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A Rare Pediatric Case Report of a Hybrid Lesion of Congenital Pulmonary Airway Malformation and Bronchopulmonary Sequestration

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Abstract | **Case Report**

Congenital malformations of the lungs are rare. Congenital pulmonary airway malformation and bronchopulmonary sequestrations are commoner malformations amongst them. Co-existence of both in a single lesion is extremely rare. Such hybrid lesions or rather malformations have been sparsely reported. We report a similar lesion in an eleven years old male who presented with hemoptysis and diagnosed to have a hybrid lesion.

Keywords: Bronchopulmonary sequestration, Congenital pulmonary airway malformations, Hybrid lung lesion.

INTRODUCTION

Congenital lesions of lung are rare, with overall incidence of 1/10000 to 1/25000 births [1]. Congenital pulmonary airway malformations (CPAM) and Bronchopulmonary sequestration (BPS) are the more commo malformations [2, 3] Their occurrence in hybrid form is extremely rarer [4]. Only 40 such cases have been reported in the literature world-wide so far. Hybrid malformations usually present in the neonatal life as respiratory distress and if left undiagnosed can lead to repeated chest infections in the pediatric age group. Diagnosis can be made antenatally in-utero or postnatally due to complications of the lesion, such as recurrent pneumonias, lung abscesses and pneumothorax [3]. Here we present a case of an eleven year old male with a hybrid CPAM and the management obstacles of the same.

CASE REPORT

An eleven years old male child presented to the casualty with the chief complaints of hemoptysis, hematemesis and cough since 4-5 days associated with a single episode of high grade fever 4 days back. On examination patient had tachycardia without tachypnea or fever, and on local examination, there was decreased air entry in the right middle and lower zones. Plain Roentgenogram of the chest revealed right sided pleural effusion with a cavitary lesion in the right middle lobe with an air fluid level. Patient underwent Computed Tomography Pulmonary Angiography(CTPA) which revealed a large well-defined multi-cystic lesion with inter-communicating cysts, showing air-fluid level and hyperdense dependent area within involving the posterior and lateral basal segment of the right lower lobe along with multiple tortuous tuft of vascular channels along its wall with arterial feeders from pulmonary artery, right intercostal arteries and infra-diaphragmatic abdominal aorta with early venous drainage via right inferior pulmonary vein draining into supra-diaphragmatic IVC (Figure 1).

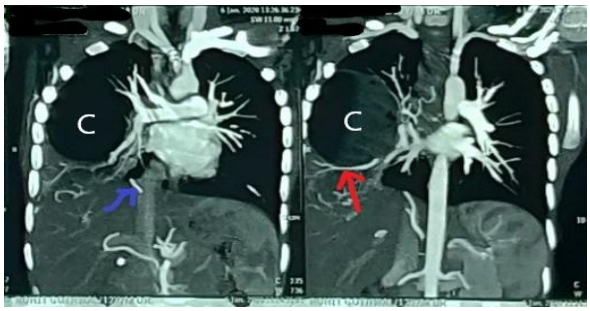

Fig-1: CTPA of the patient showing blood supply of the cystic lung lesion from the infra-diaphragmatic aorta. (Blue arrow) C-cystic lesion

There was an active contrast leak through one of the feeders into the cavitary lesion suggestive of

hybrid CPAM associated with pulmonary sequestration with an active contrast leak. Patient underwent Interventional radiology guided embolization of the tortuous feeding arteries with 50% glue-lipoidal solution and a 10Fr pigtail insertion in the right hemithorax. The differential diagnosis of arterio-venous malformation was also kept in mind. Patient then underwent a formal right posterolateral thoracotomy with excision of the cyst with right lower lobectomy after 10 days of embolization. Intra-operative findings revealed a large number of feeders to the cystic lesion which were controlled using ultrasonic scalpel (Figure 2).

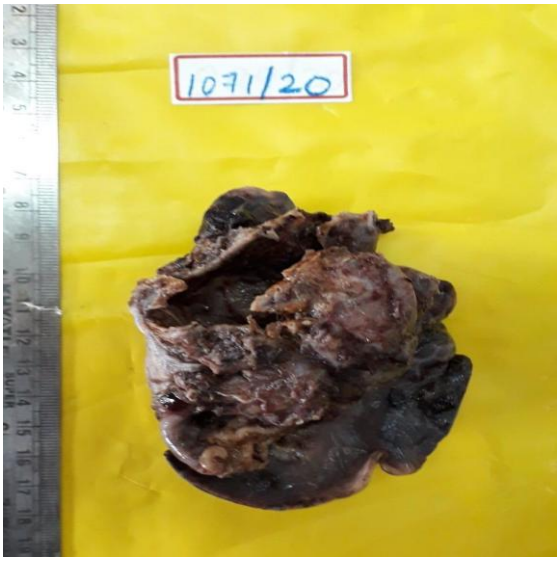

Fig-2: Gross specimen of the hybrid lung lesion showing cystic changes

The post-operative course was uneventful, Intercostal drainage was kept for 3 days and patient received antibiotics for 10 days. Histopathological examination concluded the lesion as cystic malformation of the lung (Figure 3).

