Case Report

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Anomalous Origin of Left Coronary Artery from Pulmonary Trunk

Monil P. Thakrar*, Sapna Gandhi, Alpana Jain***, Arpita Jindal******

Abstract

Anomalous origin of the left coronary artery from the pulmonary trunk also known as Garland-bland-White syndrome is a rare congenital anomaly. The usual clinical course is severe left sided heart failure and mitral valve insufficiency presenting during the first months of life. However, in some cases collateral blood supply from the right coronary artery is sufficient and symptoms may be subtle or even absent. Arrhythmias or sudden cardiac death in adult life may be the first clinical presentation in patients with this anomaly.

Keywords: Coronary Artery; Pulmonary Trunk; Congenital Anomaly.

Case Report

A 21 year old male was found unconscious at his place and was rushed to a nearby hospital where he was declared dead on arrival. A postmortem examination was requested to ascertain the cause of death.

Physical Examination

Subject was averagely built, well nourished male. Rigor mortis was present all over the body. Postmortem lividity was present on back and dependent parts of the body. Fingernails of both hands had bluish tinge.

Gross Examination of the Heart

The heart weighed 380 gms, measured 12x7x4 cms. On opening heart by inflow outflow method, the cut surface of the heart showed ventricular cavities dilated. Right ventricular wall thickness was 0.3 cms. Left ventricular wall thickness was 1.7 cms and its anterior wall was 0.3 cms thick suggesting dilated cardiomyopathy. Interventricular septum thickness was 2 cms. Right coronary artery was found to be dilated and tortuous like a vein from origin to insertion. Left coronary artery was found to be arising from the pulmonary trunk. Aortic stump was 1.3 cms in length and its inner surface was smooth. Left anterior descending artery was unremarkable. Tricuspid valve circumference was 12 cms and its external surface was unremarkable. Mitral valve circumference was 10 cms and its external surface was also unremarkable. Pulmonary valve circumference was 8 cms and its cusps were within normal limits. Aortic valve circumference was 7 cms and its cusps were within normal limits. See Figure 1, Figure 2.

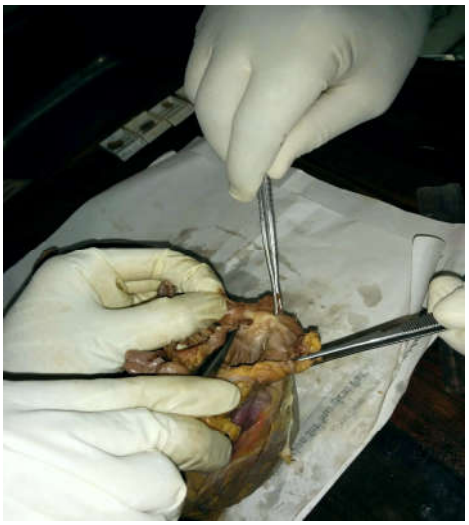

Fig. 1: Figure depicts anomalous origin of the left coronary artery from pulmonary trunk

Authors Affiliation: *2nd Year Resident, **Assistant Professor, ***Senior Demonstrator, ****Associate Professor, Department of Pathology, SMS Medical College, Jaipur.

Reprints Requests: Monil Thakrar, 102, Pearl Grands, A-57, Shantipath, Tilaknagar, Jaipur – 302004.
E-mail: monil.p.thakrar@gmail.com

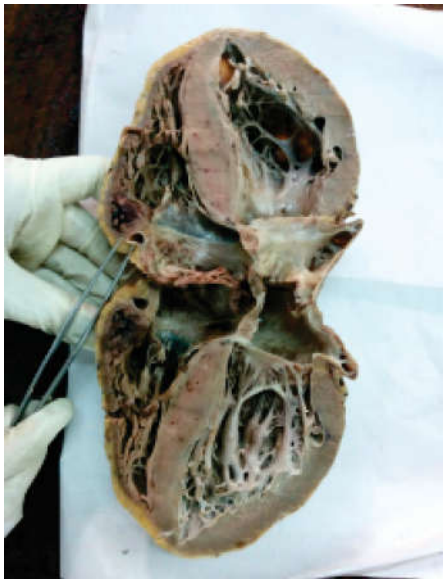

Fig. 2: Figure depicts cut surface of the heart with dilated left ventricle and thinned out papillary muscles

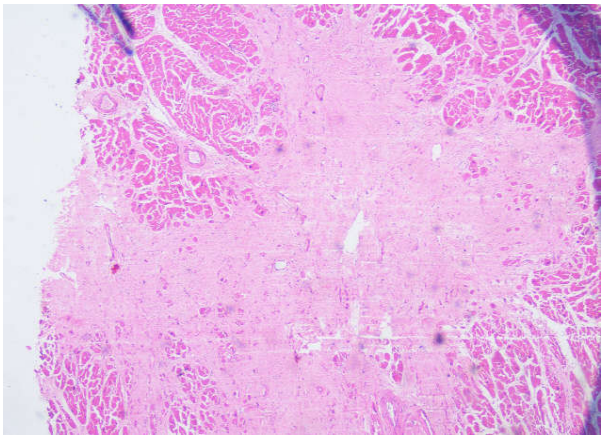

Fig. 3: Microscopic picture of left ventricular wall showing fibrosis of myocardial muscle fibres due to recent myocardial infarction

