Coronary Artery Fistula

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Abstract

Coronary Artery Fistula (CAF) is the most common coronary arterial malformation but it is a rare cardiac anomaly. Incidence of CAF is 0.002% in general population and 0.4% in all cardiac malformations. Most patients remain asymptomatic. Only few cases of left coronary artery fistulas in infants have been reported worldwide. Here we present one case of left coronary artery fistula draining to right atrium and another case of right coronary artery fistula draining to right ventricle with sub-pericardial aneurysm giving origin to left circumflex as well as anterior descending coronary artery. Main left coronary artery absent, because of their rarity. Early diagnosis is the key for successful management and prevention of complications. Almost all cases can be correctly diagnosed by echocardiography and color Doppler. Currently options available for treatment are conservative management, surgery and trans-catheter closure.

Introduction

Coronary Artery Fistula (CAF) is the most common coronary arterial malformation but it is a rare cardiac anomaly. It was firstly reported by Krause in 1865 and the first surgical correction was done by Crafoord in 1947 [1,2]. The reported incidence of CAF is 0.002% in general population and 0.4% in all cardiac malformations [3]. Approximately half of the CAF patients remain asymptomatic and some CAF might disappear spontaneously during childhood. CAF may be congenital or acquired. Some are associated with other cardiac malformations (20 to 45%) and some are isolated (55% to 80%) [4-6]. Origin of the CAF can be from any of the three major coronary arteries, including the left main trunk. The majority of these fistulas arise from the right coronary arteries or the left anterior descending & the circumflex coronary artery is rarely involved. Single origin is the most common form of CAF, ranging from 74% to 90% of the cases [5-7]. The right coronary artery or its branches is the most common site of the CAF with 55% and the second common site is the left coronary artery in about 35% of the cases [6]. The Complications secondary to CAF increases with age. The complication like aneurysmal dilatation of the coronary artery, Congestive Heart Failure (CHF), and infective endocarditis is observed in 19% to 26%, 6%, and 3% respectively [8]. Here we present two patients of coronary artery fistula draining into cardiac chamber because of it rarity.

Case Series

Case 1

A 10 months old male child born as first issue of non-consanguineous marriage admitted in pediatric casualty of MDM Hospital, Jodhpur with complaints of respiratory distress off and on since the age of 2 months for which he was admitted three times. Patient was apparently asymptomatic till the age of 2 months after which mother complaint of excessive cry and difficulty in breathing and feeding, for which baby was taken to nearby hospital where he was admitted for 15 days. Similar episode occurred at the age of 4 months for which baby again admitted for 4 days and discharged on some oral medication. At the age of 7 months baby again admitted to medical college hospital with complaints of fever, cough and respiratory distress for 10 days and informed about congenital heart disease. There they were advice to go tertiary care centre for further management. But due to some family issues parents did not go. Then child was admitted to our hospital with similar complaints. Development is normal as per age and baby is fully immunized. Anthropometric measurements are as follows weight 5.8 kg & length 64 cm, head circumference 43 cm & MUAC 10.5 cm which suggest failure to thrive. There is no significant family history present. On physical examination, his respiratory rate was 64 per min, pulse rate 140 per min, BP 92/50, temp 98.2 F and SPO2 98%. The child had Pallor but no clubbing, no cyanosis, no edema, Hepatomegaly with liver span 8 cm, Chest in shape with no precordial bulge, apical impulse in left 4th ICS with no thrill or para-sternal heave. On auscultation continuous murmur was present at aortic area. Hemogram shows Hb 8.9
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2 gm/dl, TLC 18660/Cumm and Platelets 66000/Cumm. Chest X-ray shows cardiomegaly. ECG revealed Cardiac axis +90 with Right ventricular hypertrophy. Para-sternal short axis view and apical four chamber view on echocardiography revealed dilated left coronary artery at origin and apical four chamber view demonstrated enlarge right atrium and ventricle with tubular channel draining to right atrium at fossa ovalis (Figure 1). Doppler study revealed continuous flow from tubular channel to right atrium at fossa ovalis with trivial tricuspid and pulmonary regurgitation. CT angio chest revealed LCA is originating from left posterior coronary sinus (Figure 2). Left coronary artery (8 mm maximum diameter) dilated with a solitary retro-aortic fistulous tract (5 mm to 6 mm maximum diameter) communicating between left coronary artery trunk and right atrium suggestive of coronary cameral fistula. Left anterior descending artery was normally opacified and left circumflex artery was faintly opacified. After admission baby kept on medical oxygen by nasal prongs and oral furosemide along with breast feeding. After five days baby improved and gradually oxygen weaned off. Baby discharged on day 8th day on oral furosemide and Aldactone.

