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Case Report

Surgery Section

Surgical Repair of Atrial Septal Defect and Lung Lobe Resection for Pulmonary Sequestration and Cystic Malformation in One Stage: A Case Report

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ABSTRACT

Incidence of congenital cystic malformation of lungs ranges from 1 in 11,000 to 1 in 35,000 live birth. Concurrence of Congenital Pulmonary Airway Malformation (CPAM) with congenital heart disease is even infrequent. Even rarer, is the incidence of congenital pulmonary vascular and cystic parenchymal lesions with cardiac defects. The non functioning lung tissue is separated from bronchial tree and vascularised by aberrant artery from the systemic circulation. Simultaneous repair of cardiac and pulmonary defects, though challenging, but are preferable because of surgery being done in single stage with better cosmesis. A 10-year-old female child presented to the hospital with recurrent chest infections and respiratory distress while running since five years of age. The patient underwent atrial septal defect closure and right lower lobectomy for CPAM in one stage through midline sternotomy. We conclude that such complex cases can be managed successfully with meticulous planning and multidisciplinary team approach.

Keywords: Congenital pulmonary airway malformation, Congenital heart disease, Congenital cystic adenomatoid malformation, Right lower lobectomy

CASE REPORT

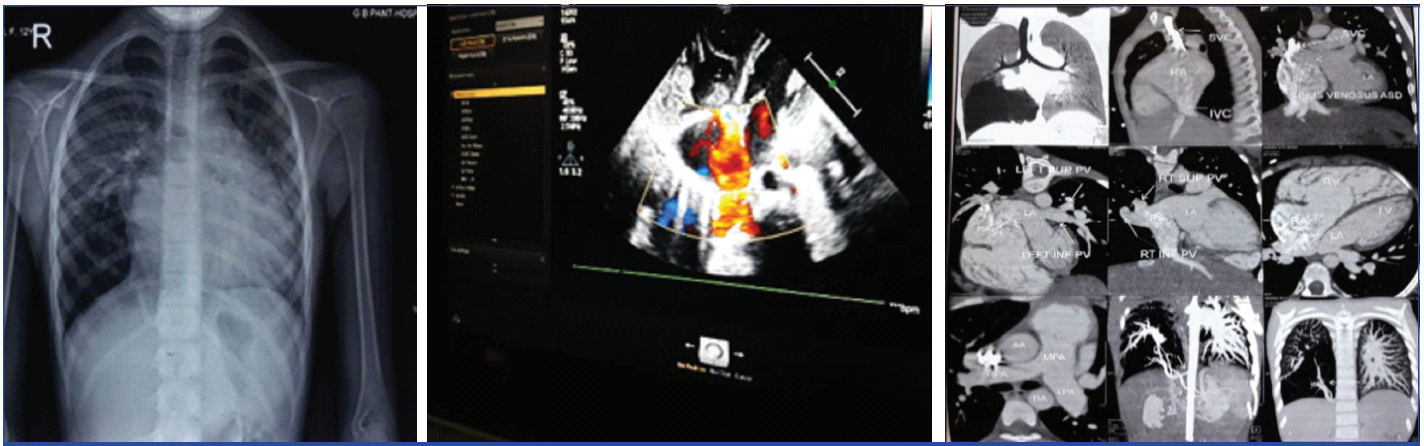
A 10-year-old female patient, presented with recurrent chest infections and respiratory distress, while running since five years of age. Her clinical examination was done. On auscultation widely split S2, grade 3/6 mid diastolic murmur was noted at lower left sternal border with loud P2. Bilateral lung field auscultation revealed; decreased air entry at right lower lung base. Electrocardiogram revealed incomplete right bundle branch block.

Chest X-ray showed hyperlucent right lower lobe with prominent vascular markings. Cardiac silhouette revealed, enlarged right atrial shadow and prominent pulmonary trunk [Table/Fig-1].

Echocardiography revealed, large ostium secundum atrial septal defect; extending to Superior Vena Cava (SVC) margin with right pulmonary veins draining to right atrium and bidirectional shunt across the septal defect. There was moderate pulmonary arterial hypertension with right ventricular systolic pressure of 40 mmHg [Table/Fig-2].

