Short Communication

Coronary Artery Origin Anomalies with Especial Emphasis on Delayed Diagnosis of Anomalous Origin of the Left Coronary Artery from Pulmonary Artery, A report from South of Iran

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Background: The aim of this study was to determine the clinical course and outcome of coronary artery anomalies including anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) diagnosed by coronary artery angiography (CAA) at our institution. The term coronary artery anomaly refers to a wide range of congenital abnormalities, involving the origin, course and the structure of epicardial coronary arteries. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an extremely rare, but potentially fatal congenital coronary anomaly.
Patients and Methods: The review included 21280 patients undergoing CAA at Medical Centers of Shiraz University between years 1997 -2006. The hospital records of all patients with diagnosis of ALCAPA reviewed for analysis. Data were expressed as percentages.
Results: There were 149 (0.7 %) cases of coronary anomalies, including 9 (6.04%) proven cases of ALCAPA. In ALCAPA group ,five patients presented with congestive heart failure (CHF) and a non-specific systolic murmur on examination. Isolated mitral valve regurgitation (MR), cerebrovascular accident due to embolization of a mural thrombus, and chest pain were the main presentations in three patients. Two of the patients were under follow up with presumptive diagnosis of coronary artery fistula. The patients aged between 4 months and 35 years. Surgical correction done for all the patients with ALCAPA by direct reimplantation in 8 patients and Takeuchi technique in one. We had one early hospital mortality in our group.
Conclusion: ALCAPA should be considered in differential diagnosis of any patient presenting with dilated cardiomyopathy, CHF, MR or chest pain syndrome. Awareness of such pathology can prevent or decrease the morbidity and mortality of a potentially fatal congenital heart disease.

Keywords: Coronary Artery Anomalies, ALCAPA, Delayed Diagnosis, Reimplantaion

Introduction

Congenital anomalies of coronary arteries occur in 1 to 2 % of general population undergoing CAA.[1-4] A study based on autopsy population of congenital heart disease has revealed a frequency of 2.2% among 1200 specimens.[5] Most of these anomalies are asymptomatic and not causing myocardial ischemia or cardiac sudden death.[1] However certain anomalies are associated with myocardial ischemia or predispose patients to sudden death.[5,6] The clinical presentation of this group depends on underlying pathology and the degree of adaptation of cardiovascular system. Associated signs and symptoms include angina, heart failure, myocardial infarction, dilated cardiomyopathy (DCM), valve insufficiency ,or sudden death.[7,8]

Angelini P in 1988 proposed the nomenclature

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and definition of both normal variants and anomalous coronary arteries.[9] A consensus report by the Society of Thoracic Surgeons-Congenital Heart Surgery Database Committee uses the following nomenclature: 1) anomalous pulmonary origins of coronaries, 2) anomalous aortic origins of coronary artery, 3) congenital atresia of left main coronary artery, 4) coronary arteriovenous fistula, 5) coronary artery bridging, 6) coronary aneurysms and coronary stenosis.[10] Of these, the most common types of clinically significant anomalies are origins of coronary artery from pulmonary artery, anomalous aortic origin of coronaries and coronary artery fistulas.[1,10-12]

The ALCAPA anomaly, also called Bland-White- Garland syndrome, account for 0.26-0.5% of congenital heart diseases, and if untreated has a mortality rate of more than 90% in the first year of life.[13-15]

The aim of this study was to determine the clinical course and outcome of congenital coronary artery origin anomalies in our area, detected by CAA between 1997- 2006, with focus on patients with diagnosis ALCAPA during the same time interval at Medical Centers of Shiraz University, Shiraz, Iran.

Patients and Methods

A 10 years review of both the pediatric car diac angiography and adult coronary artery angiography database including 21280 cases were conducted to determine isolated coronary artery anomalies. Having obtained the Institutional review board approval, and expressing the data as percentages the hospital records and operation notes of all the patients with diagnosis of ALCAPA were included for analysis.

Results

Coronary artery anomalies were found in 149(0.7%) of the patients, including 9 (6.04%) cases of ALCAPA. None of the patients with ALCAPA had associated congenital heart disease (Table 1).

Most of the patients with ALCAPA presented with signs and symptoms of CHF, DCM and a non- specific systolic cardiac murmur on physical examination. One of the patients presented with isolated murmur of mitral valve regurgitation with no signs of heart failure, and a 35 years-old adult female patient presented with chest pain. Also two patients were under follow-up with diagnosis of coronary artery fistula. The patients aged between 4 months and 35 years, including five females and four males. The clinical characteristics and surgical techniques used for repair of patients with definite diagnosis of ALCAPA are described in Table 2.

