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Congenital Cystic Adenomatoid Malformation: A Case Report with Clinical, Radiological, Histological, and Surgical Features

Karim Lahrache[1,2*], Samia Malki[1,3], Soulaimane M'harzi[1,2], Anas Ayyad[1,2], Sahar Messouadi[1,2], Houssain Benhaddou[1,4], Amal Bennani[1,3], Rim Amrani[1,2]

[1]Faculty of Medicine and Pharmacy of Oujda, Mohammed I University of Oujda, Oujda, Morocco
[2]Department of Neonatology and Neonatal Reanimation, Mohammed VI University Hospital of Oujda, Oujda, Morocco
[3]Anatomopathology Department, Mohammed VI University Hospital of Oujda, Oujda, Morocco
[4]Paediatric Surgery Department, Mohammed VI University Hospital of Oujda, Oujda, Morocco
Email: *ka.lahrache@gmail.com

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Abstract

Background: Congenital cystic adenomatoid malformation (CCAM) is a congenital anomaly of lung development, accounting for approximately 25% of congenital lung lesions. Respiratory distress often occurs during the neonatal period, and in 80% to 85% of cases, the diagnosis is made before the age of 2 years following respiratory infections. **Case Report:** We report a case of MAKC diagnosed in the neonatal period. The diagnosis was based on clinical, radiological and histological elements. Our patient underwent surgical resection. Histological examination confirmed the diagnosis of MAKC without any sign of malignancy. The postoperative evolution was good. **Conclusion:** Clinicians and pathologists should recognize the early discovery of MAKC in neonatal age. The clinical diagnosis strongly guided by the radiological approach is confirmed by the pathological anatomy insofar as the therapeutic sanction is surgical in the majority of the cases.

Keywords

Congenital Cystic Adenomatoid Malformation, Congenital Disorder, Lung, Respiratory Distress, Newborn

1. Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare disease that accounts for approximately 25% of all pulmonary congenital malformations. It is due to an early cessation of bronchiolar maturation, which affects the channeling process of the distal lobular branches, beyond which the terminal respiratory

structures develop an adenomatoid appearance [1]. We use chest X-rays, which show images of aerated cysts, to make the diagnosis in postnatal. We can also use prenatal ultrasonography to diagnose it; it shows fluid cysts or a homogeneous hyperechogenic mass [2]. Historically, these have been divided by Stocker into 3 types:

Type I: large cyst (>2 cm). These consist of large sometimes multiple or multiloculated cysts. They are not true cysts and always communicate with the proximal airway and distal lung parenchyma. Histologically, they are lined with respiratory ciliated cuboidal or columnar Epithelium. *Type II*: small cyst (<2 cm). These consist of small uniform multiple or multilocular cysts. They are not true cysts and are also lined by ciliated columnar or cuboidal epithelium. *Type III*: solid lesion. These are macroscopically and microscopically solid lesions without cysts. Pathologically these are grouped into the pulmonary hyperplasia group.

We present a case of a newborn diagnosed with CCAM after experiencing respiratory distress.

2. Case Summary

A male full-term newborn who came from a non-consanguineous family; his mother was 37 years old; she had three miscarriages; the pregnancy was well followed, and the birth was by vaginal delivery. The APGAR score was 10 at the start of the first minute. From the third hour of life, respiratory distress occurs with a Silverman score of 2/10, requiring oxygen therapy.

Clinical examination revealed a pink, toned, reactive, suck reflex, as well as the rest of the archaic reflex. The newborn weighed 3.2 kg, measured 56 cm, and had a cranial girth of 36 cm. He was apyretic at 37˚C, with a heart rate of 140 beats per minute, a capillary refill time of less than 3 seconds, a blood pressure of 80/45 mmHg, a breathing rate of 54 cycles per minute, and a SaO_2 of 90% in ambient air and 97% under 2 L of oxygen.

Physical examination revealed normal pulmonary, cardiovascular, and abdominal exams, as well as no congenital anomalies.

The biological test reveals a normal complete blood count of 11,370/µL for white blood cells, 14.8 g/dL for hemoglobin, 249,000/µL for platelets, 7 mg/L for CRP, 387 for creatine kinase, 905 for LDH, and 98 mg/L for calcemia.

