

Early Surgical Resection for Symptomatic Congenital Cystic Adenomatoid Malformation (CCAM) in the Newborn Child

Karavdic K^{1*}, Pilav I², Mešanovic S², Spahovic R³, Mesic A⁴, Sefic Pasic I⁵ and Udovicic Gagula D⁶

¹Department of Pediatric Surgery, Clinical Center of University Sarajevo, Bosnia and Herzegovina

²Department for Thoracic Surgery, Clinical Center of University Sarajevo, Bosnia and Herzegovina

³Department for Neonatal Intensive Care, Pediatric Clinic, Clinical Center of University Sarajevo, Bosnia and Herzegovina

⁴Department for Anesthesiology, Clinical Center of University Sarajevo, Bosnia and Herzegovina

⁵Department for Radiology, Clinical Center of University Sarajevo, Bosnia and Herzegovina

⁶Department for Pathology, Clinical Center of University Sarajevo, Bosnia and Herzegovina

***Corresponding author:** Kenan Karavdic, Department of Pediatric Surgery, Clinical Center of University Sarajevo, Patriotske lige 81, 71000 Sarajevo, Bosnia and Herzegovina, Tel: 38761202508; E-mail: kenan.kv@bih.net.ba

Case Report

Volume 2 Issue 4

Received Date: September 17, 2018

Published Date: October 04, 2018

Abstract

Congenital cystic adenomatoid malformation (CCAM) of the lung is a rare congenital abnormality. We present here a one month male child who had no respiratory distress after birth, but gradually developed dyspnea. The diagnosis was made prenatally. After further diagnosis (chest CT which reveals a large lesion with mediastinal shifting) due to the deterioration of the clinical picture, surgical treatment is indicated in the first month of life.

Keywords: Lung; Malformation; Neonatal; Early; Lobectomy

Abbreviations: CCAM: Congenital cystic adenomatoid malformation; CPAM: congenital pulmonary airway malformation; CT: Computed Tomography.

Introduction

Congenital cystic adenomatoid malformation (CCAM) of the lung is a rare congenital abnormality. The incidence of CCAM has been reported in approximately 1 out of 25,000 pregnancies [1]. The pathological feature of the lesion is adenomatoid proliferation of bronchioles that form cysts at the expense of normal alveoli. Its exact pathogenesis is still uncertain although latest literatures

state it is due to fetal maldevelopment of the lung bud between the 6th to 8th weeks of gestation [2]. It is diagnosed by prenatal screening at 18-20 weeks of gestation. CCAM has communication between the cystic malformation and the tracheobronchial tree, so many of the cysts are filled with air, and blood supply usually receives pulmonary circulation. The wall of the cyst is made of smooth muscles and elastic tissue, does not contain cartilage. It is limited to a single lobe in 95% of cases and bilateral in less than 2% of cases [3]. CCAM is usually discovered in neonates because of respiratory distress. Eighty percent (80%) are diagnosed before two years of age [4]. The spectrum of this anomaly ranges from asymptomatic

lesion to pulmonary compromise. CCAM also may occasionally be discovered in older children or adults who have recurrent infectio.

First classification of this lesion as congenital cystic adenomatoid malformation (CCAM) was given by Chin and Tang in 1949 [5]. Although originally described as "congenital cystic adenomatoid malformation," this entity renamed as "congenital pulmonary airway malformation" (CPAM) because cystic morphology in not present in all subtypes, nor adenomatoid. Stocker et al. 1977 classified CPAM into three types, based on clinical, macroscopical and microscopical criteria. In 1993 Adzick introduced a classification system. Microcistical lesions (cysts <5 mm) are usually associated with fetal hydrops and have a poor prognosis. Macrocyt lesions (cysts > 5 mm) are not usually associated with hydrops and have a good prognosis [6].

