Case Report

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Late Onset Congenital Cystic Adenomatoid Pulmonary Malformation

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ABSTRACT

Congenital cystic adenomatoid pulmonary malformations are defined as the presence of a cystic mass of undifferentiated lung tissue; a third of the cases affect one of the lower lobes, and multilobar or bilateral affection is rare. The entity is a rare congenital malformation in the lungs, with an incidence of one in 11,000 to one in 35,000 live births. It is mainly discovered during the fetal period, although a low percentage of patients will be diagnosed postnatally. The disease has a wide spectrum of clinical manifestations, from asymptomatic to life-threatening respiratory symptoms. The diagnosis is made by imaging studies, and the treatment depends on the patient's clinical status. We reported the case of an infant with a rare congenital pulmonary airway malformation and a late diagnosis, which makes the case even rarer, who required open lobectomy as treatment.

Keywords: Congenital abnormalities; cystic adenomatoid malformation of lung; lung diseases.

INTRODUCTION

Congenital Cystic Adenomatoid Pulmonary Malformations (CCAPM) is defined as the presence of a cystic mass of undifferentiated lung tissue.[1,2] It has been associated with an abnormal branching of the bronchioles during morphogenesis, predominantly in the pseudoglandular stage between five and 16 weeks of pregnancy.[3] A third of the cases affect one of the lower lobes, and multilobar or bilateral affection is rare.[2] Imaging studies in utero or postnatally are required for diagnosis.[4] Preventive resection aims to reduce the risk of pneumonia and lung abscess.[2] We reported the case of an infant with late-onset CCAPM, who required surgical treatment.

CASE REPORT

A three-month-old male from a rural area with a medical history of infantile colic and bronchiolitis, which occurred at one month old, at which time a chest x-ray was taken with no evidence of alterations of any kind consulted with her mother for poor weight gain associated with a persistent cough. Vital signs with a heart rate of 151 beats per minute (bpm), breath rate of 34 breaths per minute (bpm), oxygen saturation of 98%, temperature of 36.3 C, weight of 4.1 kilograms, height of 58 centimetres, and weight-for-height Z-score compatible with severe acute malnutrition. On physician examination, she had wheezing in both lung fields and intercostal retractions.

Initial laboratories reported anemia (hemoglobin 8.4 g/dL, NR: 10.7 to 17.1 g/dL, hematocrit 25.7%, NR: 33 to 55%), without leukocytosis of 14500 mmol/L (NR: 5000 to 19500 mmol/L), and a high platelet count of 840000/mm3 (NR: 150000 to 450000/mm3). Normal results of glucose (99.5 mg/dL, NR: 65 to 99 mg/dL) and renal function (creatinine 0.35 mg/dL, NR: 0.2 to 0.4 mg/dL), mild hyponatremia (133.4 mmol/L, NR: 135 to 145 mmol/L), normal potassium (4.35 mmol/L, NR: 3.5 to 4.5 mmol/L), normal albumin (4.26 g/dL, NR: 3.4 to 5.4 g/dL), elevated ferritin (748.6 ng/mL, NR: 50-200 ng/mL), and total proteins in normal ranges (7.04 g/dL, NR: 6.0 to 8.3 g/dL).

Chest radiography suggested a left diaphragmatic hernia (Figure 1A). Due to these initial findings, the pediatrician in charge decided to perform advanced imaging studies to characterize this finding and therefore requested a chest computed tomography (CT) that reported a right basal consolidation focus, right pneumatocele, multiple thin-walled cystic lesions in the lingula, and a left lower lobe, the largest of 43 x 55 cm, suggestive of congenital pulmonary airway malformation (Figure 2). The diagnosis of CCAPM was made, with an important delay making the case even rarer; the patient was transferred to an

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institution where a pediatric surgeon was available. In that first period in a hospital of higher level of complexity, he required a hospital stay of 20 days for oxygen weaning, with a stay in the pediatric intermediate care unit. He was discharged without the need for home oxygen with the intention of deferring the surgical procedure in case no new symptoms would occur. Unfortunately, the patient continued with respiratory symptoms and five months after the first medical consultation and with eight months of life, he was to be taken to surgical management with a left lower lobectomy, a left closed decortication and a left closed thoracostomy performed without complications (Figure 1B). He continued in the hospital, staying in a pediatric intensive care unit for three days and continued his hospital stay for recovery for a total of 20 days since the first surgical procedure and he was discharged without any further requirements. The latest medical appointment, two months after the surgical procedure, reported that the patient improved his respiratory symptoms, although he presented complex convulsive episodes with no associated etiology, which is why he is currently in treatment with anticonvulsants.

