International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Congenital Cystic Adenomatoid Malformation (CCAM): A Case Report

Jyotsna Wader[*], Neha Gajbi[**@], Sujata Kumbhar[***]

[*]Associate Professor, [**]MD Student 2nd year, [***]Assistant Professor
Department of Pathology, Krishna Institute Of Medical Sciences, Karad, Maharashtra, India.

[@]Correspondence Email: dr.komalgajbi@gmail.com

Received: 01/02//2013 *Revised: 19/03/2013* *Accepted: 26/03/2013*

ABSTRACT

Introduction: Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development. CCAM is a cystic area within the lung that stems from abnormal embryogenesis. An adenomatous overgrowth of the terminal bronchioles with a consequent reduction in alveolar growth occurs.
Case report: We report a case of a premature male fetus with 19.2 weeks gestational age was born to a 26-year-old G2P0L0A2 mother by emcredil instilled termination of pregnancy.
Antenatal USG detected anomalies – congenital cystic malformation in right lung and prominent left renal pelvis necessitating the misoprost induced termination of pregnancy.
At autopsy body of the fetus revealed multiple thin walled cystic spaces, largest cyst m. 0.5cm in diameter on lower lobe of right lung. Rest of the lobes of right lung and left lung revealed no pathology. On microscopy right lower lobe of the lung showed multiple evenly spaced cysts resembling dialated terminal bronchioles lined by ciliated cuboidal to columnar epithelium with scanty connective tissue beneath the epithelium. Plain X ray of the abortus revealed – Scoliosis of the vertebral column.
Diagnosis: Congenital cystic adenomatoid malformation (CCAM)
Conclusion: we report the case because of its rarity.
Keywords: Congenital cystic adenomatoid malformation, rare, lung abnormality.

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development. CCAM is a cystic area within the lung that stems from abnormal embryogenesis. An adenomatous overgrowth of the terminal bronchioles with a consequent reduction in alveolar growth occurs.

CASE REPORT

We report a case of a premature male fetus with 19.2 weeks gestational age was born to a 26-year-old $G_2P_0L_0A_2$ mother by emcredil instilled termination of pregnancy.

Antenatal USG detected anomalies – congenital cystic malformation in right lung and prominent left renal pelvis necessitating the misoprost induced termination of pregnancy.

At autopsy body of the fetus revealed multiple thin walled cystic spaces, largest cyst m. 0.5cm in diameter on lower lobe of right lung. Rest of the lobes of right lung and left lung revealed no pathology.

On microscopy right lower lobe of the lung showed multiple evenly spaced

cysts resembling dilated terminal bronchioles lined by ciliated cuboidal to columnar epithelium with scanty connective tissue beneath the epithelium.

Plain X ray of the abortus revealed – Scoliosis of the vertebral column.

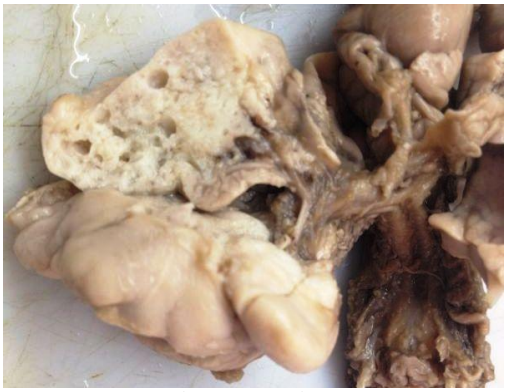

Fig 1 - Cut section of lower lobe of right lung showing multiple cysts.

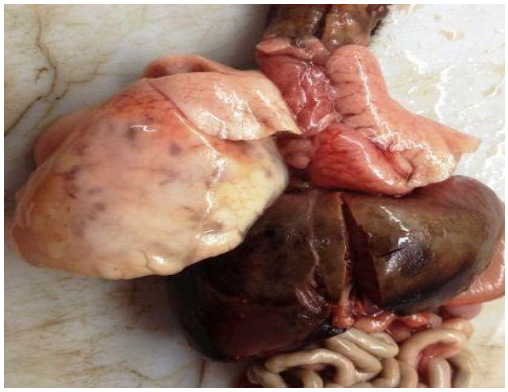

Fig 2- Gross appearance of right lung externally showing multiple cysts.

Microscopy:

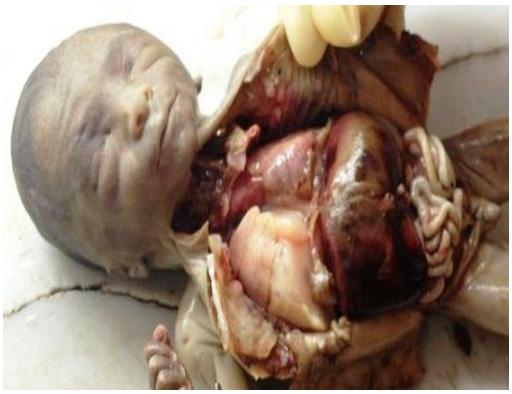

Fig 3- Abortus with multiple small cystic areas on right lung with multiple mediastinal shift to left.