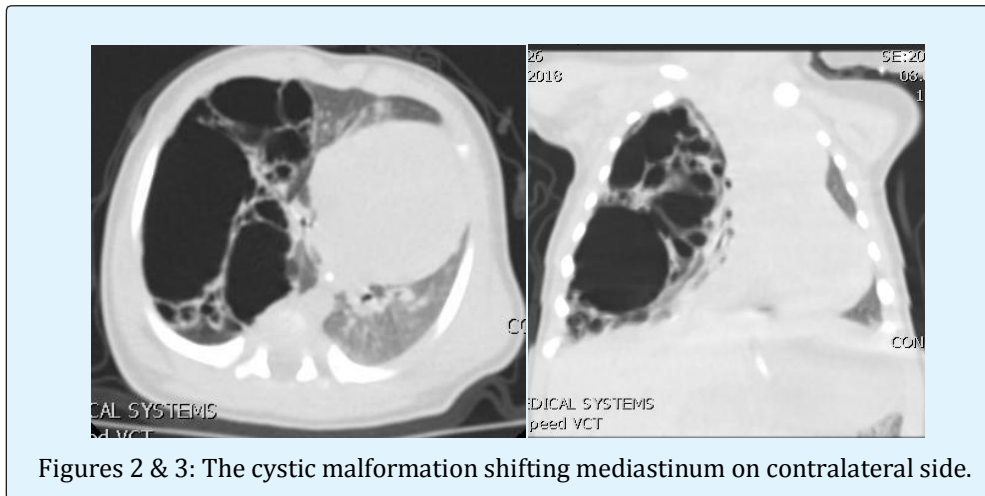
Case report

Male infant, terminal, body weight 3440 g and length 57 cm. API = 9/10. There was no need for oxygen support. Prenatal ultrasound examination suspected on congenital diaphragmatic hernia. X ray chest showed right right side with multiple cystic changes with different levels of airborne pulmonary parenchyma (Figure 1).



Figure 1: X ray chest - right right side with multiple cystic changes.

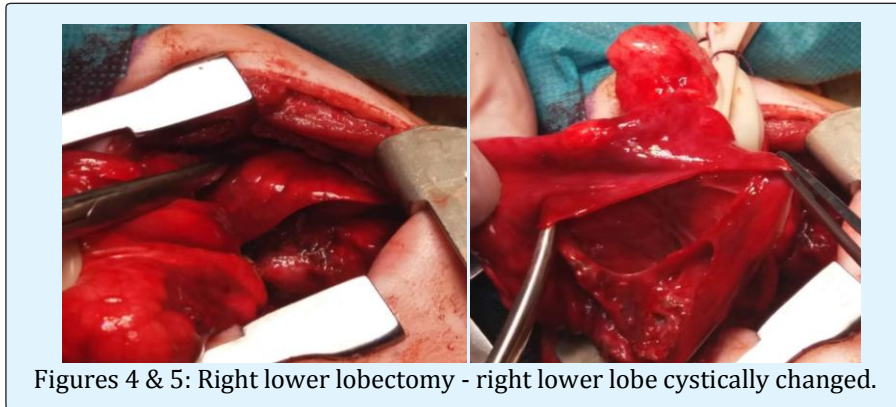
The child is with a normal respiratory status. CT chest is performed: a modified appearance of pulmonary parenchyma in the right half of the thorax. A lumpy parenchyma mostly occupied by hypodontic zones filled with air, thin walls, unrecognizable parenchyma in this area. The cysts are of different sizes from 25 to 61mm. In one of the cysts, noticeable fluency. The cysts are placed in the lower lobe right and compress the pulmonary parenchyma. Cysts suppress mediastinum and large blood vessels on the contralateral left side. No abnormal vascularization of pulmonary parenchyma (Figures 2 & 3).



Figures 2 & 3: The cystic malformation shifting mediastinum on contralateral side.

In the course of further observation, the patient becomes dysfunctional and is indicated for surgical treatment due to deterioration of the respiratory status as well as due to the extent of the radiologically described changes. On examination, pallor was present, breath sounds were decreased and percussion note was dull in

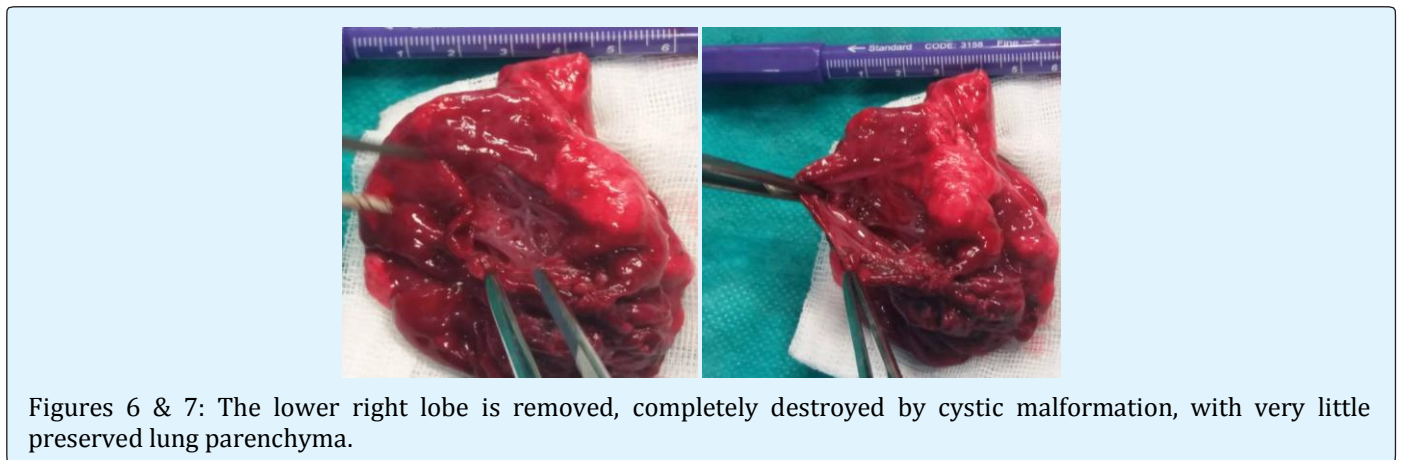
right region. During the surgery the right thoracotomy is performed and there is a large cystic change taking the lower lobe while the middle and upper pulmonary lobes are atelectical [7]. Typical lower right lobectomy was performed (Figures 4 & 5).



