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CASE REPORT

A 19 DAYS OLD NEONATE WITH CONGENITAL PULMONARY AIRWAY MALFORMATION: A CASE REPORT

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ABSTRACT

Introduction: *Congenital Pulmonary Airway Malformation (CPAM), previously known as Congenital Cystic Adenomatoid Malformation (CCAM) is a rare dysplastic lesion of the fetal tracheobronchial tree. Here we report a 19 days male child with respiratory distress. Right sided Congenital Pulmonary Airway Malformation Type 1 was diagnosed. Right thoracotomy with upper and middle lobectomy was carried out with successful management of the child.*

Keywords: *Congenital pulmonary airway malformation, Lobectomy, Respiratory distress, Thoracotomy.*

INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM) is a rare congenital birth defect that includes a single, large, multiloculated, cystic mass of abnormal lung tissue. It presents in 0.004%of all pregnancies and constitutes <25% of all congenital pulmonary airway malformation.[1] The incidence of CPAM is reported as 1 in 10000 to 35000 live births.[2] It is due to abnormalities throughout embryogenesis and occur at different stages during lung development, leading to anomalous bronchial morphogenesis.[3] CPAM is diagnosed early during antenatal period due to new advancements.

CASE SUMMARY

A 19-day-old male neonate was admitted to neonatal intensive care unit of our hospital with history of fast breathing and fever for 1 week. There was no history of cough, cyanosis, choking or difficulty at breastfeeding. He was admitted at another hospital for the same complain but there was no improvement because of which he was referred to this hospital.

On examination he was tachypneic, febrile with respiratory rate of 80/minute, heart rate of 165/minute, SPO_2 of 85% in room air. Intercostal indrawing were present with hyper-resonant percussion notes over right lung. On auscultation there was decreased breath sound on right side. Chest X-ray showed hyper lucent right lung with cystic component and shifting of mediastinum to

the left side (Figure 1). CT-scan of chest showed a large well defined non enhancing thin-walled cystic lesion with minimal collection on the dependent part measuring approx. 3.9 x 4.4 x 4.63 cm at apical and posterior segment of right upper lobe. A differential diagnosis of congenital pulmonary airway malformation Type I was made by the radiologist (Figure 2).

Patient was planned for open right thoracotomy with posterolateral approach. At surgery, the malformation was seen involving upper and middle lobe following which upper and middle lobectomies were carried out and the specimen sent for histopathology (Figure 3). He has an uneventful postoperative course. Patient was extubated on 2nd POD with complete resolution of respiratory distress by 4th POD. Feeding was started on 6th day and he was discharged on 11th days after surgery.

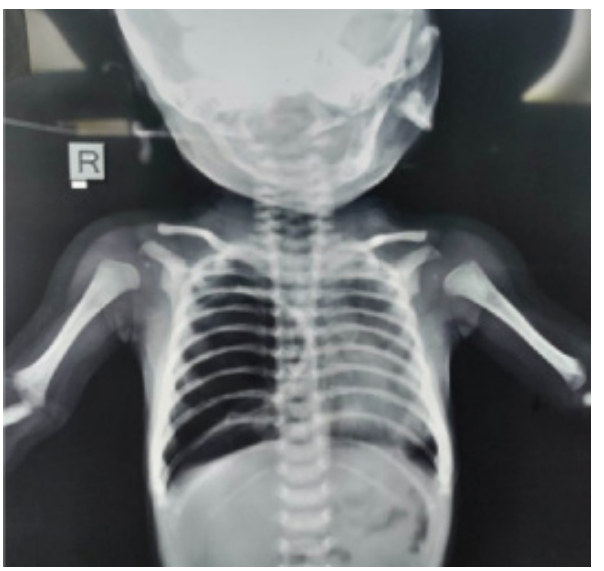

Figure 1: Chest X-ray of patient with congenital pulmonary malformation

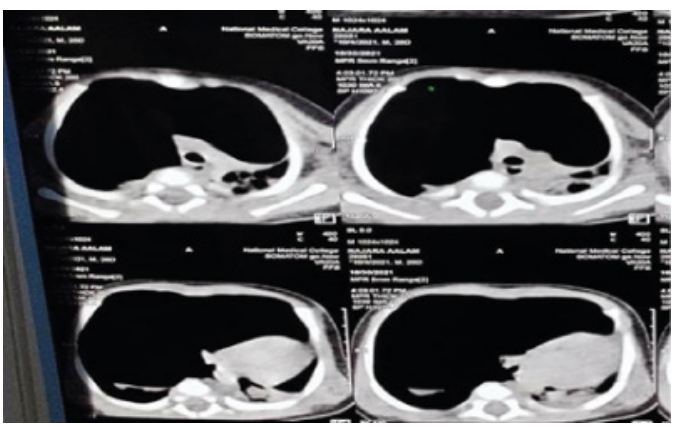

Figure 2: Computed tomography scan of 19-days infant with congenital pulmonary malformations

