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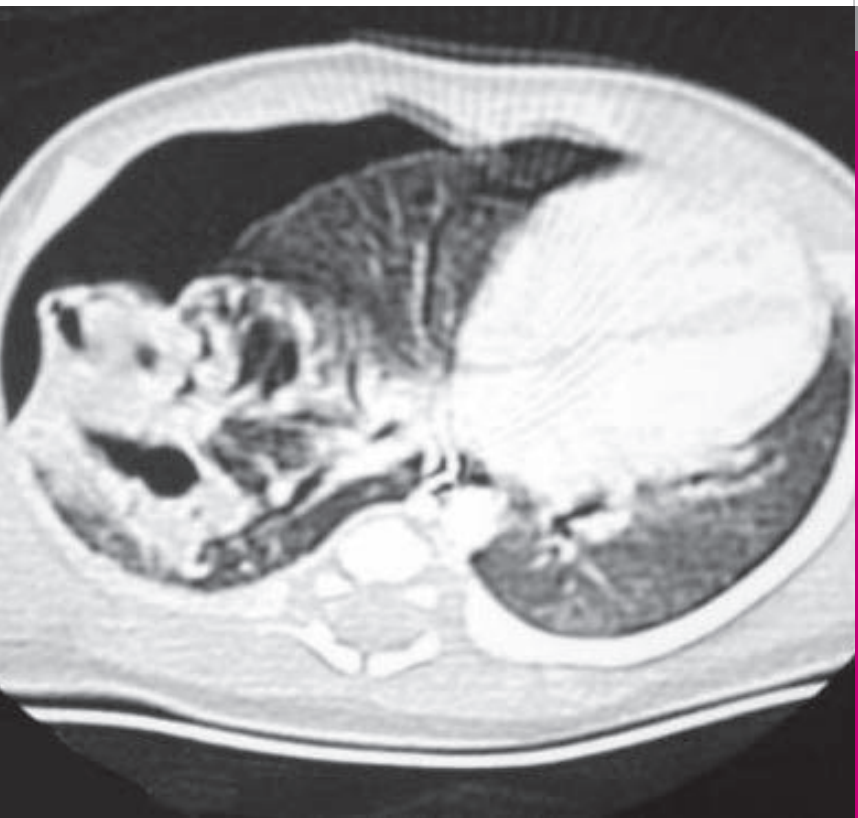
Congenital cystic adenomatoid malformation



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INTRODUCTION

Many congenital anomalies of the lung have been described. In the era of prenatal diagnosis and in-utero treatment, the pediatrician, neonatologist and intensivist are challenged by specific aspects in the management of babies suffering from these malformations, either in the pre-, peri- or postnatal periods.

CASE REPORT

During the second trimester of an otherwise uneventful pregnancy, a fetal ultrasound examination disclosed a cystic lesion of the right lung. Fetal MRI suggested the diagnosis of a cystic adenomatoid malformation of the lung (Fig. 1). At 33 weeks of gestation, a pigtail drain was placed into a large cyst of the right lower lobe because of a severe mass effect on the mediastinum and adjacent lung parenchyma. This resulted in a reduction in the size of this cyst and a reduction in the mediastinal shift. Other smaller cysts were detected on follow-up exams.

A female baby was born by elective Caesarean section during the 37th week of gestation. Birth weight was 2640 g (P 50-90), birth length 48.2 cm (P 50-90) and head circumference 34.5 cm (> P 90). Apgar scores were 4, 4 and 8 at 1, 5 and 10 minutes, respectively. Respiratory distress was apparent after a few minutes and the baby was intubated at 25 minutes of life.

On arrival in the ICU, respiratory sounds were more audible on the left than on the right side. A small thoracic drain (pigtail) was noticeable on the right hemithorax. Around it, the skin looked umbilicated and the hole was a tiny bit bigger than the drain.

A tension pneumothorax developed after 12 hours and required transcutaneous thoracic drainage. Chest X-ray revealed an inhomogeneous opacity of the lower part of the right lung (Fig. 2). A CT scan performed at 10 hours of life revealed a tension pneumothorax and an inhomogeneous mass with several cysts of varying sizes, located in the right pleural cavity (Fig. 3-6). The baby was extubated after 48 hours but required a bilevel non-invasive ventilation for seven additional days.

At one week of age, surgical excision of the right inferior pulmonary lobe was performed. The child was easily weaned from the ventilator on the second post-operative day. The baby left the ICU on postoperative day 5 with supplemental oxygen and a slight tachypnea.

Histopathology of the resected lobe was compatible with a cystic adenomatoid malformation of the lung, type II.