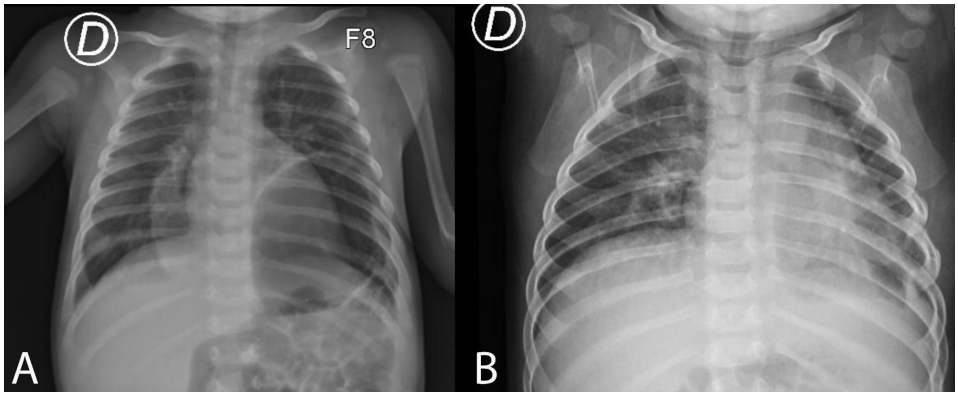

Figure 1. Chest X-rays in anteroposterior projection show in 1A an image suggestive of gastric chamber occupation in the left hemithorax compatible in the first instance with diaphragmatic hernia, and in 2B a control image is appreciated after a surgical procedure with the resolution of the CCAPM.

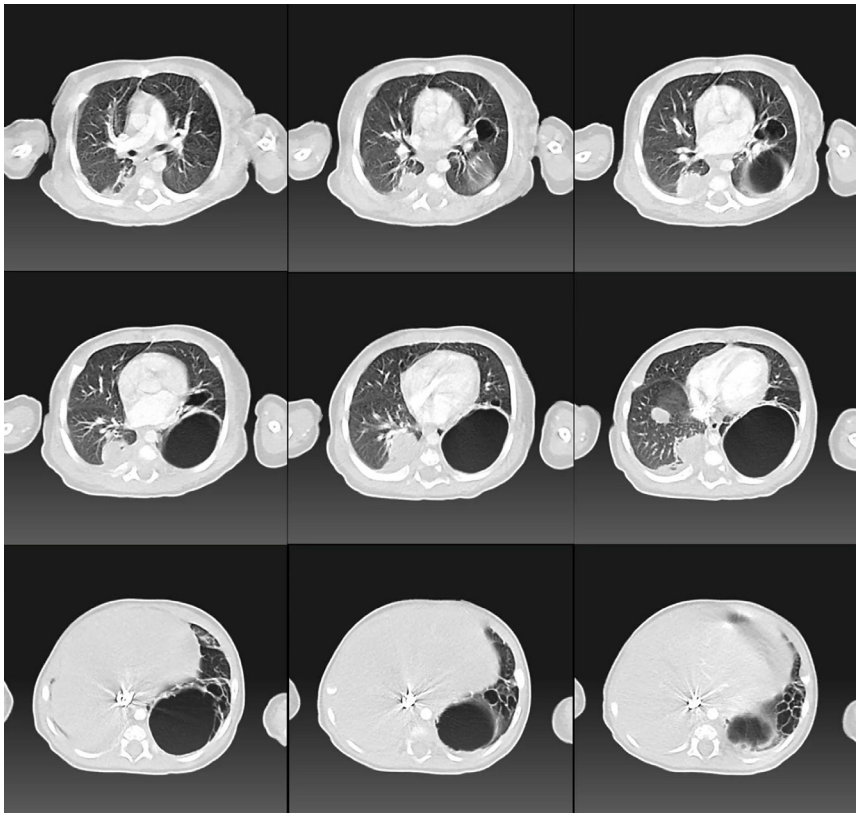

Figure 2. Chest computed tomography with contrast medium in axial slices showing CCAPM.

DISCUSSION

Despite CCAPM being the most common type of lung malformation, it has an incidence of one in 11,000 to one in 35,000 live births, predominantly males.[2,3] Classically, these entities had a histological classification into three groups: type I macrocystic (2–10 cm), type II cystic (up to 2 cm) and type III microcystic (less than 0.5 cm).[5] Posteriorly, Adzick proposes to classify them only into microcystic and macrocystic according to prenatal ultrasound.[2] The disease has a huge spectrum of clinical manifestations, from asymptomatic to life-threatening respiratory symptoms. Approximately 75% of the neonates are asymptomatic; the average age of initial symptoms is seven months; some of the symptoms include infections, chronic coughs, and recurrent wheezes.[6] The majority of pulmonary diseases are unilateral and restricted to the lower lobe, being rare on the lower left lobe.[6] Additionally, it can coexist with other congenital defects (up to 26%) in different systems, such as diaphragmatic, cardiovascular, gastrointestinal, and osteomuscular.[4,6,7] Our patient presented respiratory symptomatology at the month of birth with a persistent cough, which should have alerted the medical group to expand studies.

While the diagnosis can be made before delivery, there are instances where it is not detected until after the baby is born. Chest radiography has limited sensitivity, so it is preferable to utilize CT scans for confirming diagnoses. In situations where information about arterial supply and venous drainage is necessary, a CT scan with contrast is beneficial.[5] In our case, the initial chest radiography cannot evidence the cystic lesion that would be found a month later. The second chest radiography was inconclusive, suggesting a diaphragmatic hernia; nevertheless, the chest CT scan may accurately describe the abnormality.