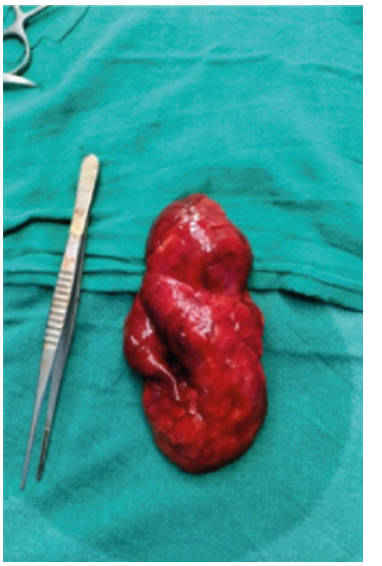

Figure 3: Excised right upper & middle lobe showing a large cystic cavity

DISCUSSION

Although CPAM is rare; it is the most common type of congenital lung disease that can present from prenatal period to childhood. About 95% of congenital cystic lung disease are caused by congenital pulmonary airway malformation with incidence of 1 in 10000 to 35000 livebirths[2,3] with no gender predominance. These malformations occur sporadically with no genetic predisposition (except Type 4), and no association with maternal factors. It results from the cessation of lung development during different stages of embryogenesis.[4] There are various genes associated with this condition which include thyroid transcription factor gene (Nkx2), sex-determining region Y- box 2 gene (Sox2), Hox gene (Hoxb-5), Ying Yang 1 gene (Yy1), fatty acid-binding protein-7 gene (FABP-7), acyl-CoA synthetase 5 (ACSL5) platelet-derived growth factor B gene (PDGF-B), sonic hedgehog (SHH), bone morphogenetic protein 4 (BMP4), sprouty 2 (SPRY2), Wnt signaling pathways, transforming growth factor B (TGFB), and fibroblast growth factors 10, 9 and 7 (FGF10, 9, 7).[5,6]

New Stocker et al.'s classification of CPAM[7]

Type 0	Acinar dysgenesis and dysplasia of the airways. Incompatible with life
Type 1	One or multiple cysts over 2 cm in diameter of bronchus or bronchiole
Type 2	One or multiple cysts under 2 cm in diameter of bronchiole

Type 3	Solid lesion with some cyst under 0,5 cm of bronchiole and alveolar duct
Type 4	Acinar origin multiple cysts

Clinical presentation of congenital pulmonary airway malformation may be symptomatic or asymptomatic. Respiratory distress is the most common presentation in neonate. It may range in severity from grunting, tachypnea, and a mild oxygen requirement to fulminant respiratory failure requiring aggressive ventilator support or extracorporeal membrane oxygenation (ECMO). Pulmonary hypoplasia may arise as a consequence of a large CCAM, mediastinal shift may compromise cardiac and respiratory function, spontaneous pneumothoraxes may occur, and air trapping within the cyst leads to compression of functional pulmonary tissue. Associated anomalies are very uncommon.[8]

CPAM usually arises from one lobe of the lung, lower lobes being the most common site. Bilateral lung involvement is rare. CPAM lesion have an equal left and right sided incidence.

Fetal ultrasound is diagnostic modality for Congenital pulmonary airway malformation during prenatal period. For further evaluation of the lesion fetal MRI may be indicated. CPAM types 1 and 4 appear as one or two large air-filled cysts while Type 2 appears as multiple smaller air-filled cysts, giving a “bubbly” appearance on a chest X-ray; Type 3 presents as a solid, homogenous mass on chest X-ray, with a mass effect on the mediastinum.[2,9] For further evaluation, CT chest is indicated.

Neonate with respiratory compromise due to CPAM require surgical resection, usually by lobectomy. In asymptomatic neonates an early elective resection is warranted because of the risk of infection and occult malignant transformation.[10]

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