Case Report

A case of Tetralogy of Fallot with left coronary artery to pulmonary artery fistula

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URL: http://keralaheartjournal.in/ojs/index.php/KHJ/article/view/58

Abstract:

The congenital coronary artery to pulmonary artery fistula is a rare anomaly. It is often reported in patients with ventricular septal defect and pulmonary atresia. Herein we report a case of left anterior descending coronary artery to pulmonary artery fistula in a patient of tetralogy of Fallot.

Key words: Tetralogy of Fallot, fistula, coronary artery, pulmonary artery, pulmonary atresia

Case Report:

An 8-year old girl presented with cyanosis since 1 year of age and dyspnea on exertion NYHA functional class II. She had history of cyanotic spells during infancy. On examination her pulse rate was 86 per minute with blood pressure of 100/70 mmHg. There was cyanosis, pan digital clubbing and polycythemia (hematocrit of 68%). Cardiovascular system examination revealed normal first heart sound, single second sound and continuous murmur best heard in left 3rd intercostal space in the parasternal area. Chest X-ray showed decreased pulmonary blood flow with right ventricular type of apex, concave pulmonary bay and a prominent aortic knuckle. Electrocardiogram revealed sinus rhythm with features of right ventricular hypertrophy and right axis deviation. The transthoracic echocardiography showed large sub aortic ventricular septal defect with 60 % aortic override, right ventricular hypertrophy with severe infundibular and pulmonary valve stenosis. Cardiac catheterization and angiography study confirmed the diagnosis of tetralogy of Fallot with large ventricular septal defect, right ventricular hypertrophy and aortic override. There was only faint filling of pulmonary artery on right ventricular injection (Figure 1A). The aortic root injection showed presence of large communication from left coronary system to pulmonary trunk. Selective left coronary injection confirmed presence of fistulous communication arising from left anterior descending coronary artery to the main pulmonary artery (Figure 1B). Patient underwent intra-cardiac repair with ventricular septal defect patch closure, infundibular resection, pulmonary valvotomy and right ventricle outflow tract augmentation with patch. Fistulous communication between left coronary artery and pulmonary artery was ligated. Post-operative period was uneventful and discharged on day 7.
Discussion:

The congenital coronary artery to pulmonary artery fistula is a rare anomaly, reported to occur commonly in patients with ventricular septal defect and pulmonary atresia with an incidence of 10%. Failure of obliteration of the intramyocardial trabecular sinusoids with anomalous development of the intra-trabecular spaces, through which blood is supplied to the myocardium during intrauterine life results in congenital coronary to pulmonary artery fistula. Reports of large coronary artery to pulmonary artery communication are very rare in tetralogy of Fallot. Physiologically coronary artery to pulmonary communications in tetralogy of Fallot helps in increasing pulmonary blood flow and reduction in degree of cyanosis. The essential goals of surgical correction are to close the fistulous communication, to ensure unhindered coronary arterial flow, to close the ventricular septal defect with relief of right ventricular outflow obstruction, and to establish unobstructed continuity between the right ventricle and the intra-pericardial pulmonary arteries.

Conclusion:

The surgical importance of coronary artery to pulmonary artery fistula lies in the fact that the communication needs to be identified and ligated before commencing cardiopulmonary bypass to prevent loss of volume to pulmonary vascular bed. Failure to close such communications prior to application of aortic cross clamp results in loss of cardioplegic solution to pulmonary circulation and may result in incomplete myocardial protection during administration of cardioplegia.

References: