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CONGENITAL CORONARY ARTERY ATRESIA: A RARE CAUSE OF DILATED CARDIOMYOPATHY

Paediatrics

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ABSTRACT

Congenital atresia of coronary artery is a rare disease. We report a case of congenital coronary anomaly in a 3 year old male child. Patient was admitted with symptoms of heart failure. Echocardiography revealed a dilated heart. A diagnosis of Dilated cardiomyopathy was made (? Post myocarditis). Further coronary angiography showed atresia of left coronary artery as a cause of dilated cardiomyopathy.

KEYWORDS

Dilated Cardiomyopathy, Atresia Of Left Main Coronary Artery

Introduction:

Congenital anomaly of the coronary arteries is a rare disease occurring in 1%-2% of all congenital heart disease. Atresia of Left Main Coronary Artery (LMCA) is one of its least frequently observed variations. Only a few cases of left main coronary artery atresia in children have been described manifesting as DCMP.[1,2,3] To best of our knowledge this is first case reported from India. Its initial signs and symptoms are similar to those of an Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA). Angiography is confirmative. For its rarity and uncommon presentation we are reporting this case of a 3 year old child who presented with dyspnea which was subsequently diagnosed to have coronary artery atresia.

Clinical Summary: A 3 years old male child was admitted with diagnosis of Myocarditis/Cardiomyopathy with poor left ventricular function. He was asymptomatic 6 months prior to presentation. On physical examination he was afebrile with tachycardia, tachypnea (RR=56/min), subcostal and intercostal retractions, basal crepts and hepatomegaly with apical soft systolic murmer (II/IV) on auscultation. His BP was 88/48 mm Hg (50th centile). X-ray showed cardiomegaly with CT Ratio of 0.70. ECG showed diffuse non specific ST-T changes. Echocardiography revealed globular dilated left ventricle (LVIDd- left ventricular internal diameter in end diastole = 4.7cm), mild-moderate mitral regurgitation with decreased left ventricular systolic function (EF = 30%).

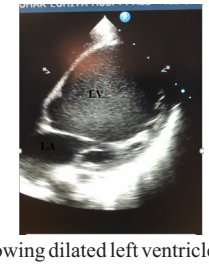

Image 1: 2D Echo showing dilated left ventricle (LV)

The right coronary artery, 2.5mm at ostia, was seen originating from right coronary sinus. Ostia of left MCA was difficult to delineate.

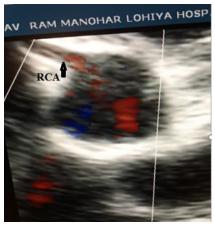

Image 2: 2D Echo showing Right coronary artery arising from Aorta.

Branches of LCA were seen i.e. Left Anterior Descending (LAD) and Left circumflex (LCX) artery with decreased blood flow. A diagnosis of Dilated Cardiomyopathy with congestive heart failure was kept and patient was treated with diuretics, digoxin, enalapril and carvidilol. Patient improved on above treatment.

Additional tests like complete blood count (CBC), kidney function test (KFT), liver function test (LFT), CPK, Cardiac troponin T, S. lactate, S. ammonia, S. Calcium, Vit D, Thyroid function tests, Viral markers for myocarditis were negative. TMS/GCMS and urine for organic acids were normal.

Cardiac CT was done to evaluate for coronary artery anomaly but report was inconclusive. In view of strong suspicion, coronary angiography was done which showed right coronary artery arising from aorta. Direct communication of Left Main Coronary Artery was not visualized with aorta. Multiple collaterals were seen between Right Coronary Artery and branches of Left Main Coronary Artery which were filling the Left Anterior Descending (LAD) and Left circumflex (LCX) artery in retrograde fashion. **(Image 3, 4).**

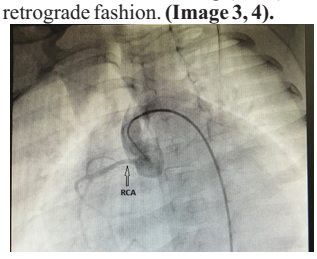

Image 3: Non selective angiography showing origin of RCA (right coronary artery) from aortic sinus. Left coronary artery was not seen.

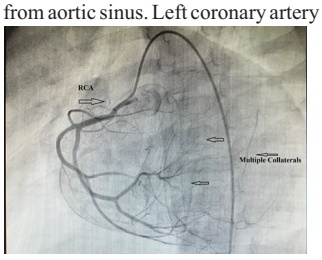

Image 4: Right coronary artery selective angiography showed multiple collaterals arising from RCA.

Discussion: Coronary Ostial Stenosis or Atresia (COSA) is spectrum of rare development conditions with an incidence between 0.01-0.04% of "sole coronary" cases. COSA affects the left coronary artery (L-COSA) more frequently than it does the right coronary artery (R-COSA).[3]

Embryologically, the coronary ostial buds develop soon after truncal septation. The defects that lead to coronary ostial atresia are likely to be acquired during later stages of fetal life. As coronary circulation begins to flow, the coronaries become the predominant source of blood supply for development of myocardial tissue. Therefore, the distribution and size of the major coronary arteries become related to the extent of development of myocardium. A lack of coronary circulation during this stage of development would induce hypoplasia of the dependent myocardium. Infrequently, the left coronary ostia, although possibly normal during early embryologic development, may become occluded in later fetal development, which leaves distal coronary bed intact. In these cases, in order for normal myocardial development to persist, an intact alternative blood supply must exist. In LMCAA, the distal coronary vascular bed is entirely supplied by the evolution of collateral circulation from the opposite side. Stage and manner of presentation is dependent on the adequacy of the collaterals.[4]

In congenital Atresia of LMCA, a single right coronary artery supplies the entire heart with flow in LAD & left circumflex artery in retrograde manner via collaterals from right coronary artery. Ostium of Left Main Coronary Artery is absent and proximal left main trunk ends blindly. Left Anterior Descending (LAD) & Left Circumflex arteries (LCx) are located in normal anatomical position and connect in usual fashion.[5] These findings were also seen in our case.

Depending on adequacy of collaterals, coronary artery atresia has different age of presentations. Literature reviewed by A Musiani et al [6] in 26 patients of LMCAA concluded that syncope, failure to thrive and myocardial ischemia were commonest presenting symptoms in pediatric age group. Adults are symptomatic in later life with angina pectoris. Our patient presented at 3 years of age with heart failure, without any prior symptoms.

If cardiac dysfunction is caused by congenital coronary anomaly, its recognition and prompt surgical treatment radically modifies the prognosis. Early surgical revascularization is the best choice. CABG (Coronary Artery Bypass Graft) using the internal mammary artery is procedure of choice in adults. In his study of A Musiani et al, 11 patients underwent surgical revascularization; CABG was done in 7 and in 3 cases coronary angioplasty was attempted. Although result of surgical angioplasty was not satisfactory, all patients of CABG were reported to be doing well after surgery. However, long term follow up is missing in these patients.

However, Bonnet et al [7] reported that surgical angioplasty in children is more suitable technique for coronary revascularization.

To conclude, congenital atresia/hypoplasia of the left coronary artery is a very rare disease. It should be considered when diagnosis of dilated cardiomyopathy is made. Prompt surgical treatment with coronary revascularization radically modifies the prognosis in these patients.

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