There is no consensus on the management of this entity; asymptomatic patients may continue under strict observation, and in cases of symptomatology, surgical management would be required,[8] which also helps to prevent complications such as abscess, empyema, recurrent pneumonia, air leak, pneumothorax, and malignancy that is primarily associated with bronchoalveolar carcinoma, sarcoma, or blastoma.[7,9] The surgical options include lung-sparing resections (LSR), for instance, the wedge section or non-anatomic resections, which theoretically have fewer complications.[10,11] The use of complete resection procedures such as lobectomy has been the traditional surgical strategy in congenital pulmonary airway malformations.[10] Lobectomy can be performed in an open approach or by thoracoscopy; the latter is preferred due to the better aesthetic results and low complication rate[12]; however, it is feasible in those weighing more than 10 kilograms or older than 18 months.[10] Our patient underwent an open lobectomy, considering the location of the thin-walled cystic lesions, size, and age, with favorable evolution and outcomes. Notice that it is necessary to perform genetic studies to rule out other possible associated pathologies.

CONCLUSIONS

CCAPM is a rare congenital malformation. The majority of patients are asymptomatic. Postnatal imaging studies are necessary for the diagnosis. The patient's clinical status determines the treatment; asymptomatic patients may opt for observation. Surgical strategy depends on the patient's age, clinical condition, location, and the size of the lesions.

CONFLICT OF INTERESTS

The authors declare no conflict of interests.

REFERENCES

1. Garzi A, Ferrentino U, Ardimento G, Brongo S, Rubino MS, Calabrò E, et al. Congenital Cystic Adenomatoid Malformation of the Lung Type II: Three Cases Report. Transl Med UniSa. 2019;20:4-8. PMID: 31850245

2. Kunisaki SM. Narrative review of congenital lung lesions. Transl Pediatr. 2021;10(5):1418-1431.doi: https://doi.org/10.21037/tp-20-133

3. Gosain R, Motwani R. Congenital cystic adenomatoid malformation of the lung: a case report with embryological review. Anat Cell Biol. 2022;55(2):264-268. doi: https://doi.org/10.5115/acb.21.260

4. Annunziata F, Bush A, Borgia F, Raimondi F, Montella S, Poeta M, et al. Congenital Lung Malformations: Unresolved Issues and Unanswered Questions. Front Pediatr. 2019;7:239.doi: https://doi.org/10.3389/fped.2019.00239

5. Thimmesch M, Khamis J, Senterre G, Dôme F, Dirix M. L□image du mois. Malformation adénomatoïde kystique congénitale [Congenital cystic adenomatoid malformation]. Rev Med Liege. 2021;76(2):69-70. PMID: 33543849.

6. Salerno M, Sessa F, Cocimano G, Roccuzzo S, Esposito M, Pomara C. Congenital Cystic Adenomatoid Malformation (CCAM) Type II: A Rare Case of Sudden Infant Death. Children (Basel). 2022;9(12):1830. doi: https://doi.org/10.3390/children9121830

7. Badour M, Hussain B, Hammed A, Sawssan Ali, Falyon S. A rare case of congenital cystic adenomatoid malformation: Mimics pneumonia manifestations. Ann Med Surg (Lond). 2021;66:102433.doi: https://doi.org/10.1016/j.amsu.2021.102433

8. Sime H, Abera G, Mengistu A, Lamessa S. Congenital Pulmonary Airway Malformation (CPAM): A Case Report, Jimma University Medical Center, Southwest Ethiopia. Ethiop J Health Sci. 2021;31(4):907-910. doi: https://doi.org/10.4314/ejhs.v31i4.27

9. Brucknerová J, Babala J, Ujházy E, Mach M, Juránek I, Brucknerová I. Rare causes of respiratory insufficiency in newborns. Physiol Res. 2020;69(Suppl 4):S637-S647. doi: https://doi.org/10.33549/physiolres.934613

10. Bhende VV, Sharma TS, Krishnakumar M, Shah DM, Pankhaniya RN, Parmar ZN, et al. Surgical Management of Congenital Pulmonary Airway Malformations (CPAM) in an infant and a toddler: case report depicting two distinct surgical techniques with successful outcomes. Cureus. 2024 Feb 3;16(2):e53526. doi: http://dx.doi.org/10.7759/cureus.53526

11. Zeng Z, Liu C, Liu C, Pu Q, Mei J, Liao H, et al. Clinical characteristics and surgical treatment of congenital cystic adenomatoid malformation in adults: the largest cohort of 46 patients. Ann Transl Med. 2022 May 1;10(10):596. doi: http://dx.doi.org/10.21037/atm-22-1747

12. Khan H, Kurup M, Saikia S, Desai A, Mathew M, Sheikh A, et al. Morbidity after thoracoscopic resection of congenital pulmonary airway malformations (CPAM): single center experience over a decade. Pediatr Surg Int. 2021;37(5):549-554. doi: https://doi.org/10.1007/s00383-020-04801-1