CONGENITAL ABSENCE OF LEFT PERICARDIUM AND MEDIASTINAL PLEURA: A RARE INCIDENTAL FINDING DURING OPEN HEART SURGERY.

Cardiology

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ABSTRACT

9 Year old male child planned for elective Atrial Septal Defect closure Surgery. He had a incidental Intra Operative finding of Partial pericardial defect on left side with Pericardial Herniation in left pleural cavity.

KEYWORDS

Congenital Pericardial defect, pericardial herniation, atrial septal defects.

Introduction- Congenital Pericardial defects are a rare finding and is usually diagnosed incidentally at the time of cardiac surgery. Patient is usually asymptomatic but rarely can have sudden cardiac death due to pericardial herniation. We report incidentally detected congenital pericardial defect in a case of 9 year old male child with Ostium Secundum ASD with a size of 25mm and left to right Shunt. Patient was referred to higher centre and was further evaluated by the Cardiologist. Echocardiography detected only dilated RA/RV with 27mm Ostium Secundum ASD L→R >2:1 shunt, with normal pulmonary artery anatomy and biventricular function.

Case History: A 9-year-old boy had a history of high grade fever with dry cough for which he presented to a local paediatrician and was diagnosed as having upper respiratory tract infection. On Auscultation patient had wide and fixed split second heart sound for that he was advised echocardiography and was diagnosed as Ostium Secundum ASD with a size of 25mm and left to right Shunt. Patient was referred to higher centre and was further evaluated by the Cardiologist. Echocardiography detected only dilated RA/RV with 27mm Ostium Secundum ASD L→R >2:1 shunt, with normal pulmonary artery anatomy and biventricular function.

X-Ray chest PA view revealed Enlarged RA and Pulmonary Plethora.

ECG showed sinus rhythm of 84/min. Axis qrs +135 and P +115, P wave is Negative in 3, aVF, QRS Shows RBBB with T Inversion in V1 TO V6.

Pre-op chest X-ray PA and Lat.

Intra op Findings- After median Sternotomy pericardium was separated and was found to have a defect between Right Atrium and Right Ventricle through which rest of the heart chambers were herniating into the left pleural cavity with slight shift of the SVC and IVC also towards the left side.

Intraoperative picture showing pericardial defect between Right Atrium and Right Ventricle.

Post op CT Non-coronary cardiac doesn't reveals any pericardial defects and shows shifting of heart towards left pleural cavity.

DISCUSSION:

Congenital absence of total or partial pericardium are very rare findings and mostly the patient is asymptomatic. Left sided pericardial defects are more than the right sided ones with prevalence rate of more than 70%. Complete absence of the pericardium (9%) and absence of the right pericardium (17%) are fewer than the absence of the left pericardium. Diagnosis for the criteria are simple, ensuing from the geometric consequences of the absent left pericardium: shift the heart out of the mediastinum into the left hemi thorax, heart hyper mobility, and modified heart silhouette due to the interposition of lung tissue.

Congenital defects can be further characterized by location and whether there is partial or complete absence of the pericardium. Patients with partial left sided pericardial defect, however, are at risk of possibly symptomatic herniation of the Atrial Appendage or herniation of the left ventricle, eventually leading to fatal myocardial strangulation.

Apical defects need to be detected as soon as possible because the time from the onset of symptoms of apical herniation and strangulation of the left ventricle to death varies from one hour to two days. In our case Echocardiography and CT scan could not be able to find the defect and found it incidentally Intra Operatively.

In our case there is partial left side pericardial defect along with left pleural defect through which right and left ventricle along with left atrium herniated to left side of pleural cavity. After Correction of Atrial Septal Defect, pericardial defect is Removed and pleura also enlarged to Reduced the chances of Future Pericardial Herniation.

Other congenital anomalies of pericardium described by Ellis et al: total absence, right sided defects (complete or partial) and diaphragmatic defects. Saint Pierre and Froment reported total absence of pericardium in 9% of cases which coexists with incomplete closure of chest wall and anomalies of transverse septum. In rarer form there is ectopia of heart, either extra thoracic (ectopia cordis) or inside and omphalocele (anterior celosomia). In less severe forms the heart remain Intra thoracic and associated diaphragmatic defects of Intrathoracic Hamartoma.

Embryology

Developmental embryology – Development of pericardium started at

3rd week of gestation and after division of embryonic coelom. Septum transversum formation leads to separation of peritoneal cavity with pericardial cavity⁹. These pericardial cavity grows on left and right side to form right and left pleura pericardial folds. These folds join together to form incoming cardinal vein of sinus venosus, fold on ventral side forms the pleuropericardial membrane, while the dorsal forms the pleuroperitoneal membrane¹⁰. Defects in pericardium occurs either due to incomplete formation of septum transversum or pericardial folds. Premature atrophy of common pericardial vein leads to compromised blood supply in pericardial folds and is the leading cause of congenital pericardial defects.

CONCLUSIONS-

1. Pericardial defects can be missed in Imaging and Echocardiography and surgeon should be more rely on Intra operative findings.
2. The partial displacement of the heart into the left hemithorax in supine position of the patient is pathognomonic for partial left sided pericardial defect.
3. Contrary to previous reports the noticeable prominence of the left atrial appendage cannot be taken as a reliable sign of a partial left sided pericardial defect. It can be seen in both complete and partial left sided pericardial defects.
4. The bulging of the left atrial appendage with absence of gross cardiac displacement is indicative of a partial left sided pericardial defect. No fatal outcome has been reported in these defects, in which strangulation of the left atrial appendage can occur.

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