The radiological evaluation revealed a chest radiograph in favor of a dense and homogeneous lower left lobar opacity without an aeric bronchogram (**Figure 1**). The results of the chest scanner are shown in **Figure 2**.

The head ultrasound was normal, and echocardiography showed an atrial septal defect of 3 mm.

The newborn was operated on at 12 days of age for a lower left lung lobectomy (**Figure 3** and **Figure 4**). Surgical pathology examined the specimens removed during surgery.

The pulmonary parenchyma contains cystic cavities of varying sizes, surrounded by smooth muscle fibers and separated by regular alveoli, according to histological examination (**Figures 5-7**). The newborn is returned to his family

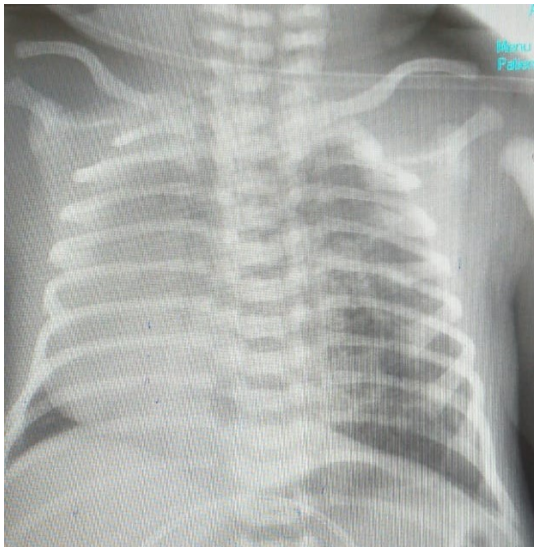

Figure 1. Chest radiograph in favor of a dense and homogeneous lower left lobar opacity without an aeric bronchogram.

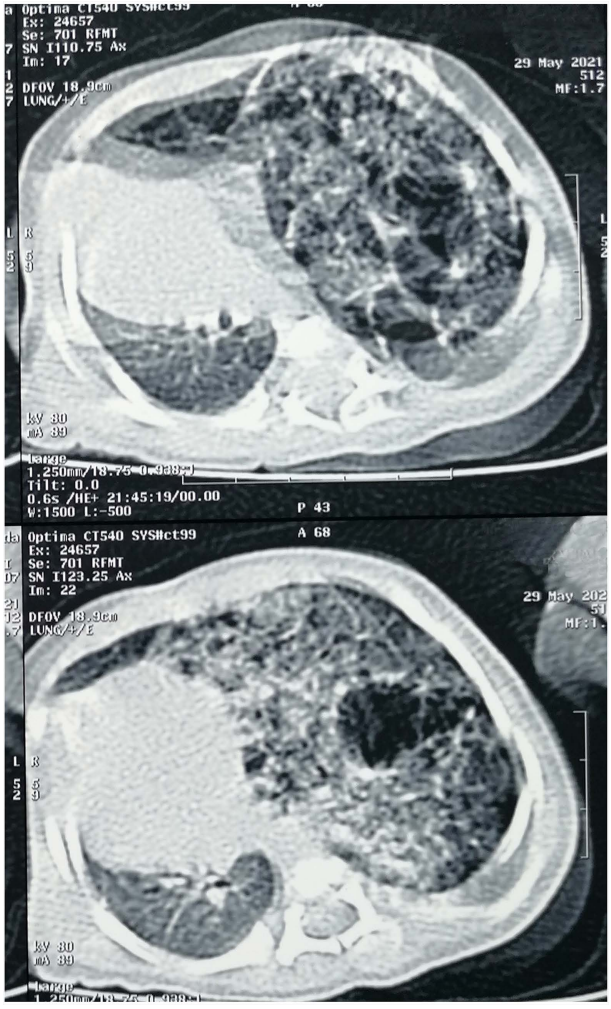

Figure 2. CT scan reveals several aerated cysts that are separated from the healthy lung parenchyma and linked to the bronchi.