Thoracic CT angiography of heart and great vessels showed large ostium secundum atrial septal defect with right pulmonary veins, draining into right atrium. Right lower lung lobe showed, a well-defined area of reduced attenuation measuring 9.3×9.3×8.3 cm with intralesional cystic areas. Intralesional bronchovascular bundle was noted. Two aberrant feeding arteries, both originating from descending aorta at the level of diaphragmatic hiatus and at the level of origin of celiac axis was noted. However, normal venous drainage into the pulmonary veins was noted. The arteries were draining into right inferior pulmonary veins directly [Table/Fig-3].

Diagnostic confirmation was made through transoesophageal echocardiogram, which confirmed the diagnosis of large atrial septal defect and right-sided pulmonary veins draining to right

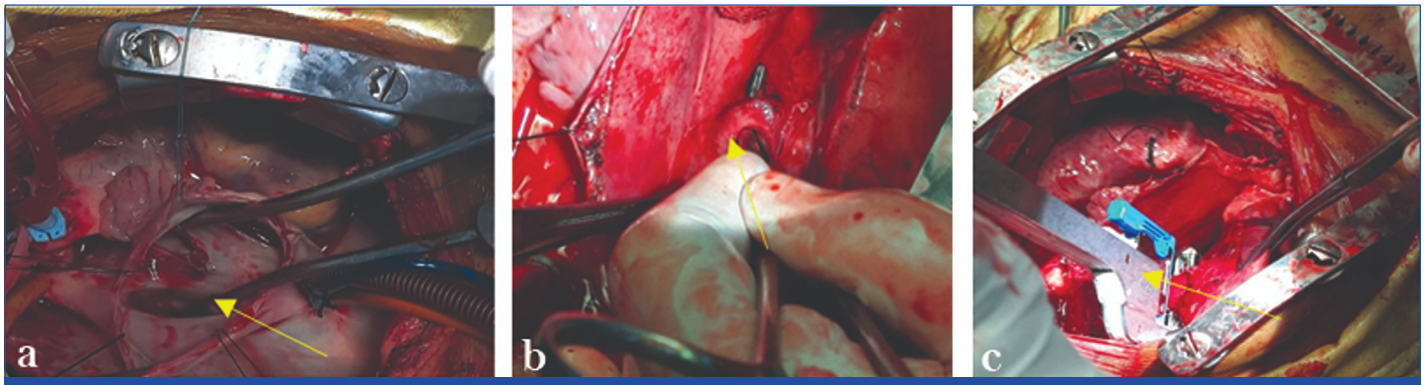


[Table/Fig-1]: Chest X-ray posteroanterior view showing hyperlucent right lower lobe with prominent vascular markings. **[Table/Fig-2]:** Echocardiographic image showing Atrial Septum Defect (ASD). **[Table/Fig-3]:** Computed tomography angiography heart and great vessels showing two aberrant feeding arteries both originating from descending aorta at the level of diaphragmatic hiatus and at the level of origin of celiac axis. (images from left to right)

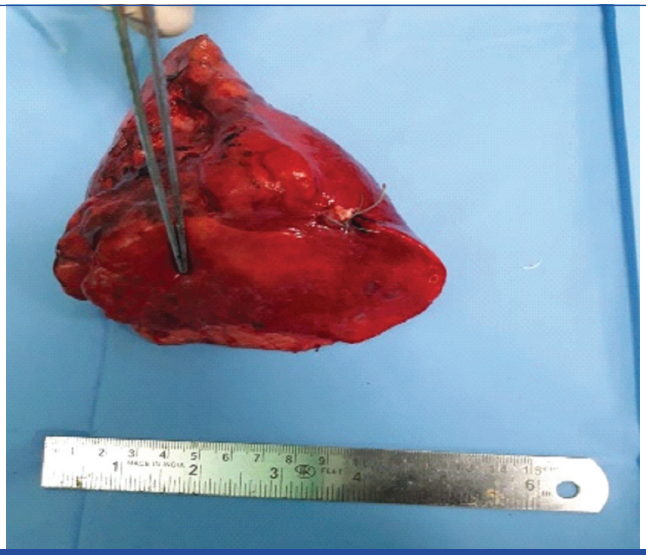
atrium. Midline sternotomy was done. Cardiopulmonary bypass was initiated and heart was arrested in diastole. Pericardial patch closure of atrial septal defect and baffling of right pulmonary veins to left atrium was done. The pleura was widely opened. Aberrant vessel supplying the sequestration was identified and ligated. Hilar dissection was done. Interlobar groove was dissected and intersegmental vessels were ligated and bronchus was divided by bronchial stapler [Table/Fig-4a-c]. The surgical specimen was sent for histopathological examination which confirmed to be Congenital Cystic Adenomatoid Malformation (CCAM) type 2 [Table/Fig-5,6]. Postoperatively patient had grade 1 air leak, which resolved spontaneously on day three and patient was discharged on 10th postoperative day. Outcome of patient was uneventful after one month follow-up.

