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Selecting An Early Neonatal Timeframe For The Surgical Management Of High Risk Congenital Pulmonary Airway Malformation Type 1 : A Case Report

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Abstract

BACKGROUND:Congenital Pulmonary Airway Malformation Type 1 can be managed conservatively in low risk individuals.Identifying high risk factors in the postnatal period is important to plan for surgical intervention

CASE PRESENTATION: We describe a case of antenatally diagnosed CPAM Type 1 with high risk features on radiological imaging and its surgical management

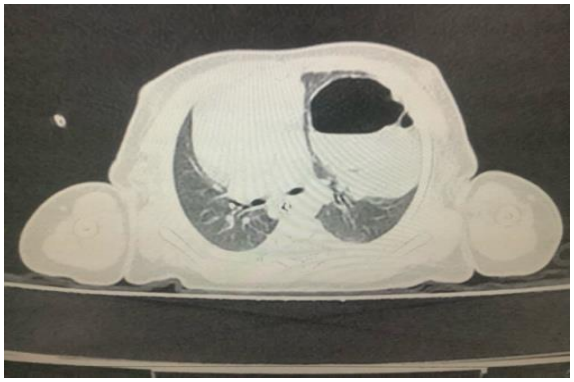
CONCLUSION: Early surgical intervention avoids complications of pneumothorax,pulmonary infections and probable malignant transformation

Introduction

Pulmonary Airway Malformation (CPAM) previously also known as Congenital Cystic Adenomatoid Malformation (CCAM) is a developmental anomaly usually resulting from sporadic abnormalities of branching morphogenesis of the lung often as a result of in utero airway obstruction or atresia[1] It is the most common congenital lung lesion and is composed of hamartomatous cystic and adenomatous elements arising from tracheal, bronchial, bronchiolar, or alveolar tissue.The widespread use of routine prenatal ultrasound has led to an increase in prenatal diagnosis.[2] The classification of CPAMs is based on their origination at different levels of the tracheobronchial tree and their subsequent pathology.Type 1,the most common,arises from distal bronchi or proximal bronchioles[3]This paper studies the stratification of a high risk case of Type 1 CPAM and its surgical management in the early neonatal period.

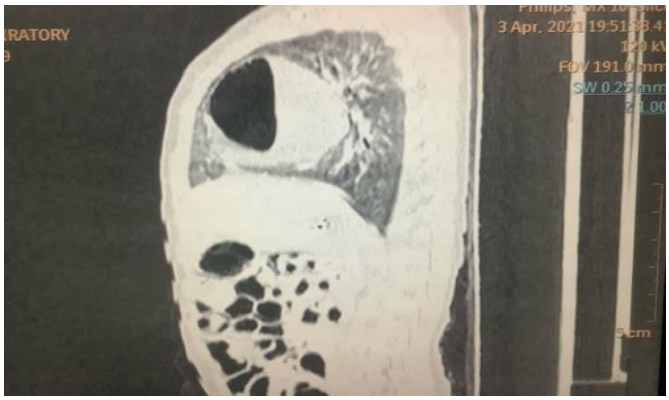
Case Report

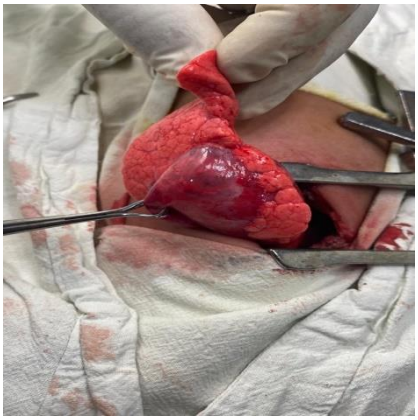
A 23 year old female primi underwent a full term normal vaginal delivery. The 2.6kg female child was shifted to the neonatal intensive care unit as the antenatal ultrasound done at 31 weeks of gestation was suggestive of Congenital Pulmonary Airway Malformation Type 1.The baby was asymptomatic in the postnatal period and was feeding well.Blood investigations and arterial blood gas values were within normal limits.