Figures 4 & 5: Right lower lobectomy - right lower lobe cystically changed.

The lower right lobe is removed, completely destroyed by cystic malformation, with very little preserved lung

parenchyma (Figures 6 & 7).



Figures 6 & 7: The lower right lobe is removed, completely destroyed by cystic malformation, with very little preserved lung parenchyma.

The postoperative course was satisfactory, antibiotic therapy is ordered. The thoracic tube is removed. The Control Rtg is neat and the patient's postoperative course is satisfactory (Figure 8).

variable-sized cystic formations (larger than 2 cm) outlined by thin walls with honeycomb appearance (Figures 9-11).

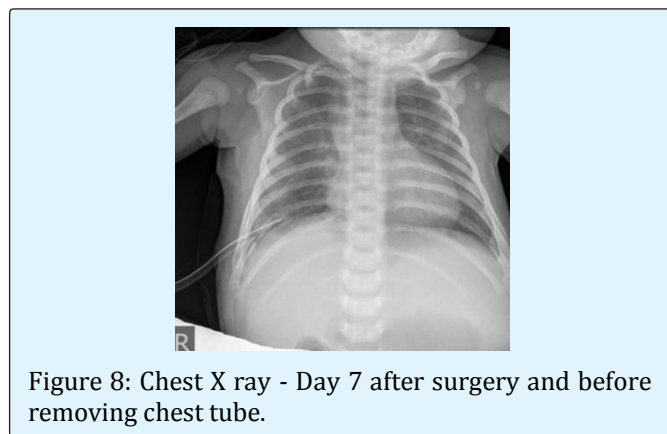


Figure 8: Chest X ray - Day 7 after surgery and before removing chest tube.

The pathohistological finding has shown a gross appearance of the pulmonary lobe showing several

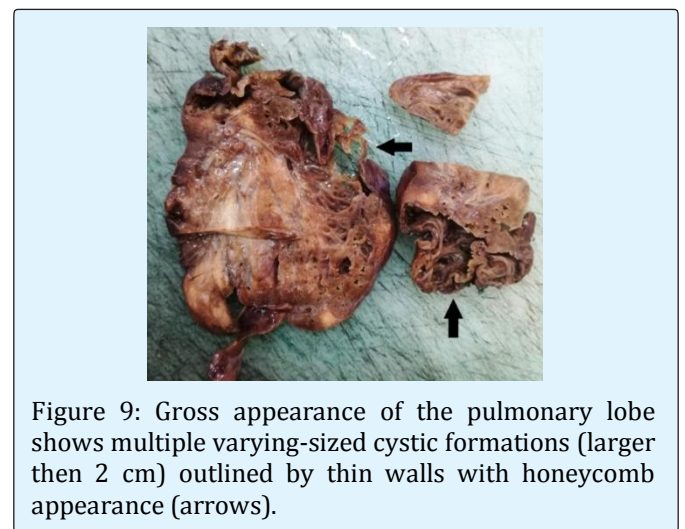


Figure 9: Gross appearance of the pulmonary lobe shows multiple varying-sized cystic formations (larger than 2 cm) outlined by thin walls with honeycomb appearance (arrows).

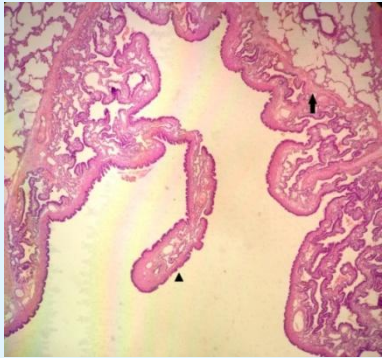


Figure 10: Histology shows several cystic structures varying size with polypoid projections of the mucosa, lined by respiratory-type epithelium (arrowhead), and transition between the cystic lesion and surrounding lung (arrow).