DISCUSSION

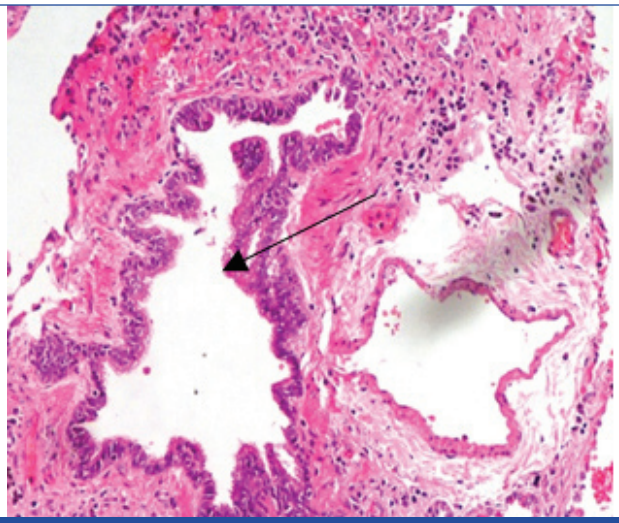
The most common complication of CHD is PH and it occurs more commonly in subjects with left-to-right (systemic-to-pulmonary) shunts. In Atrial septal defect Pulmonary Hypertension (PH) can occur as a complication, although less frequently. The common condition responsible for the development of PH is Congenital Pulmonary Airway Malformation (CPAM). Development of PH is associated with increased risk of perioperative morbidity and mortality [1]. During the branching and proliferation of the bronchial structures CCAM can occur. It is a hamartomatous condition, in which the non functioning lung tissue gets separated from the normal pulmonary structure and it has malignant potential [2]. In addition, congenital cardiac defect and pulmonary sequestration can co-exist as hybrid lesions. Thus, although the pathogenesis of these lesions is poorly understood, they may have a common origin [3]. The condition responsible for the formation of CCAM includes airway obstruction, dysplasia and metaplasia of normal tissues [4]. The most common symptoms are the respiratory symptom, which may be present at the time of birth, or it may occur secondary to the compression or rupture of the cyst [4]. Highest incidence of associated anomalies, up to 60% occur in type 2 CCAMs [5]. Type 2 CCAMs account for 15-30% of cases and arise from terminal bronchioles. The association of CCAM and CHD is rare and accounts for 15-20% of cases [6]. They are composed of smaller cysts, measuring 0.5-2 cm, as well as solid areas that may be difficult to distinguish from surrounding tissue. These are lined by ciliated cuboidal or columnar epithelium, and elements of bronchioles or alveoli may be seen [6]. Type 2 CPAM has the worst prognosis, because they are often associated with other defects such as Atrial Septum Defects (ASDs), Ventricular Septum Defects (VSDs), and Patent Ductus Arteriosus (PDA), and other renal and skeletal anomalies [7]. Pretricuspid shunt patients (i.e., ASD or unobstructed anomalous pulmonary venous return) generally do not develop PH at all or present with PH in adulthood [8]. Most of the reported procedure for these hybrid lesion include either thoracotomy or partial sternotomy for cardiac defect closure and separate anterior and posterior thoracotomy for lung lobe resection. Similar case report published, in which a 6-month-old child, with atrial septal defect and PH underwent right lobectomy for CPAM. They concluded, that with careful preparation and multidisciplinary team approach, such difficult cases can be managed efficaciously [9].



[Table/Fig-4]: a): Intraoperative image with arrow showing Atrial Septum Defect (ASD); b): Arrow showing vessel supply to lung; c): arrow showing use of stapler during division of lung.



[Table/Fig-5]: Surgically resected specimen.



[Table/Fig-6]: Histopathological examination showing cystic areas in specimen.

CONCLUSION(S)

Atrial septal defect with lung sequestration is rare presentation. Both atrial septal defect closure and lobectomy was done in same siting to reduce hospital cost and morbidity of the patient, as well as cosmesis. Hence, through midline sternotomy atrial septal defect closure and right lower lobectomy can be performed as a single stage procedure.

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