Lung herniation into pericardial cavity: A case of partial congenital absence of right pericardium

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ABSTRACT

Congenital absence of pericardium is rarely seen, often diagnosed intraoperatively during cardiac and thoracic surgeries. Left-sided pericardial defects are more common than right-sided ones. We present a case of an incidentally detected congenital absence of right pericardium with herniation of part of the right lung during ventricular septal defect closure surgery in a male child aged 4 years.

Keywords: Congenital absence of pericardium, lung herniation, partial absence, right pericardium

A male child aged 4 years presented to us with history of repeated respiratory tract infections. On clinical examination, the child was acyanotic with normal physical appearances. Holosystolic murmur of grade 4/6 was appreciated at fourth intercoastal space along the left sternal border. Echocardiogram revealed a diagnosis of non-restrictive perimembranous ventricular septal defect of moderate size having left to right shunt with biventricular hypertrophy. Chest X-ray postero–anterior view showed increased cardiothoracic ratio of 0.6 with increased vascularity of lung, and was remarkable for a radiolucent shadow in the superior mediastinum [Figure 1]. The electrocardiogram showed normal sinus rhythm with subtle and non-specific findings.

The child was taken up for elective closure of the venricular septal defect. A standard median sternotomy incision was performed and midline pericardiotomy was done. To our surprise, lung tissue was noticed inside the pericardium [Figure 2]. Following complete pericardiotomy, it was noticed that part of the upper lobe of the right lung was herniating into the pericardial cavity through a right-sided pericardial defect posterior to the superior vena cava [Figure 3]. The herniating part of the right lung was not compressing on the superior vena cava or right atrium. Routine ventricular septal defect closure was performed and no intervention was carried out for the pericardial defect. The outcome of the surgery and post-operative well being of the child was not affected by the right-sided partial pericardial defect. The child was asymptomatic at 6 months, although a follow-up of longer duration is required to comment on the benefits of the non-interventional measure taken up for partial absence of right pericardium.

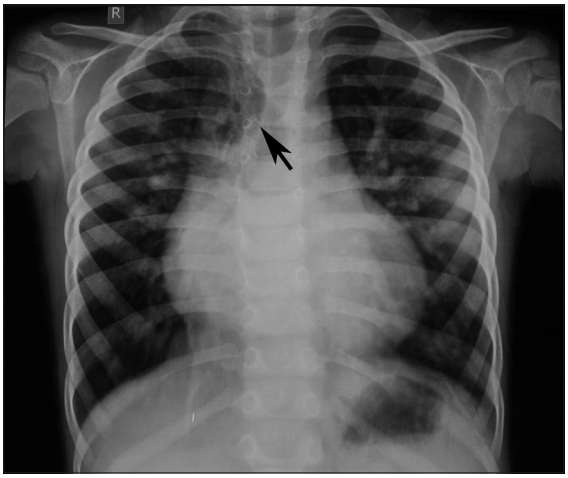

Figure 1: Chest X-ray showing radiolucent shadow (arrow) in the superior mediastinum

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DISCUSSION

Congenital pericardial deficiency was first described by Realdus Columbus in 1559.[1] Congenital absence of

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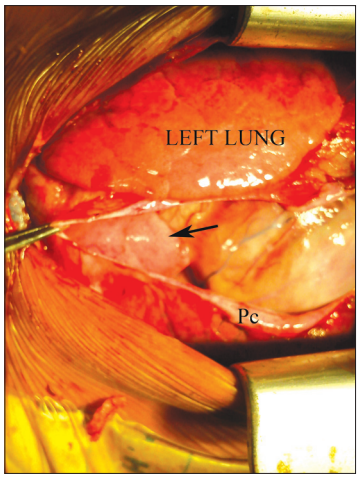

Figure 2: Part of the right lung (arrow) seen in the pericardial cavity following pericardiotomy. Pc: pericardium

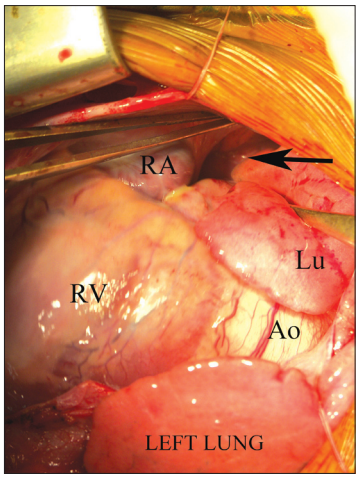

Figure 3: Partial congenital absence of right pericardium (arrow) seen with herniation of part of the right lung (Lu) into the pericardial cavity. RV: right ventricle; Ao: aorta; RA: right atrium

pericardium (CAP) occurs 1 in 14000, and has a male predominance.[2,3] CAP is a rare anomaly, and is almost always detected intraoperatively during intrathoracic surgeries.[1,2] Partial absence of right pericardium is rare due to persistence of the right cardinal vein, which eventually develops into right superior vena cava. Right cardinal vein (right duct of curvier) ensures closure of right pleuropericardial membrane. Early atrophy of left cardinal vein leads to partial or complete absence of left pericardium, depending on the blood supply.[1,4] Chest X-ray only raises suspicion and has no diagnostic role in pericardial defects.[4] Right-sided defects are difficult to image echocardiographically.[4] The Mayo clinic has reported 15 cases of partial or complete absence of pericardium detected among 34,000 cardiovascular surgeries performed over 40 years, none of which were diagnosed pre-operatively.[1] Right-sided defects and complete forms are rare compared with left-sided defects.[1-5]

Surgical intervention for CAP is mandated only in symptomatic cases, and is usually recommended for left-sided defects due to the risk of cardiac herniation.[1-4]

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