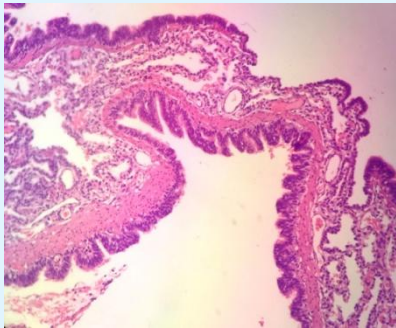


Figure 11: The cysts are lined by pseudostratified columnar epithelium that overlies a fibromuscular layer, without presence of cartilage.

The patient is released from the hospital at home treatment.

Discussion

CCAM is a very rare lung abnormality but the most common congenital cystic lung diseases. CCAM most commonly present with respiratory distress due to the expanding cystic lung mass while others are asymptomatic and later discovered on chest radiograph. Cough, fever, and repeated respiratory infections are the less common presentations. Routine prenatal US screening has become increasingly valuable in detecting CCAM [8] with an accuracy of 65–91% [9,10]. Chest X-ray, computed tomography (CT) scan and clinical follow-up after birth is required in all cases to confirm the diagnosis

and to initiate adequate treatment [11,12]. Significant advances in neonatal intensive care have allowed support, either with conventional ventilation or high-frequency oscillation ventilation, optimising oxygen delivery. A postnatal echocardiogram is essential for assessing cardiovascular function.

Emergency operation for CCAM is performed when the patient has symptoms of respiratory distress in the neonatal period and subsequent diagnostic work-up reveals CCAM. Some newborns with large lesions even asymptomatic, may require a neonatal surgical resection in order to save viable lung parenchyma and reverse mediastinal displacement [7]. In asymptomatic patients, the diagnosis of CCAM is not usually made until childhood or later, when symptoms of frequent pneumonia develop or an incidentally checked chest roentgenography reveals the lesion. In those patients, elective surgical resection is recommended at the time of diagnosis. There is no clear indication for deciding the best timing for operation when the diagnosis is made in utero by fetal USG.

The development of neonatal care, thoracic surgical techniques, anesthesiology, nursing care, and postoperative management all enable safe surgery, even for a neonate. Many reports recommend early surgery for infants in all cases of CCAM [13]. Our opinion that the respiratory distress syndrome in patients with CCAM is sufficient reason for emergency operation. If the patient is asymptomatic and the CT scan shows mediastinal shifting and large cystic lesion, then urgent surgery is required. Aziz and al. suggested that the non-operative approach might be a reasonable alternative for asymptomatic children with perinatally diagnosed CCAM because there was no increase in the postoperative complication rate even if surgery was delayed until the onset of symptoms [14]. Another problem of conservative follow up is the risk of malignancy in which true incidence is still unclear. Our opinion is that an asymptomatic patient with CCAM should be clinically monitored and operated after the first year of life and without symptoms of the respiratory tract to prevent complications such as lung infections, pneumothorax and questionable malignancy Young Tae Kim et al. decided to perform an operation if the patient showed any signs of respiratory distress or there was any evidence of the lesion progressing to develop symptoms. Among five emergency cases which they had, only one patient suffered dyspnea and the remaining four were operated on an urgent basis as the lesion was too big or had increased in size, even though patients exhibited no symptoms [15].

The extent of surgical resection is also controversial, but since most lesions are limited to the lobe, lobectomy is the mainstay surgical method. A more conservative policy of segmentectomy, when possible, seems to have been employed by some investigators [16-18]. Lobectomy is the procedure of choice for patients with CCAM, especially when the lesion is confined to a single lobe. Infants and children tolerate lobectomy well, with compensatory lung growth, so that the total lung volume and gas exchange capacity return toward normal during somatic maturation [19,20]. Our opinion is that this conservative surgery has several disadvantages, such as persistent air-leak or incomplete excision of the lesion. That is why we always apply lobectomy. Only when multiple lobes are affected in patients with CCAM, then extensive resection should be avoided and performed parenchyma-saving procedure.

