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Case Report

Complete Left Pericardial Agenesis associated with Ventricular Septal Defect

Case Report

9 years old male patient admitted to our department with complaints of recurrent respiratory tract infection since 2 years of age. No History of cyanosis His left eye is absent and it was the abnormality at birth.

On physical examination, there was no cyanosis or clubbing. Left eye Enophthalmous was present. Pulse rate was 88 per minute and regular. Blood pressure on Right upper limb in sitting position was 100/60 mm Hg. Apical impulse was displaced and prominent down and out with prominent systolic thrill. On Auscultation, first and second heart sounds were normal and 4/6 pan systolic murmur was heard on the left lower border of the sternum.

Electrocardiography showed features of Normal sinus rhythm, Left axis deviation, Left ventricular hypertrophy.

Chest X-Ray Postero anterior View showed the following features (4)

- Cardiomegaly with Cardiothoracic ratio of 0.6.
- Prominent pulmonary bay.
- Lung shadow between Pulmonary artery and Aorta.
- Straight Left heart border (Figure 1).

2D Echocardiography revealed Perimembranous VSD with intact Inter Atrial septum, pulmonary stenosis with gradient of 50 mm Hg across the pulmonary valve and Good biventricular function.

Intra operatively, both heart and left lung were seen after opening the anterior pericardium. There was no separate pericardial cavity with absent posterior pericardium and it was continuous with left pleural cavity – Complete left pleruo-pericardial agenesis (Figure 2).

There was a Restrictive Perimembranous VSD of 5 mm diameter partially closed by the septal leaflet of Tricuspid valve. Pulmonary valve was dysplastic with valvular pulmonary stenosis.

Transatrial 0.6 mm Polytertafluroethylene patch closure of VSD and open pulmonary valvotomy was done. No surgical correction done for left pleuropericardial agenesis as there was no major herniation of the heart in to the left pleural cavity. Chest tubes were placed in the mediastinum and pericardium. Post-operative course was uneventful and patient discharged on 6th post-operative day with stable hemodynamics. There were no symptoms post operatively at 6 month follow up of the patient. 2D ECHO at the time of review showed intact IVS with no gradient across the pulmonary valve.

Discussion

A congenital pericardial defect is a rare clinical condition, involving the left (86%) more than the right [1-3]. Males are more commonly affected than females (3:1). In most of the

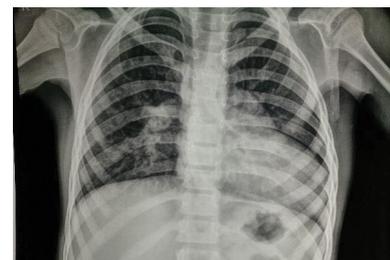


Figure 1: Preoperative Chest Roentgenogram showing features of CPD.

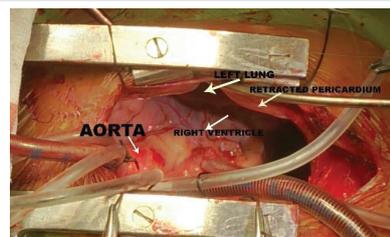


Figure 2: Intra operative picture showing Heart and Left Lung after opening pericardium.

cases, the anomaly is asymptomatic. The reported incidence of isolated congenital pericardial defects is 1/14000 [1].

The main cause is due to the premature atrophy of the left duct of Curvier, which leads to the persistence of the left pleuropericardial foramen [2].

Chest pain is the commonest symptom, which is typically precipitated by left lateral decubitus position and relieved by turning to the right [4]. Sudden death is reported [1]. Chest pain symptoms are often attributed to coronary ischemia from the torsion of great vessels, herniation of cardiac structures through the pericardial defect and tension of pleuropericardial adhesions [4]. The reasons for sudden death may be the herniation of the left ventricle, left appendage and the involvement of left circumflex artery [1].

On chest X-Ray film, the following features can be seen

- 1) Shifting of heart to the left and hence irregular left side border of the heart and loss of right side border of the heart due to its superimposition over the spine.
- 2) Air shadow between the aorta and pulmonary bay due to the presence of lung tissue.

On echocardiography, right ventricular dilatation and paradoxical septal motion are commonly seen, but ventricular function is usually normal [5].

At cardiac catheterization, one can detect protruding of left atrial appendage through the defect [4,6]. Magnetic resonance imaging is the most useful diagnostic test. It determines the extent of the defect, excludes herniation of cardiac structures and any additional defects [7,8]. Video assisted thoracoscopy can also be helpful.

If the patient is symptomatic or the defect is associated with other cardiac defects, surgery might be indicated. Because of the rarity of congenital pericardial defects and variability

of their presentation, no standard surgical approach has been recommended. Surgical closure techniques consist of primary closure, patch closure, widening of the defect depending on the anatomic size and location of the defect. If there is acute apical appendage strangulation, left atrial appendectomy is also might be considered [4]. Larger defects are typically well tolerated and repair can be technically difficult. Reconstruction is not recommended for such large defect like in our case because heart typically adapts to the distorted anatomy and corrective attempts may result in unstable flow patterns.

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