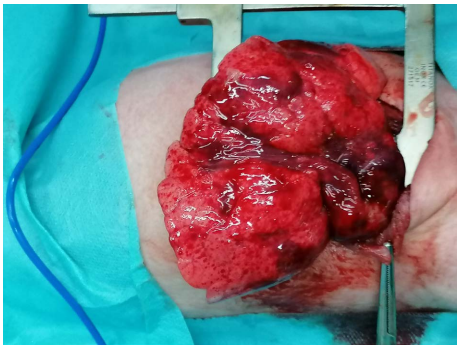

Figure 3. Adenomatoid cystic malformation at the time of surgery.

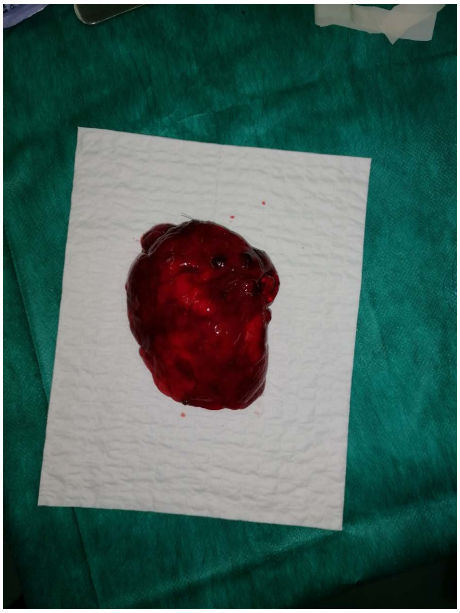

Figure 4. Adenomatoid cystic malformation after surgical removal.

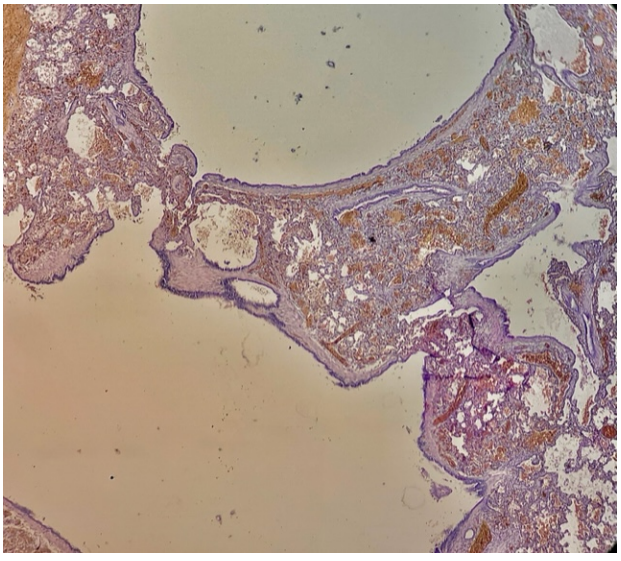

Figure 5. Microphotograph showing a lung parenchyma with cystic cavities of variable size.

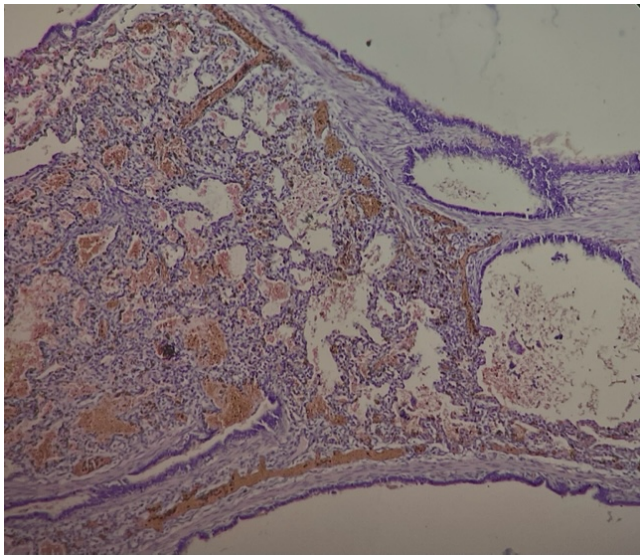

Figure 6. Cystic cavities are surrounded by smooth muscle fibers and separated by regular alveoli sometimes containing red blood cells.