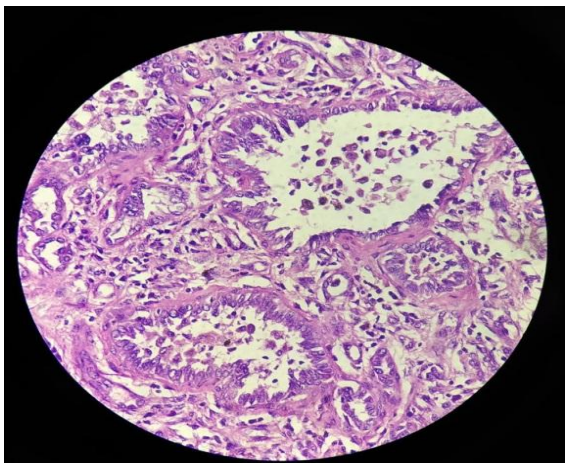

Fig-3: H & E section, 400X, Cyst lined by respiratory epithelium showed Type II pneumocyte proliferation

Patient was followed up monthly for three months and the post- operative follow-up till one year was uneventful.

DISCUSSION

CPAM is an uncommon developmental lung malformation that is thought to occur early in fetal lung development, around the fifth to the eighth weeks of gestation [2]. It is suspected that there is an arrest in normal fetal pulmonary maturation caused by primary bronchial atresia or failure of normal bronchial segmentation leading to dysplasia [5]. This dysplastic broncho-pulmonary tissue consists of numerous, intercommunicating, bronchioles like structures of variable size with many immature cells surrounded by a rim of lung tissue. Its incidence is of the order of 1 in 25,000 to 35,000 live births. CPAM often affects the lower lobes and is often unilateral and more common in boys [5]. Histologic classification of CPAM was first described in 1977 [1]. The Stocker classification system divides the lesions into three groups- Type I, Type II and type III. The most commonly seen CPAM lesion is the Stocker type I accounting for 50-60% of cases [3, 5].

BPS: It constitutes 10-30 % of congenital cystic lung lesions. These are solid non-functioning congenital lung lesions which derive blood supply from aorta rather than pulmonary artery, with absence of communication with the bronchial tree. The incidence of BPS is 0.15 to 1.7%. CPAM and BPS abnormalities occur during branching and proliferation of the bronchial structures. Both the lesions have malignant potential [2, 3]. It is subdivided into intra-lobular and extra-lobular types.

Hybrid malformations: Hybrid malformations are extremely rare and have features of both CPAM and BPS. This suggests a common developmental origin for both and perhaps each represent two ends of a broad spectrum of pathologies. Theories of their pathogenesis include abnormal proliferation of tissues, dysplasia and metaplasia of normal tissues and airway obstruction [6].

In our case report, we present a similar rare congenital cystic lung lesion in a child found on right lung base as an intra-lobar sequestration with systemic arterial blood supply but having a histological appearance of CPAM [4]. CPAM receives arterial supply from pulmonary vasculature, whereas BPS receives arterial supply from aorta [5]. CPAM tissue has connections with the tracheobronchial tree, whereas pulmonary sequestration does not connect with the bronchioles. The incidence of hybrid cases of CPAM and BPS is rare, with only 40 such cases reported in English literature so far. A study conducted in Taiwan showed an incidence of 19% of mixed type hybrid lesions from 1995 to 2008 [6]. Khaladkar SM et al reported a similar case in a 4 year old male child in 2015 [7]. Another such case in neonates was reported

by Khushdil et al in 2018 [8]. Thus, hybrid lesions of CPAM and BPS are very rare and their data sparse. A high index of suspicion should be raised when such lesions are encountered in any neonate or a child. Management should include tackling of the feeding vessels and excision of the lesion.

CONCLUSION

CPAM and BPS can co-exist together (hybrid malformations). They share same developmental ancestry and most likely represent a similar pathogenesis. A high degree of suspicion is needed and the diagnosis of hybrid lesion has to be kept as a differential. This case report represents the successful treatment of such a malformation using interventional radiology and surgery.

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Conflict of Interest: None.

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