Microscopic Description of the Heart

- *Left ventricular walls* showed features of recent myocardial infarction. Anterior wall of the left ventricle showed old healed infarct undergone dystrophic calcification. See Figure 3.
- Right ventricular wall, both coronary arteries, aorta and left anterior descending artery were within normal limits.

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was first described by Brook in 1886, and, in 1933, Bland, White, and Garland reported the clinical syndrome of this disease for the first time [1,2,3,7,10]. Its occurrence is rare (present in one of 300,000 live births or about 0.26% in patients with congenital heart disease) [1,2,7,10]. Ever since its first introduction, it was generally an isolated genetic anomaly although there are reports of its association with other diseases including ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and AP window [2,5,6,10]. Anatomically, the whole left main coronary artery or only the left anterior descending or circumflex branch connects anomalously to the proximal pulmonary trunk or very rarely to the proximal right pulmonary artery. Very rarely, both coronary arteries connect to the pulmonary artery by a single trunk [3,7]. The anomalous main LCA connects most often to the sinus of valsava immediately above the left or posterior cusp of the pulmonary trunk and rarely above the right cusp [2,7]. Branching pattern of the anomalous left coronary artery remains normal. Because of its high mortality rate (up to 90%)[2], early diagnosis and prompt surgical interventions are necessary to provide gradual myocardial recovery and good clinical outcome, and, due to this reason, the majority of the cases were diagnosed before the patient reaches the age of one year. But even with clinical awareness and early intervention, 65% of the infants born with this anomaly die within the first year of life [7,10]. In the remaining infants who survived beyond the age of one year, the hazard lessens considerably, and the chronic phase ensued. Among the patients who are in the chronic phase and lived into the adulthood, as in the patient reported in this case, rich collateral from the right coronary artery, which arises normally from the aorta, feeds the left coronary artery and the flow is reversed, in which the left coronary artery drains into the pulmonary artery [1,2,4,7,10]. Many such patients are in good health, and few even have normal ECGs. Survival beyond the first year may be related to marked RCA dominance, supplying not only the diaphragmatic portion of LV but also much of the septum and lateral wall [7]. Patient with these arrangements may occasionally only have papillary muscle ischemia and fibrosis, and mitral regurgitation may dominate the clinical picture [7]. Some adults remain asymptomatic or complain only of fatigue, dyspnea, or palpitations. About half have effort angina. The resting ECG is always abnormal, with ST changes or evidence of old anterolateral infarction [3,7]. Exercise ECG usually shows an abnormal ischemic response, while the stress thallium myocardial imaging is usually abnormal [7]. CXR may be normal or may show cardiac enlargement. Cineangiography shows collaterals from the RCA and usually a near normal LV ejection fraction, but

with anterolateral hypokinesia [7,8,10]. However, patients who survived infancy continue to be at risk of death from chronic heart failure secondary to ischemic left ventricular cardiomyopathy, and diagnosis is an indication for operation even among the older patients [7]. In older patients, internal thoracic artery grafting with or without mitral valve repair is a reasonable alternative when size of the graft permits, and this case is achieved simply by the standard coronary arterial bypass grafting technique and ligation or obliteration of the anomalous left coronary artery [2,4,7,8,9]. The risk of premature death depends on the perioperative status of the left ventricle (especially the left ventricular myocardium) and mitral regurgitation [2,7]. The postoperative functional class depends primarily on preoperative LV status and it was generally good late postoperatively. The LV, size (including cardiothoracic ratio) is nearly always markedly reduced after operation [7]. Signs of myocardial ischemia are also reduced. However, the myocardial flow reserve is reduced, and exercise tolerance is lower than normal among the survivors [2,7,10]. In terms of mitral regurgitation, when operation is performed in infancy, even important mitral regurgitation can regress postoperatively. However, if the mitral regurgitation was severe before surgery, it would not regress, and reoperation would require a few months to a few years later [2,7].

Conclusion

Anomalous origin of the left coronary artery from the pulmonary trunk, also known as Garland-Bland-White syndrome, is an extremely rare but potentially fatal congenital cardiovascular anomaly. With its high mortality within the first year of life, even fewer infants who were born with this anomaly. Our case was thereby presented not only because of its rarity but also for the mild clinical onset the patient presented, and a simple operation utilizing the well-established and common cardiac procedures could have effectively corrected this highly fatal defect.

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