

## A Rare Case of Giant Coronary Aneurysm in a 5- Year- Old Child

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### **Abstract**

Coronary aneurysms are typically seen in association with Kawasaki disease and only in rare instances, they are reported to be congenital or idiopathic. Here we report a case of a five- year- old boy who was found to have an idiopathic right coronary artery aneurysm. He presented with complaints of recurrent respiratory tract infections. His examination and investigations did not reveal any of the common etiological conditions. Echocardiography showed the right coronary artery was arising from the right coronary sinus and was dilated proximally. Cardiac catheterization revealed dilated proximal right coronary artery draining into a large pulsatile aneurysm. The child was surgically treated with an aortocaval bypass, resection and surgical ligation of the aneurysm. He has no further complications and is on follow- up.

**Key Words:** Child, Coronary artery aneurysm, Idiopathic aneurysm, Cardiac catheterization.

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## 1- INTRODUCTION

Childhood aneurysms are a rare entity and an often neglected topic in pediatric literature. Aneurysmal dilation of the coronary arteries was first described by Bourgon in 1812, and is characterized by abnormal dilatation of a localized or diffuse segment of the coronary arterial tree. Coronary artery aneurysm (CAA) is defined as a focal dilation greater than 1.5 times the diameter of the adjacent normal coronary artery and involving < 50% of the length of the coronary artery (1). It is a rare finding even on angiography, most of which remain clinically asymptomatic (2). Classifying coronary aneurysms into small, medium and large depending on their internal diameter tends to underestimate its true incidence (3). Many causes have been attributed to the etiology of giant aneurysm in paediatrics; Kawasaki Disease being the most common. Very rarely, no cause can be established as the etiology. Only 14 cases of such "idiopathic" aneurysm have been reported in pediatric and surgical literature from various parts of England, Japan and the United States (4). Here we report a case of a five year old boy referred to us for cardiac evaluation was incidentally found to have a giant right coronary artery aneurysm.

## 2- CASE REPORTS

A five- year- old boy was referred to our centre (AJ Hospital and Research Centre, Mangalore, India) for cardiac evaluation with complaints of recurrent respiratory tract infections. Neither his history nor did his examination reveal any significant information. Primary investigations including electrocardiograph and chest X-ray were normal. Echocardiography showed right coronary artery was arising from the right coronary sinus and was dilated proximally. It was measuring 6 mm at the ostium and proximal right coronary artery was 5.5 mm

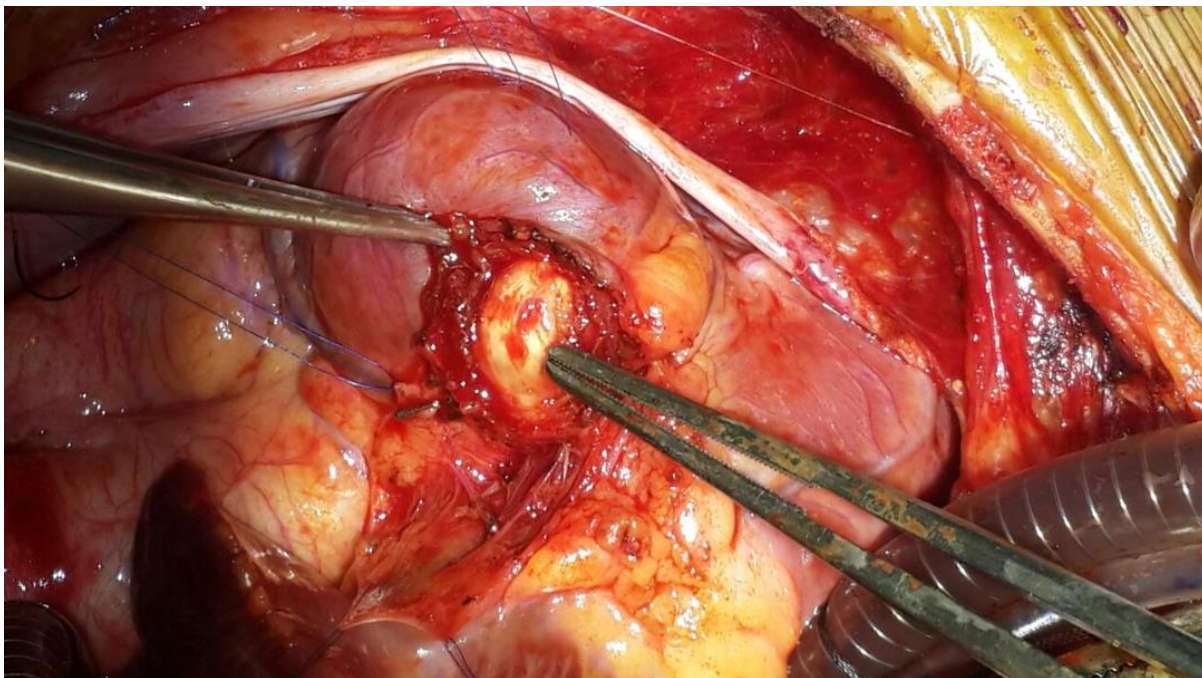
distal right coronary artery was draining into a circular chamber. There was no evidence of coronary cameral fistula. Ventricular chamber size was normal and left main coronary artery was normal. Neither his examination nor his investigations revealed any of the common etiological conditions associated with coronary artery aneurysm. In view of the above findings, cardiac catheterization was planned to delineate detailed coronary anatomy and intervene if necessary. Selective coronary injections revealed dilated proximal right coronary artery draining into a large aneurysm measuring 32 mm x 28 mm in diameter (**Figure.1**). Blood supply to the myocardium was arising proximal to the aneurysm. The aneurysm was pulsatile. Surgical ligation of the aneurysm was planned in view of potential complication of rupture.