Conclusions

- CCAM neonatal patient with signs of respiratory distress, need an emergency operation.
- CCAM neonatal asymptomatic patient with the chest CT which reveals a large lesion with mediastinal shifting need urgent operation.
- Lobectomy is the procedure of choice in neonatal patients with CCAM

References

1. Kanavi JV, Thangaraj J, Thomas A (2017) Congenital cystic adenomatoid malformation (CCAM): antenatal and postnatal management *Int J Reprod Contracept Obstet Gynecol.* 6(6): 2447-2451.
2. Singh J (2011) Congenital Cystic Adenomatous Malformation: A Case Report. *JIMSA* 24(2).
3. Carias MJB, Orillaza M (2007) Congenital Cystic Adenomatoid Malformation Type II with Associated Cardiac Anomalies. Accepted paper for PHC 15th Annual research Paper Competition 2007 and for 38th PHA Annual Convention held on May 16-18, 2007 at Edsa Shangrila Hotel, Philippines 13(2): 161-167.
4. David M, Lamas-Pinheiro R, Henriques-Coelho T (2016) Prenatal and Postnatal Management of Congenital Pulmonary Airway Malformation. *Neonatology* 110(2): 101-115.
5. Adzick NS (2003) Management of fetal lung lesions. *Clin perinatol* 30(3): 481-492.
6. Vu L, Tsao K, Lee M, Nobuhara K, Farmer D, et al. (2007) Characteristics of congenital cystic adenomatoid malformations associated with non-immune hydrops and outcome. *J Pediatr Surg* 42(8): 1351-1356.
7. Kotecha S, Barbato A, Bush A, Claus F, Davenport M, et al. (2012) Antenatal and postnatal management of congenital cystic adenomatoid malformation. *Paediatr Respir Rev* 13(3): 162-171.
8. Breysse L, Bosmans H, Dymarkowski S, Schoubroeck DV, Witters I, et al. (2003) The value of fast MR imaging as an adjunct to ultrasound in prenatal diagnosis. *Eur Radiol* 13(7): 1538-1548.
9. Tsai AY, Liechty KW, Hedrick HL, Bebbington M, Wilson RD, et al. (2008) Outcomes after postnatal resection of prenatally diagnosed asymptomatic cystic lung lesions. *J Pediatr Surg* 43(3): 513-517.
10. Farrugia MK, Raza SA, Gould S, Lakhoo K (2008) Congenital lung lesions: classification and concordance of radiological appearance and surgical pathology. *Pediatr Surg Int* 24(9): 987-991.
11. Raychaudhuri P, Pasupati A, James A, Whitehead B, Kumar R (2011) Prospective study of antenatally diagnosed congenital cystic adenomatoid malformations. *Pediatr Surg Int* 27(11): 1159-1164.
12. Ankermann T, Oppermann HC, Engler S, Leuschner I, von Kaisenberg CS (2004) Congenital masses of the lung, cystic adenomatoid malformation versus congenital lobar emphysema. *J Ultrasound Med* 23(10): 1379-1384.
13. Miller JA, Corteville JE, Langer JC (1996) Congenital cystic adenomatoid malformation in the fetus: natural history and predictors of outcome. *J Pediatr Surg* 31(6): 805-808.
14. Aziz D, Langer JC, Tuuha SE, Ryan G, Ein SH, et al. (2004) Perinatally diagnosed asymptomatic congenital cystic adenomatoid malformation: to resect or not? *J Pediatr Surg* 39(3): 329-334.
15. Kim YT, Kim JS, Park JD, Kang CH, Sung SW, et al. (2005) Treatment of congenital cystic adenomatoid malformation—does resection in the early postnatal

- period increase surgical risk? *European Journal of Cardio-thoracic Surgery* 27 (4): 658-661.
16. Sapin E, Lejeune V, Barbet JP, Carricaburu E, Lewin F, et al. (1997) Congenital adenomatoid disease of the lung: prenatal diagnosis and perinatal management. *Pediatr Surg Int* 12(2-3): 126-129.
 17. Waszak P, Claris O, Lapillonne A, Picaud JC, Basson E, et al. (1999) Cystic adenomatoid malformation of the lung: neonatal management of 21 cases. *Pediatr Surg Int* 15(5-6): 326-331.
 18. Kim HW, Choi YS, Kim K, Shim YM, Ku GW, et al. (2008) Treatment of Congenital Cystic Adenomatoid Malformation: Should Lobectomy Always Be Performed? *Ann Thorac Surg* 86(1): 249-253.
 19. Sauvat F, Michel J, Benachi A, Emond S, Revillon Y(2003) Management of asymptomatic neonatal cystic adenomatoid malformations. *J Pediatr Surg* 38(4): 548-552.
 20. Zach MS, Eber E (2001) Adult outcome of congenital lower respiratory tract malformations. *Thorax* 56(1): 65-72.