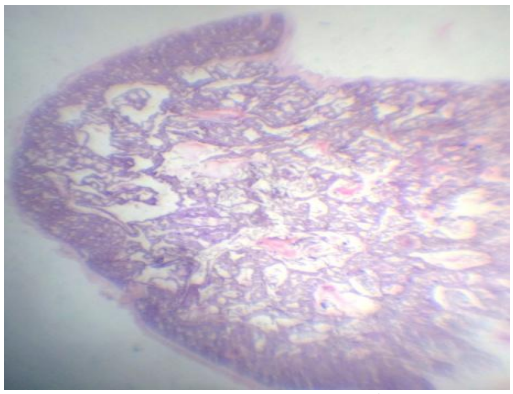

Fig 4- H & E – Scanner view - show lobe of lung with evenly spaced cysts.

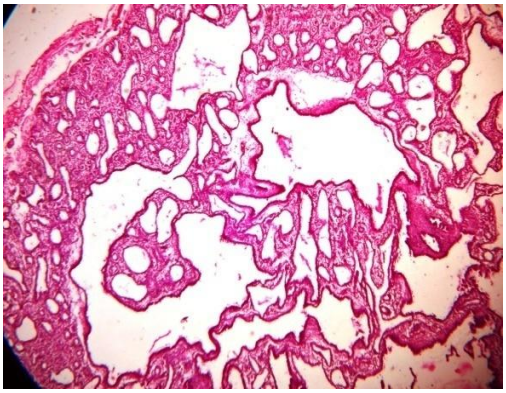

Fig 5: H & E: 50x- Sections through the right lower lobe of the lung - multiple cysts resembling dilated terminal bronchioles.

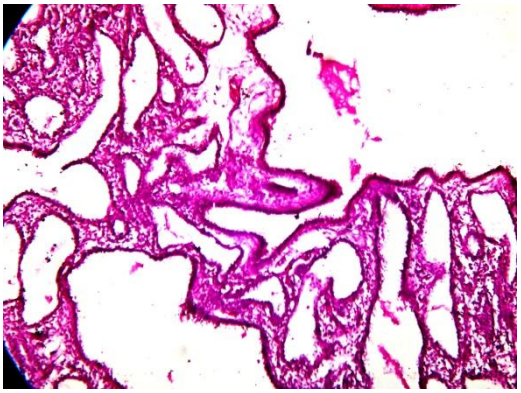

Fig 6: H & E: 100x - Sections through the right lower lobe of the lung show multiple evenly spaced cysts resembling dilated terminal bronchioles lined by ciliated cuboidal to columnar epithelium with scant amount of connective tissue beneath the epithelium.

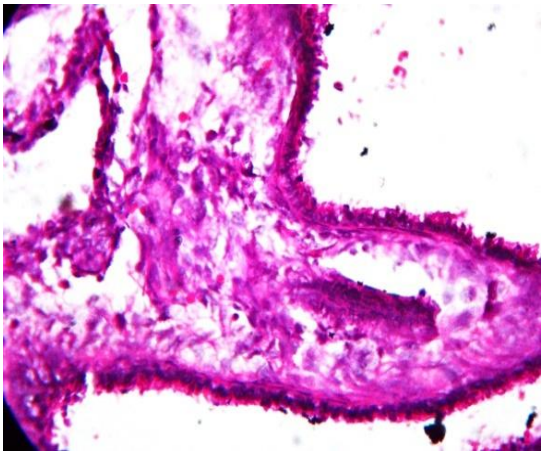

Fig 7 : H & E : 400x - Sections through the right lower lobe of the lung show multiple evenly spaced cysts resembling dilated terminal bronchioles lined by ciliated cuboidal to columnar epithelium with scant amount of connective tissue beneath the epithelium.

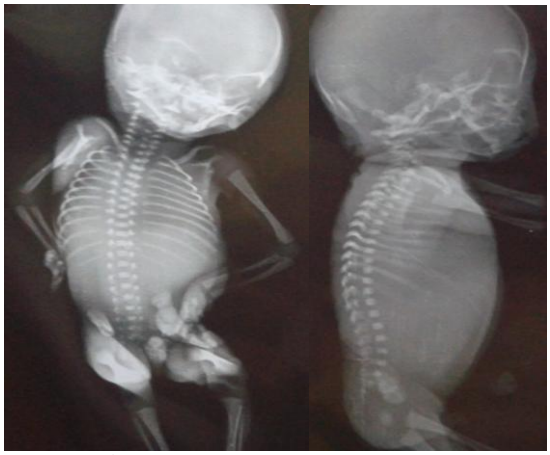

Fig 8 X ray fetus: showing scoliosis.

DIAGNOSIS: Congenital cystic adenomatoid malformation (CCAM)

DISCUSSION

Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development. CCAM is a cystic area within the lung that stems from abnormal embryogenesis. An adenomatous overgrowth of the terminal bronchioles with a consequent reduction in alveolar growth occurs. [1]

In CCAM, usually an entire lobe of lung is replaced by a non-working cystic piece of abnormal lung tissue. This abnormal tissue will never function as normal lung tissue.