In most of the patients electrocardiography showed signs of ischemic myocardial lesion. Two-dimensional echocardiography and color Doppler studies revealed DCM with mild to moderate mitral regurgitation and abnormal flow in ventricular septum with dilation of RCA (Fig. 1). In two patients abnormal diastolic flow were detected in pulmonary artery, while in two patients origin of LCA was reported to be normal

Table 1. Type of congenital coronary artery anomalies among 149 patients from Shiraz, Iran

Type of anomaly	Percent
LCX and LAD with separate orifice	34.22
LCX from RCA	27.1
RCA from left aorta sinus	19.46
LCA from right aorta sinus	10.04
Single coronary orifice	6.04
LCA from PA (ALCAPA)	6.04

LCX: Circumflex Artery , LAD: Left Anterior descending artery
RCA: Right Coronary Artery , PA: Pulmonary Artery

Table 2. Clinical characteristic and surgical techniques for repair of patients with ALCAAPA

No patient	Age at presentation (years)/ gender	Clinical presentation	Presumptive diagnosis By ECG + echo	Age at operation / year of operation	Surgical technique
1	6/ male	CVA + seizure	LV aneurysm suspicious to ALCAPA by echo	6 y / 1997	Reimplantation aneurysmectomy
2	0.83/ male	Symptoms of CHF	DCM	8 y / 1999	Reimplantation
3	0.33/male	Symptoms of CHF	DCM + MR	9 y / 1999	Reimplantation aneurysmectomy
4	8/ male	Cardiac murmur	MR, coronary fistula	14 y / 2003	Reimplantation
5	35/female	Chest pain	Coronary artery disease	35 y / 2004	Reimplantation
6	5 y/female	Cardiac murmur	Coronary artery fistula	5 y / 2006	Reimplantation
7	0.33/female	Symptoms of CHF	DCM suspicious to ALCAPA	2 y / 2006	Reimplantation
8	0.25/female	Symptoms of CHF	DCM + MR	7 y / 2007	Reimplantation
9	0.33/female	Symptoms of CHF	DCM suspicious to ALCAPA echo	0.25 y / 2007	Tunnelling (Takeuchi technique)

MR: mitral regurgitation, DCM: dilated cardiomyopathy , LV: left ventricle, CHF: congestive heart Failure, Echo: echocardiography
ECG: electrocardiography

on echocardiography. Because of diversity of clinical presentations and unawareness of physicians regarding unusual findings of ALCAPA on echocardiographic studies, the definitive diagnosis in most patients was missed, until confirmation was made by CAA (Fig. 2). Surgical correction was done by reimplantation of LCA to aorta for 8 patients with change of 1-coronary perfusion system to 2- coronary perfusion system. We had only one early mortality in a 4 months old baby girl on the third day of hospitalization for which Takeuchi technique was used for re-establishment of coronary perfusion. She was the youngest patient in this study who died due to ventricular fibrillation.

Discussion

In our review for anomalies of origin of coronary arteries among 21280 CAA, we found a total number of 149 (0.7%) patients with coronary anomalies including 9(6.0.4%) cases of ALCAPA with delayed diagnosis in 8 patients. The delay in diagnosis of ALCAPA reported by Frescurac et al. who diagnosed 5 cases (18.5%) of ALCAPA among 1200 autopsy specimens collected from 1968 to 1996.[5] In their study 4 out of 5 cases died in early infancy, either suddenly or due to congestive heart failure due to myocardial infarction.

Survival and symptomatology of the patients with ALCAPA depends on the extent of collateral circulation and, to a lesser degree on the pulmonary vascular resistance.[16-20] If an adequate collateral network is not well established early in infancy, with decreasing pulmonary resistance, the myocardium perfused by the anomalous artery will often become ischemic. Patchy areas of fibrosis develops

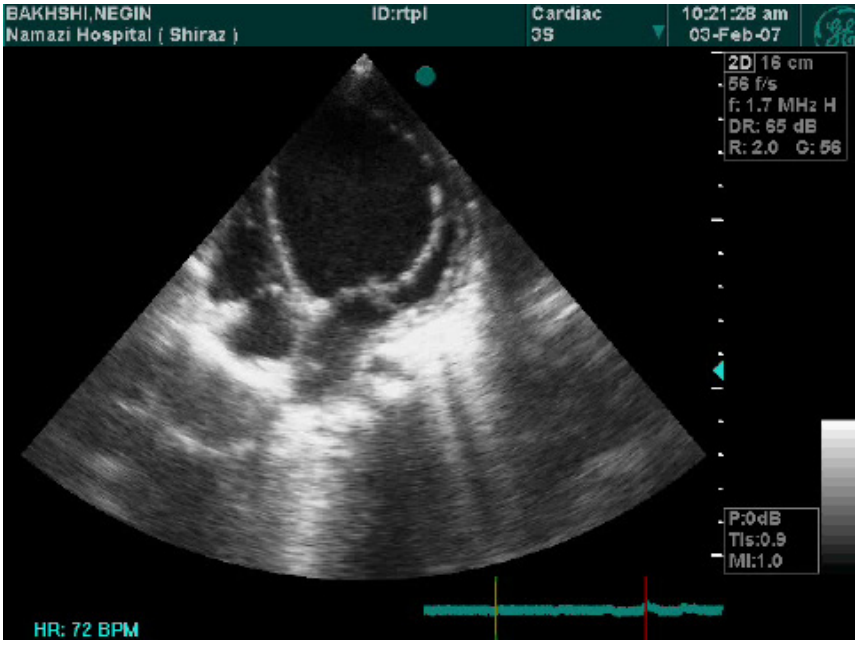

Figure 1. Two-dimentioanal echocardiography (4 chamber view) of patient No. 3, reveals severely dilated left ventricle, as sign of dilated cardiomyopathy in our patient with ALCAPA.