Intramyocardial pulsatile swelling in the right ventricle wall arising from right coronary artery was noted intraoperatively. Right coronary artery was isolated. Proximal right coronary artery was dilated. Just proximal to the neck of the aneurysm, the distal branches of right coronary artery were seen taking off. Since ligation of neck of aneurysm would cause occlusion of distal branches, we went for an aorto-caval bypass and cardioplegia was given. Intramyocardial portion of aneurysm was dissected (**Figure.2**).

Aneurysmal neck was closed off with pericardial patch using a 7.0 prolene continuous suture. Small opening in the aneurysmal sac was closed directly and the myocardium was approximated over the sac. We came off bypass with minimal inotropic support. No ST changes were detected post operatively. Post procedure echocardiogram showed dilated proximal right coronary artery with normal branching. Child was discharged on the fifth post-operative day and is now on follow up.



**Fig.1:** CT Angiogram showing giant aneurysm.



**Fig.2:** Intra operative picture showing the giant aneurysm.

### 3- DISCUSSION

The overall incidence of CAAs ranges from 0.3 to 5.3%, while the incidence of giant CAAs is as low as 0.02%. They are commonly associated with congenital arterial fistulae with an incidence of 5.9% (5); an association which was not seen in our case. Coronary atherosclerosis and Kawasaki disease are found to be the principal causes of coronary artery aneurysms. Atherosclerosis being responsible for 50% of the cases in adults and Kawasaki disease may lead to coronary artery aneurysm in 25 – 30% of untreated children. In young patients, aneurysmal disease is uncommon mostly associated with underlying causes including Marfan's syndrome and Ehlers-Danlos syndrome, polyarteritis nodosa, Takayasu disease, syphilis and metastatic tumours (6). They are rarely found secondary to infection, neurofibromatosis, and trauma from previous umbilical artery catheterization, and tuberous sclerosis (4).

Giant coronary artery aneurysms have been reported from the United States and Korea in the recent past secondary to juvenile polyarteritis nodosa and Kawasaki disease (7, 8). It has been suggested by other authors that all of these "idiopathic" aneurysms are variants of Ehlers-Danlos syndrome, lacking the clinical skin and joint changes but with a propensity for aneurysm formation. An alternative approach would be to consider these cases as part of a syndrome of idiopathic childhood aneurysms (4). Patients with giant CAAs can present in a variety of ways, such as with superior vena cava syndrome or with a mediastinal mass, which can often be misdiagnosed as a cardiac tumor (5). However, in most cases, they remain asymptomatic as how the presentation was in our case. It has been suggested that the syndrome of idiopathic childhood aneurysms may progress during active growth until adolescence, at which

point it may stabilize (4). Factors that contribute to development of complications include distal embolization with myocardial infarction, rupture with associated fistula, cardiac tamponade or haemopericardium, dissection, vasospasm and vessel compression (9). Although no fixed guidelines have been developed for the management of giant CAA in the acute setting, further dilatation and even rupture of the CAA, as well as thrombus formation, should be avoided (7). Its importance lies in the potential fatality of the condition, if left untreated; which was the reason why we chose to operate. Large aneurysms are a diagnostic challenge; the differential diagnoses for these include cysts and other masses. The variation in the presentation of CAAs highlights the importance of utilizing a variety of diagnostic approaches (5). Coronary artery aneurysm may be detected by non-invasive tools including echocardiography, computed tomography and magnetic resonance imaging (6). Although coronary angiography remains the gold standard for the diagnosis of aneurysms, we planned for a cardiac catheterisation in order to be able to diagnose as well as intervene if necessary. Interventional techniques have been described for coronary aneurysms including obliteration with polytetrafluoroethylene-covered stents and surgical therapy.

Since the cardiac catheterisation revealed a pulsatile swelling, surgical ligation was considered to be the best option. The appropriate therapy is uncertain and controversies still persist regarding the better treatment option being surgical or conservative. Treatment is best customised according to patient's clinical presentation (1). The challenge in diagnosis of a coronary artery aneurysm lies in the fact that the child may be asymptomatic for a variable period of time and the aneurysm is frequently an unrecognised, incidental finding.

#### 4- CONCLUSION

This case report illustrates the importance of being aware of the varied presentations of giant coronary aneurysm and the potential danger it holds in going unrecognized.

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**6- CONFLICT OF INTEREST:** None.

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