CCAMs occur with equal frequency in both lungs.

Incidence – It is a rare congenital lesion, with a reported incidence of 1 in 25000 to 35000 pregnancies. [2]

Types

Stocker has divided CCAM into five different categories based on the proposed site of defect in the tracheobronchial tree. [5]

1. Type 0 – Acinar dysplasia; lungs are firm and small.
2. Type 1 - Is characterized by multiple large cysts, (up to 10 cm) , cyst are lined by ciliated columnar to pseudostratified tall columnar epithelium overlying a thin to moderately thick fibromuscular layer.
3. Type 2 - Shows multiple cysts 2-5 cm in diameter. Small, uniform cysts, irregular proliferation of ecstatic structures resembling bronchioles.
4. Type 3 - consists of cyst size upto 2cm. solid, bulky lesion. Irregular curving channels and small air spaces lined by plumb cuboidal epithelium.
5. Type 4 - consists of cyst size upto 7 cm. Microscopy shows multilocular, large cysts lined by flattened alveolar lining cells.

It is important to remember that these fetal CCAM's cannot, in most circumstances, be classified using the Stocker type 1-4 criteria and that attempts to do so will not be rewarding. [5]

So according to Enid Gilbert-Barness: Potter's Pathology of the fetus, infant and child, second edition, 2007, classification should not be applied to fetal lung tissue hence our case has not classified with types.

Causes [3, 4]

The underlying cause of CCAM is unknown.

- Resected CCAMs show signs of increased cell proliferation and decreased apoptosis.
- Studies have investigated the role of *HOXB5* gene and protein expression, as well as other growth factors such as mesenchymal platelet–derived growth factor-BB.

The development of the respiratory system begins at 3 weeks of gestation, and aberrations in developmental processes may give rise to a group of structural abnormalities collectively referred to as bronchopulmonary foregut malformations. These lesions include congenital cystic adenomatoid malformations (CCAMs).

So, as a consequence of abnormal embryogenesis during the first 6–7 weeks of pregnancy, involving maldevelopment of terminal branches leads to CCAM.

Differential Diagnoses

- Pulmonary Sequestration
- Congenital Pneumonia
- Hemothorax
- Pleural Effusion
- Pneumatocele
- Pneumothorax

Radiology gives the morphological assessment of the lung cavities, but it is inadequate to differentiate CCAM from other cystic lung diseases. The differential diagnosis is essential, since malignancy has been associated with large cyst-type CCAM, including rhabdomyosarcoma and bronchioloalveolar carcinoma.

Complications

May be asymptomatic, until when infection, pneumothorax or malignant degeneration may occur.Or can lead to –

respiratory distress, mediastinal shift, hypoplasia of normal lung tissue, polyhydramnios and cardiovascular compromise leading to fetal hydrops and death.

If hydrops develops, this places the mother at a slightly increased risk for "mirror syndrome."

The treatment

Treatment of choice for CCAM is a lobectomy, surgery will be performed at approximately 2 to 6 months of age.

Thoracocentesis allows drainage of a large cyst with immediate decompression of the CCAM; however, fluid rapidly reaccumulates, thus negating the benefit of the procedure. [2]

Another option is to place a thoracoamniotic shunt that continually drains fluid from the CCAM to the amniotic space. This is most beneficial when the CCAM consists of a large fluid-filled cyst. Complications such as obstruction and shunt dislodgement may occur.

CONCLUSION

We report the case because of its rarity. And it is important to diagnose as at birth, some babies born with CCAM will not be able to breathe effectively due to the large mass in their chest causing airway compression and this may have an effect on their heart rate. These babies need immediate intensive care to stabilize them until surgery is performed. If the CCAM is not removed, there is an increased risk of it becoming cancerous.

So early diagnosis and Postnatal surgery is recommended to remove the risk of direct complications, such as recurrent infection and pneumothorax. In addition, the malignant potential in later life has long been recognized.

REFERENCES

1. Luca Morelli et al, Pulmonary congenital cystic adenomatoid malformation, type I, presenting as a single cyst of the middle lobe in an adult: case report, Diagnostic Pathology *2007, 2:17:1746-1596.*
2. Curran PF et al, Prenatal steroids for microcystic congenital cystic adenomatoid malformations, J Pediatr Surg. 2010, 45 (1): 145–50.
3. Peranteau WH, Wilson RD et al, Effect of maternal betamethasone administration on prenatal congenital cystic adenomatoid malformation growth and fetal survival, Fetal Diagn Ther 2007,22 (5): 365–371.
4. Davenport M, Warne SA, Cacciaguerra S, Patel S, Greenough A, Nicolaides K: Currentoutcome of antenally diagnosed cystic lung disease, *J Pediatr Surg* 2004, 39**:**549-556.
5. Enid Gilbert – Barness, Potters pathology of the fetus, infant and child: 2nd edition, 2007.

How to cite this article: Wader J, Gajbi N, Kumbhar S. Congenital cystic adenomatoid malformation (CCAM): a case report. Int J Health Sci Res. 2013;3(3):86-90.

****