Congenital Cystic Adenomatoid Malformation of the lung : Two cases report

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Malformation adénomatoide kystique congénitale du poumon : deux nouvelles observations

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RÉSUMÉ

Prérequis : La malformation adénomatoide kystique du poumon (MAKP) est une anomalie congénitale rare avec une incidence de 1 /25000 à 1/35000 naissances. Le diagnostic des MAKP peut se faire en anténatal par l'échographie ou en postnatal suite à des signes de détresse respiratoire. Dans les cas asymptomatiques le diagnostic se fait à un âge plus tardif à l'occasion d'une radiographie systématique.
But : Rapporter deux cas de MAKP différents par leurs circonstances de découverte.
Observations : Le premier nouveau-né était de sexe masculin, à terme, dont le diagnostic de MAKP à été suspecté en anténatal par l'échographie faite à 29 semaines d'aménorrhée (SA). Le nouveau-né a présenté une détresse respiratoire néonatale immédiate. En postnatal la radiographie et la tomodensitométrie ont confirmé le diagnostic de MAKP.
Le deuxième nouveau-né également de sexe masculin était né à un terme de 30 SA 5 jours issu d'une grossesse triple induite et dont le diagnostic de MAKP était posé en postnatal devant une détresse respiratoire néonatale compliquée de pneumothorax et l'aspect radiologique caractéristique.
Conclusion : Les deux cas décrits rendent compte de la disparité clinique des MAKP. Actuellement, le diagnostic se fait en anténatal permettant une prise en charge optimale.

Mots-clés

Echographie anténatale, malformation adénomatoide kystique du poumon, nouveau-né

SUMMARY

Background: Congenital cystic adenomatoid malformation (CCAM) is a rare embryonic developmental abnormality with an incidence of one in 25 000 to 35 000 pregnancies. With advances in antenatal ultrasonographia (USG), CCAM has been increasingly diagnosed. After birth, the clinical appearance of CCAM can vary from immediately postnatal respiratory distress, to an incidental finding on chest radiography.
Aim: To report two additional cases with CCAM different in clinical features.
Case reports: The first case was a boy in witch diagnosis was suspected by antenatal USG; he was born at 37 weeks of gestation by cesarean section because of severe toxemia, and presented immediately respiratory distress. The chest x ray and computed tomography scan (CT scan) revealed a right CCAM.
The second case was also a boy of an inducted threefold pregnancy, born at 30 weeks by cesarean section. He presented immediately respiratory distress. The diagnosis of a right CCAM was confirmed by clinical and radiological findings in postnatal period.
Conclusion: The two cases described in this report show the disparity in clinical features of CCAM. Nowadays, antenatal echo-tomography can establish the diagnosis allowing optimum management.

Key-words

Antenatal ultrasonography, congenital cystic adenomatoid malformation, newborn

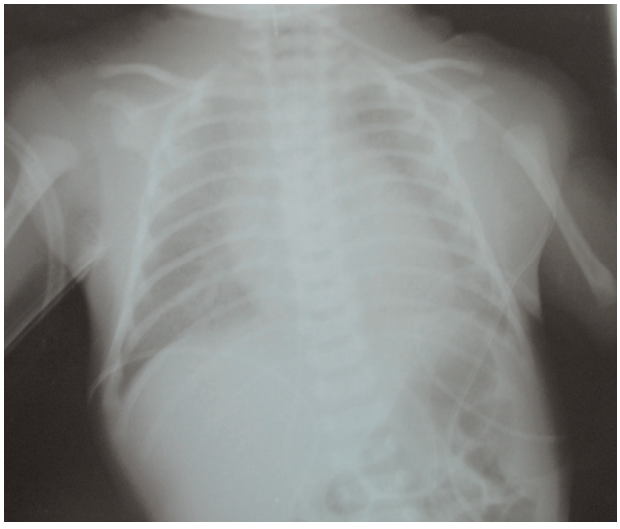
Congenital cystic adenomatoid malformation (CCAM) was first described by Ch'in and Tang in 1949 [1]. CCAM is a rare anomaly, the incidence was estimated at 1 per 25000 to 35000 pregnancies, it accounts for 25% of all congenital lung malformations and most commonly presents with respiratory distress in newborns [2]. These lesions are now often diagnosed on prenatal sonographic, a minority of cases may not be identified by prenatal imaging techniques and the pulmonary lesions are recognized postnatally [3].
We report two cases different by their moment of diagnosis.