Further imaging with High resolution computed tomography (HRCT) was done on the second day of life and the radiological features were strikingly significant.

- Hyperdense mass lesion is present in left upper lobe of size 4.6cmx4.6cmx3.7cm showing multiple cysts.
- Few of the cysts had air fluid levels
- The entire mass lesion occupied 42% of the left hemithorax
- This was causing mediastinal shift to the right
- Sagital planes showed that anteriorly the lesion was abutting the anterior chest wall and posteriorly it was surrounded by bronchial airways and vessels.This would be significant intraoperatively





Intraoperative image of the cyst that demonstrates its close relationship with the surrounding lung parenchyma.Separation of the lesion is achieved by harmonic scalpel

Operative Steps

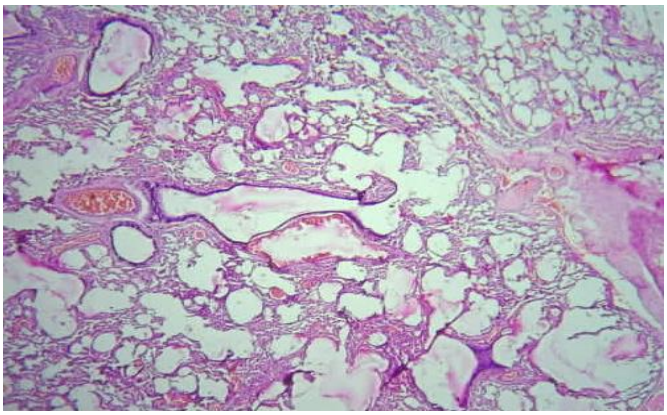
- Left Posterolateral thoracotomy was planned electively on the fifth day of life
- Incision is taken in fifth intercostal space and deepened.Latissimus dorsi and intercostal muscles are divided and parietal pleura opened to enter thoracic cavity
- Evidence of cystic lesion is seen surrounded by normal lung parenchyma
- Segmentectomy done using harmonic scalpel,

  hemostasis achieved with cautery.Sample is sent for histopath.
- Post resection, bronchial airway leaks are evident.These are closed with prolene.
- Intercostal drain no.16 is placed in the cavity and connected to an air tight bag
- Sterile dressing is done after closure and patient was shifted intubated to the Neonatal Intensive Care Unit

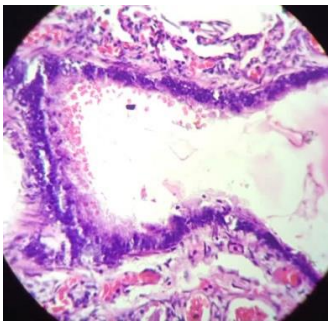
  where she was maintained on low pressure ventilation and later extubated

Histopathology

Sections studied show multiple thin walled cysts lined by pseudostratified ciliated columnar epithelium.Some of these cysts show evidence of haemorrage.Scattered cartilage plates are seen beneath the wall of large cysts. The features favour Type 1 Congenital Pulmonary Airway Malformation



Cysts with papillary infoldings demonstrated on low power



Pseudostratified ciliated columnar epithelium of the cyst with scattered red blood cells seen on high power

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Conclusion

Asymptomatic patients with CPAM can be managed conservatively.

In such cases the indications for surgical intervention are:

Large lesion (>20% of hemithorax)

Bilateral or multifocal cysts

Family history of Pleuropulmonary blastoma

In case of above mentioned factors,conservative management is associated with relapsing courses of pulmonary infections, pneumothorax, hypoxia, malignant transformation and in worst case scenarios,death.[4]Identifying high risk patients for surgery and poor outcomes in case of delay is an important step in the neonatal management of the disease.Since the patient mentioned herein had a lesion that measured 42% of the left hemithorax,she was an ideal candidate for early surgical intervention.

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