Anomalous Origin of Right Coronary Artery from Pulmonary Trunk (Arcapa Anomaly)

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Editorial

Knowledge of the normal and variant anatomy and anomalies of coronary circulation is a vital component in the management of coronary artery disease. Any coronary artery disorder may reduce the blood supply to the heart and may lead to myocardial infarction and death.

Coronary arteries represent the only supply to the myocardium. The two main coronary arteries, right and left coronary arteries arise from ascending aorta. Right coronary artery arises from right anterior aortic sinus and left coronary artery arises from left posterior aortic sinus. Patency of left coronary artery is vital for sufficient perfusion of the heart. The left coronary artery is responsible for irrigation, not only of most of the left ventricle, but also considerable portion of right ventricle [1].

Right coronary artery runs between pulmonary trunk and right auricle and then runs downwards and is divided into two segments. First segment runs along the right border of the heart up to inferior border of heart. It gives right conus artery and supplies infundibulum of the right ventricle. If the right conus artery arises separately from the aortic sinus, then it is called as Third coronary artery. The second segment runs up to the apex as right marginal artery and supplies right ventricle. Sometimes it may runs in the posterior interventricular groove and may give rise to Posterior interventricular branch. Hence according to the origin of posterior interventricular artery, the coronary artery dominance has been considered. Right coronary artery ends in the crux by anastomosing with circumflex artery. It supplies the right atrium, right ventricle, atrioventricular septum and part of left ventricle [1].

The incidence of congenital coronary artery anomalies is 5-6%. By definition, the term anomalous or abnormal is used to define any variant form observed in less than 1% of the general population [2].

Prevalence of Anomalous origin of right coronary artery from pulmonary trunk is 0.002%. Anomalous origin of left coronary artery from Pulmonary trunk is a common one [3].

Splanchnopleuric mesoderm contributes to all components of heart. The mesoderm contributes to the cardiac area that occurs during 3rd week of embryogenesis. The cardiac area subsequently forms a pair of endocardial tubes which fuses to form primitive heart tube. Normal coronary artery arises from appropriate differentiation of pleuripotent cells into their respective anatomic and functional components. Anomalies of the coronary circulation result from processes that disrupt the normal differentiation and specialization of heart tube [3]. In particular, abnormal involution, position of endothelial buds or septation of truncus arteriosus may give rise to anomalous origin of coronary artery [4].

Coronary endothelial sprouts occur at around 5th week of Intrauterine life from the bulbous cardia whose has not yet differentiated into the aorta and pulmonary trunk. The first evidence of coronary vessel development is the appearance of the blood islands at the beginning of the 5th week just under the epicardium in the sulci of the developing heart.

Anomalous origin of right coronary artery from pulmonary trunk (ALRAPA SYNDROME) is a rare congenital anomaly with an estimated prevalence of 0.002%. This is incidental anomaly. This was observed in three specimens in our study. This is in contrast with anomalous origin of left coronary artery from pulmonary trunk which is fatal during early infancy without immediate surgical management [5].

Anatomic variations of the heart vessels are common. Hence identification of normal coronary artery pattern and its branches and variations are important for cardiologists, cardiothoracic surgeons and radiologists while performing coronary angiography and surgical procedures.

Even though Anomalous origin of right coronary artery from pulmonary trunk (ALRAPA SYNDROME) is a rare, it should be known to the cardiologists, cardiothoracic surgeons and radiologists before performing intervention. This may lead to Cyanotic heart disease and even death in the newborn period. This syndrome produces less severe complication than the ALCAPA (Anomalous origin of left coronary artery from pulmonary trunk). Definitive therapy may be surgical translation of the anomalous Right coronary artery to the aortic root [6].

Coronary angiography is an imaging procedure which shows the identification of normal coronary arterial pattern and its variations. MDCT (Multi Detector Coronary Angiography) is the study to diagnose the ARCAPA and to plan for surgery.

Reference

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