OBSERVATIONS

Case 1

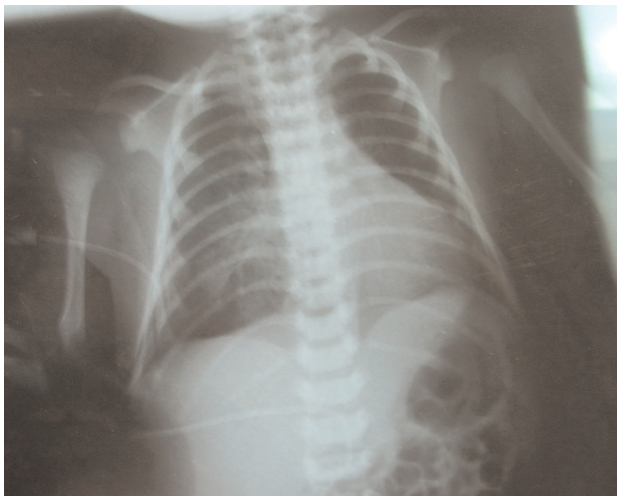
A one month old male issued from healthy parents was born at 37 weeks of gestation by elective cesarean section because of severe toxemia and scarred womb. The pregnancy was complicated with gestational diabetes; the antenatal ultrasonographia at 29 weeks showed multiples small cysts, the greater was measured 15 mm in favor of a micro cyst form d'Adziak in the right lung. No others fetal anomalies were detected. After delivery, the patient had a good Apgar score, a birth weight of 3450g (50th centile), and immediately respiratory distress with increasing oxygen need. He was admitted in intensive unit care at 18 hours of life, ventilated with intermittent conventional ventilation, then inhaled NO for 43 hours. The first chest radiographs showed micro nodular opacities in favor of respiratory distress syndrome (Figure 1).

Figure 1 : First chest radiography: micro nodular opacities of the two lungs.



He had been weaned from the ventilator at 67 hours of life. The CT scan showed a large cystic lesion with multiples small cysts in the right lower lobe of the lung. Controlled chest radiography showed a heterogeneous apical opacity in the right lung (Figure 2). As the patient was asymptomatic, the surgeon suggested a regular clinic and radiologic follow up until about 9 months.

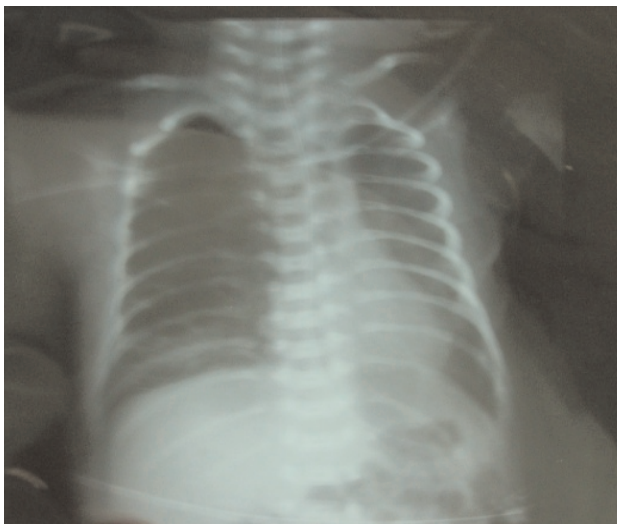
Figure 2 : Controlled chest radiography: heterogeneous opacity in the lower lobe of the right lung.