Case 2

The 10 years old asymptomatic female child born as first issue of non-consanguineous marriage referred with suspected very small Ventricular septal defect (VSD) X-ray chest and ECG were normal (Figure 3). Echocardiography revealed dilated right coronary artery 5 m to 6 m and left coronary artery absent. CT angio revealed (Figure 4) anomalous Left Coronary Artery LCA (white short arrow) arising from right coronary artery coursing anterior to right ventricle (white large arrow) with focal aneurysmal dilatation (black asterix) before giving origin to left circumflex (LCx) (black large arrow) and Left Anterior Descending Artery (LAD). That aneurysmal pouch is continuing with another intra-myocardial pouch (white asterix) before draining into right ventricle through small channel (black small arrow).
Discussion

The coronary artery fistula may involve main vessel, but the right coronary fistula is the more commonly implicated Azcuna et al. [9] the fistula terminates most frequently in a chamber or vessel of the right heart. The order of frequency being right ventricle, right atrium (including coronary sinus), pulmonary artery, and superior vena cava but drainage into the left ventricle is exceptional [10-13]. Although most fistulas occur singly, multiple fistulas are also described McNamara and Gross et al. [11]. Sabbagh et al. [14] considered that coronary artery fistula is in most instances an isolated anomaly, and other authors, notably Ogden and Stansel et al. [11] and McNamara and Gross et al. [12], have estimated that additional cardiac abnormalities are present in only 20% of patients. The clinical findings have been well documented [9]. In children it is exceptional for any symptoms to be present, although these may arise in later life in untreated cases when the shunt volume is large. Cardiomegaly may be present. The most frequent abnormal clinical sign (and for diagnosis the most useful) is a continuous murmur closely simulating that of a patent ductus arteriosus, except that it is best heard in the third or fourth interspace at the left sternal border rather than at the pulmonary area. Inappropriate surgery for a mistaken diagnosis of patent ductus arteriosus has occurred in the absence of full investigation [15]. Other conditions which require differentiation are aortopulmonary fenestration, persistent truncus arteriosus, ruptured aortic sinus aneurysm, and ventricular septal defect with aortic regurgitation [10]. Early diagnosis is the key for successful management and most relevant prognostic indicator [8]. Almost all cases can be correctly diagnosed by 2D ECHO & Doppler. Currently options available for treatment are conservative management, surgery and transcatheter closure. The management of CAF is still a controversial issue especially in small asymptomatic patients. To our knowledge only few cases of left coronary artery fistulas in infants have been reported worldwide. In 2006 Said et al. [16] studied the pediatric cohort (neonates, infants, and children) that was considered for investigation include in 62 males and 50 females and 17 whose gender was not specified. The mean age was 0.5 years (range 0.01 to 17 years). Reason for evaluation was mainly analysis of a cardiac murmur. The reported etiology of the CAFs was congenital in 127 (98%) and acquired in only 2 of the patients. In 2009 Jeewa et al. [17] reported a case of 1 year old clinically asymptomatic boy referred for a persistent murmur found at the clinical examination. An angiography showed the dilated left main coronary artery with a normal left coronary sinus of Valsalva. This dilated vessel followed along the atrioventricular groove, tapering into a 2 mm channel entering the right atrium at the mouth of the coronary sinus. A second smaller fistula was noted from the conus branch of the right coronary artery to the main pulmonary artery. Jeewa et al. [17] have already reported the case of CAF with CS stenosis in infant, but Hayabuchi et al. [18] revealed that Jeewa’s report may not be exactly correct. The LCX may drain into an abnormal vessel at first and finally drained into CS, which is not the same description as Jeewa’s. CAF may result in increase in right heart pressure, while CS stenosis could delay this process and reduce the incidence of CHF [8]. In 2017 karazisi et al. [19] described that most CAFs are asymptomatic, follow-up without intervention can be considered. To the best of our knowledge, most recommendations are empiric. Antiplatelet therapy is recommended to prevent thrombosis, especially in coronary artery dilatation. In some patients with greatly dilated coronary arteries, warfarin is recommended. Anticoagulation should also be considered postoperatively for patients who are treated surgically or with transcatheter closure, until the operative surface has healed and become sufficiently endothelialized. Warfarin should be considered for patients with greatly dilated coronary arteries in the long term. Most authors also recommend endocarditis prophylaxis. The 2008 AHA/American College of Cardiology Guideline recommends that small, asymptomatic CAFs, should not undergo closure, be followed up with echocardiography every 3 to 5 years [20]. This is recommended to exclude development of symptoms or arrhythmias, or progression of size or chamber enlargement that might alter management (class Ila, level of evidence: C). Follow-up is also essential after closure for early diagnosis of recurrence of the fistula and because of the possibility of persistent dilatation of the coronary artery, thrombus formation, calcification, arrhythmias, and myocardial infarction.

References