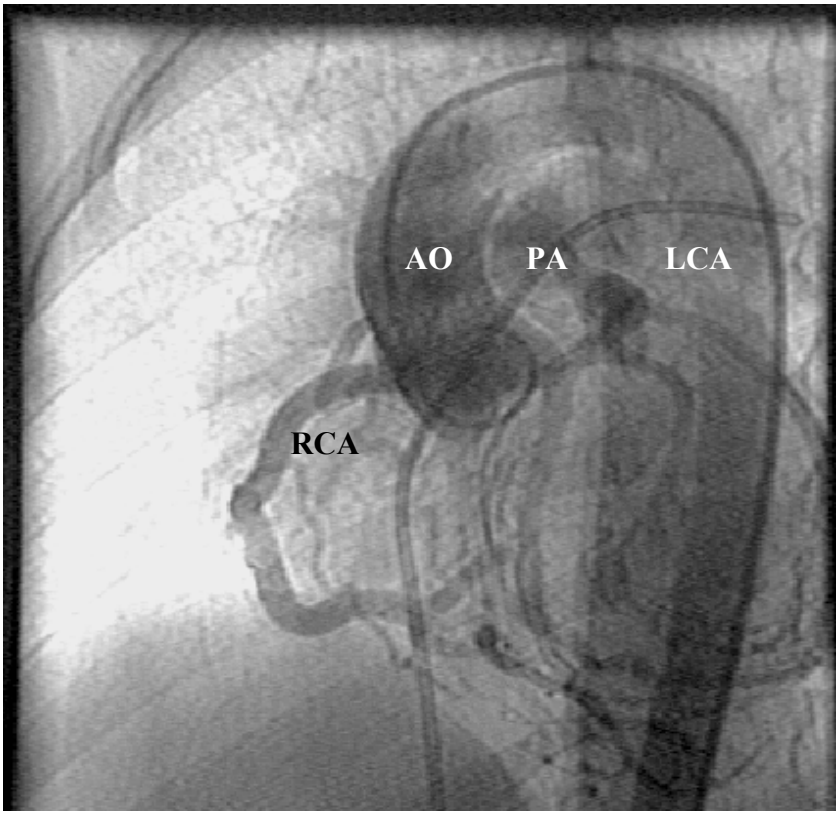

Figure 2. Aortogram (Anterior – Posterior) view of patient No 8, shows dilated RCA with collaterals to LCA and then opacification of main pulmonary artery, which confirms diagnosis of ALCAPA

resulting in ventricular dysfunction, a large aneurysm, and often mitral regurgitation. Most of the patients usually present with cardiogenic shock in early infancy and without surgical intervention, this situation is uniformaly fatal.[1] If collateral network is well developed, this results in a functional single coronary system, which helps some patients remain asymptomatic into adulthood, while some will present with signs of congestive failure, DCM or isolated mitral regurgitation. In our series, 4 out of 9 patients clinically presented with signs and symptoms of heart failure, and were under treatment for DCM. Only for a 4 months- old baby girl with heart failure, the diagnosis of ALCAPA was made by echocardiography on arrival to the hospital which was subsequently confirmed by CAA. Early diagnosis and reestablishment of a 2- coronary artery system will improve outcome and survival of these patients.[9,10] Surgical correction for ALCAPA is recommended whenever diagnosis is established, because on long term follow up of such patients with delayed presentation or missed diagnosis, improvement of myocardial function is confirmed.[20,21] In two of our patients with DCM, 2 normal coronary artery systems were reported by echocardiography and Doppler studies. This false appearance of coronary arteries has already been reported.[22] Two of our patients were under follow up with presumptive diagnosis of coronary artery fistula. This pitfall also has been reported by Azakie et al. and others.[22,23] We had one patient with presentation of isolated mitral regurgitation, and another with cerebrovascular accident, due to embolizaton of a mural thrombus from left ventricular aneurysm. In conclusion, our study demonstrated the unusual and late presentation of patients with ALCAPA, which due to unawareness of physicians, were missed . Therefore ALCAPA should be considered in differential diagnosis of any patient with DCM, CHF, isolated mitral regurgitation, chest pain syndrome or coronary artery fistula. While non-invasive diagnostic tools such as color Doppler echocardiography and multi detector computed tomomography (MDCT) can be conclusive, the final diagnosis should be confirmed by CAA if the results of such screening studies are not convincing.[23-25]

Acknowledgements

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