Case 2

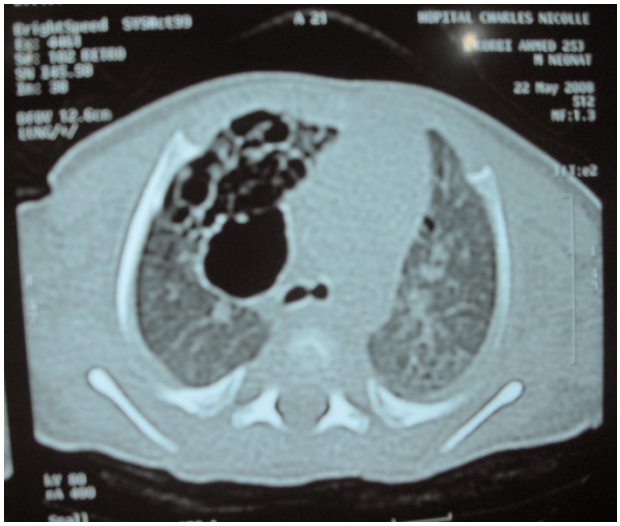
A three weeks old male issued from an inducted threefold pregnancy, the mother is thirty three years old with no evident history of pulmonary disease in the family, antenatal ultrasonography (USG) was normal. The babies were delivered prematurely at 30 weeks of gestation by cesarean section because of severe toxemia, prematurity and threefold pregnancy. Our patient was the second born of the triplets; he had good Apgar score, a birth weight of 1750g (50th centile). He developed immediately a respiratory distress so he was admitted in intensive care unit, intubated and ventilated for 14 days and had to be drained twice for a recurring air leak. He had been weaned from the ventilator at fourteen days of life. The chest radiographs (CXR) showed a large cystic lesion in the total right upper lobe of the lung (Figure 3).

Figure 3 : Chest radiographs (CXR): large cystic lesion in the right upper lobe of the lung



A computed tomography (CT) scan showed in the right lobe, small cysts filled with air (Figure 4).
The patient was addressed to a surgery team at the age of two months asymptomatic. A regular clinic and radiologic follow up was determined.

Figure 4 : CT scan showed in the right lobe, small cysts filled with air



DISCUSSION

CCAM is a rare fetal pulmonary lesion; it is thought to be due to overgrowth of terminal bronchioles that form cysts of various sizes with suppression of alveolar development ensuing between 5 and 8 weeks of gestation. The lesion communicates with the bronchial tree and derives its vascular supply from the pulmonary circulation [2].
In 1977, Stocker and al classified three types of CCAM: Type I consists of large cysts; type II consists of small cysts; and type III shows lesions resembling a homogeneous mass, with cysts only seen on microscopy [4]. Our two cases had probably a type II lesion based only on radiological features.
Diagnosis can be made in antenatal period by USG and CCAM is classified according to Adzick's classification based on sonographic features presence or absence of polyhydramnios, mediastinal shift and hydrops [5]. Various USG features have been associated with a poor prognosis, including the type and size of CCAM, presence of mediastinal shift and hydropic change [6]. We could classify the case one based on antenatal USG in type 2.
CCAM communicates with the bronchial tree at birth and therefore typically contains air soon after birth [7, 8]. The imaging appearance is determined by the size and number of cysts [9]. CCAM is observed with equal frequency in boys and girls, does not appear to be hereditary, the lesions are confined to one lobe and seem to be predominant in the lower lobes; they may or may not occupy the whole lobe [3, 9]. Our first patient had a particularity of upper localization less described in literature.
Clinical expressions of CCAM are ranging from death in utero (frequent in type III) to respiratory distress at birth (30%) as well as the absence or the presence of pulmonary symptoms. Pneumothorax and atelectasis may also occur as in our first patient who had recurring pneumothorax [7]. Antenatal diagnosis allows preparation for immediate surgery if necessary. In-utero regression was reported to occur in 6.3 to 55% of cases [10, 11, 12].
Late complications of CCAM include severe or recurrent infection of the lungs and malignant change in the lesion requiring early surgery recommended by the majority of authors [12, 13]. Overall postnatal mortality was reported to be 4 to 37% [14]. While postnatal spontaneous resolution is noted [15].
CCAM is the most common type of fetal thoracic mass diagnosed by prenatal ultrasonography. Identification of prognostic factors calls for antenatal counseling, possible fetal intervention, and preparation of perinatal management at tertiary care facilities.
The treatment is based on surgery, generally a lobectomy or a segmentectomy, rarely localized resections. There should be a long clinical and radiological follow-up.

CONCLUSIONS

The two cases described in this report show the disparity in features and moment of the diagnostic of CCAM. The malformation can cause death at birth, respiratory distress in the newborn or may be diagnosed at a later age. Nowadays, antenatal echo-tomography can establish the diagnosis allowing optimal management.

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