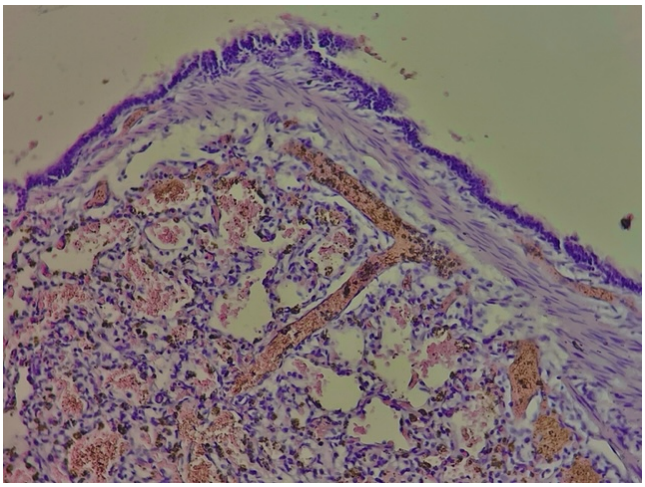

Figure 7. The cystic wall is lined by a simple ciliated cylindrical epithelium, in some places pseudostratified, surrounded by a thin layer of smooth muscle and resting on regular alveolar cavities sometimes hemorrhagic.

after 24 days. The three-month check-up reveals a healthy child, normal growth, a normal pulmonary exam, and a normal chest radiograph.

3. Discussion

Ch'in and Tang defined CCAM as an adenomatoid proliferation of the terminal respiratory structures in 1949 [3]. It is caused by the cessation of pulmonary airway maturation in the absence of alveolar tissue formation [4]. It is the most common type of fetal lung lesion, accounting for one-quarter of all fetal lung lesions. Males are the most affected, and no racial or genetic predisposition has been identified. CCAM affects only one lobe in 80% - 95% of cases, with a preference for the right lung [3].

In fact, CCAM can come in a variety of sizes and presentations. The Stocker classification, proposed in 1977, is the most widely used. Thus, the most common type, Type I, is distinguished by the presence of a small number of voluminous cysts with diameters greater than 2 cm. Type II consists of numerous cysts that are no larger than 1.5 cm in diameter. The presence of numerous small cysts measuring less than 0.5 cm distinguishes Type III [1].

Almost all cases of CCAM are diagnosed within the first two years of life, thanks to prenatal ultrasound imaging [5]. From the 20th SA, antenatal ultrasound reveals a lung mass composed of several anechoic images of varying sizes [6]. Hydramnios is seen in 60% to 65% of cases [7]. Although antenatal ultrasound can detect pulmonary malformations, it cannot determine the specific type [8].

CCAM can cause respiratory distress in newborns [8]. After a few days, dyspnea and cyanosis appear. It causes progressive dyspnea or recurrent lung infections in children. Some cases are asymptomatic and will not manifest until adulthood [9].

The first imaging technique used to diagnose bronchopulmonary malformations was chest radiography [10]. CT scanning allows for the confirmation of the diagnosis, assessment of the extent of the lesion and its anatomical reports, elimination of a differential diagnosis, and discussion of surgical procedures [11] [12].

A CT scan reveals several aerated cysts that are separated from the healthy lung parenchyma and linked to the bronchi. When making a differential diagnosis, CT angiography is helpful in searching for systemic vascularization that might support pulmonary sequestration. The interest in magnetic resonance imaging (MRI) is negligible. The diagnosis is confirmed by a histology analysis. It reveals cysts with fluid or aerated contents, restricted by a cartilage-free wall, devoid of inflammatory signs, and lined by mucous cells and ciliated pseudostratified columnar epithelia [13].

The CCAM must be surgically removed if they are symptomatic. Small CCAM only needs close monitoring if there are no symptoms [14]. However, for a number of reasons—including preventing potential problems such as lung infections and breathing difficulties and lowering radiation risks because of CT scan controls—we advise, like many authors [14] [15], conducting the surgery even if the CCAM is asymptomatic.

4. Conclusion

To detect CCAM, we use antenatal ultrasound. An early diagnosis results in a better surgical cure and helps to avoid potential complications. The possibility of recurrence in the event of insufficient excision justifies long-term surveillance of patients with this malformation.

Patient Consent

The patient's parents were contacted to explain our goal of sharing their child's case for scientific purposes.

Conflicts of Interest

All authors declare that they have no conflicts of interest.

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