sure it does not collapse. TEE is an acceptable procedure in thoracic aortic procedures[6].

No precise information is available on the aortic aneurysms occurrence in children. Aortic aneurysm is more common in the ascending aorta; however, it could be observed in other areas, such as the aortic branches amd descending aorta . Despite being rare, one of the causes of mortality in children and adolescents could be rated to aortic aneurysm. This may have genetical(such as Ehlers Danlos Syndrome, Marfan's syndrome, , Loeys-Dietz syndrome, Arterial tortuosity syndrome,Cotis Lock syndrome, Alagille syndrome, and Noonan syndrome) or non-genetical (state aorta, aortic coarctation, tetralogy of Fallot, or aortitis syndrome) causes[7-9].

In marfan syndrome aortic dissection occurs most often during adulthood ,but it rarely can affect younger patients[10].Ascending aortic disease can have devastating effects on the affected person and lead to severe diseases such as aneurysm, rupture, dissection, hemopericardium, upper valve stenosis, and AI secondary to the dilated annulus.The clinical signs of an aortic aneurysm depend on factors such as underlying disease, the aneurysm size and location , and the existence of aortic dissection.

Patients with an aortic aneurysm should pay attention to the genetic causes of associated phenomena such as hypermobile joints, thoracic deformity, long thin fingers, kyphoscoliosis, long palate amd inguinal hernia. Checking the family history would be mandatory if these symptoms were observed in patient.. In patients with an aneurysm, decreasing the femoral pulse and systolic murmur could be detected in the aortic region on cardiac examination. Diastolic murmur can be heard secondary to AI, and due to mitral prolapse, pan-systolic murmur can be heard secondary to mitral valve insufficiency. Transthoracic and transesophageal echoes are the first interventions to examine the aortic aneurysm. Cardiac catheterization, magnetic resonance imaging, and computed tomography are other imaging manners.

The factors such as life expectancy, the underlying disease, the aneurysm size and location and also the presence of dissection should be consdired in the children with aneurysms during surgical operation.. Employing beta-blockers and angiotensin-converting enzyme (ACE) inhibitors in the patients with Marfan's syndrome has been shown to delay surgery time[11]. The presence of genetic abnormalities such as the fibrillin gene may be associated with aortic dilatation in the presence of congenital heart disease. The incidence of aortic dilatation in patients with

tetralogy of Fallot is significantly common, particularly as there are lags in repairing or when the patient has a systemic pulmonary shunt to the lungs after palliative repair.Aortic coarctation can be associated with aortic aneurysm through several mechanisms. High proximal pressure causes stress to the wall and may cause an ascending aortic aneurysm. High blood pressure and damage to the walls of arteries can also lead to aneurysm formation. Dilatation of the aortic root leads to regurgitation and changes in elasticity and compliance, which leads to long-term complications in ventricular function. Blood pressure control is an important point before (to reduce rupture) and after (to prevent damage to the sutures) bypass surgery[12].

Conclusions

Aortic dilation is a rare disease in children, and the patients may not have specific cardiac symptoms and may be diagnosed with involvement of other systems such as the respiratory system. In patients with prolonged respiratory symptoms such as prolonged cough and shortness of breath more diagnostic tests should be performed, and complete clinical examination and appropriate diagnostic measures are necessary in such patients. If there are extra cardiac disorders, there is a possibility of a worse prognosis, and extra cardiac problems should be evaluated.

Abbreviations

LV: Left ventricle; AI: Aortic insufficiency; CXR: Chest X-ray;ETT: Endotracheal tube; CVL:Central venous line; ICU: Intensive care unit; HTN: Hypertension;BAV: Bicaspid aortic valve;HOTN: Hypotention;CPB: Cardiopulmonary bypass;ACP: Antegrade cerebral perfusion;DHCA: Deep hypothermic circulatory arrest;TEE: Transesophageal echocardiography;ACE: Angiotensin-converting enzyme

Declarations

Authors' contributions: All authors were involved in the management of the patient and generating the concept. All authors made an intellectual contribution and reviewing the paper. All authors read and approved the final manuscript

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Ethics Approval and consent to participate: Not applicable.

Consent for publication: Written informed consent was obtained from the parents of the patientfor publication of this case report and any accompanying images.

Competing interests: The authors declare that they have no competing interests.

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