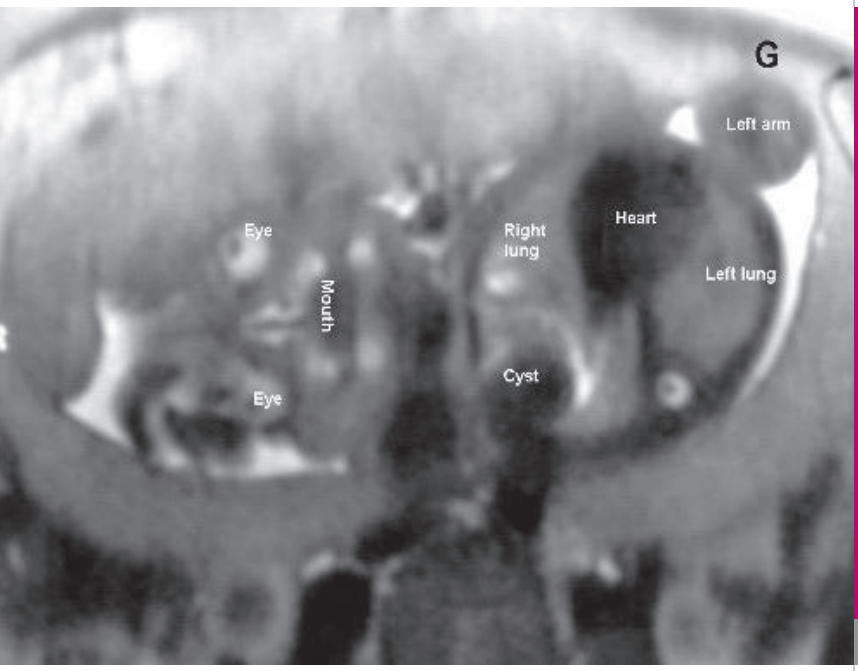

Fig. 1

Fetal MRI revealing right-sided cystic lung lesion.

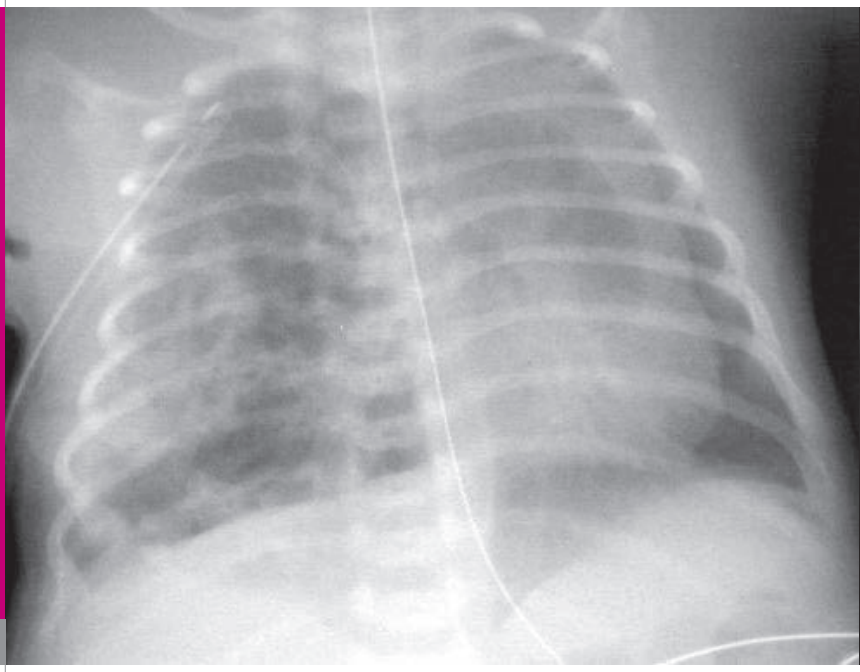

Fig. 2

Chest X-ray at 12 hours of age with residual right-sided pneumothorax.

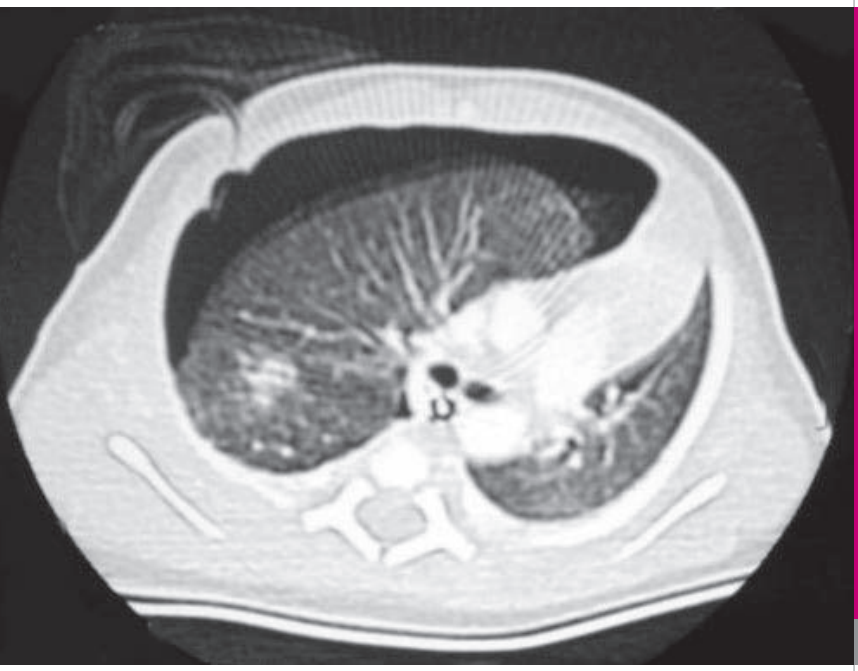

Fig. 3

Chest CT-scan on day 1 of life with right-sided tension pneumothorax.

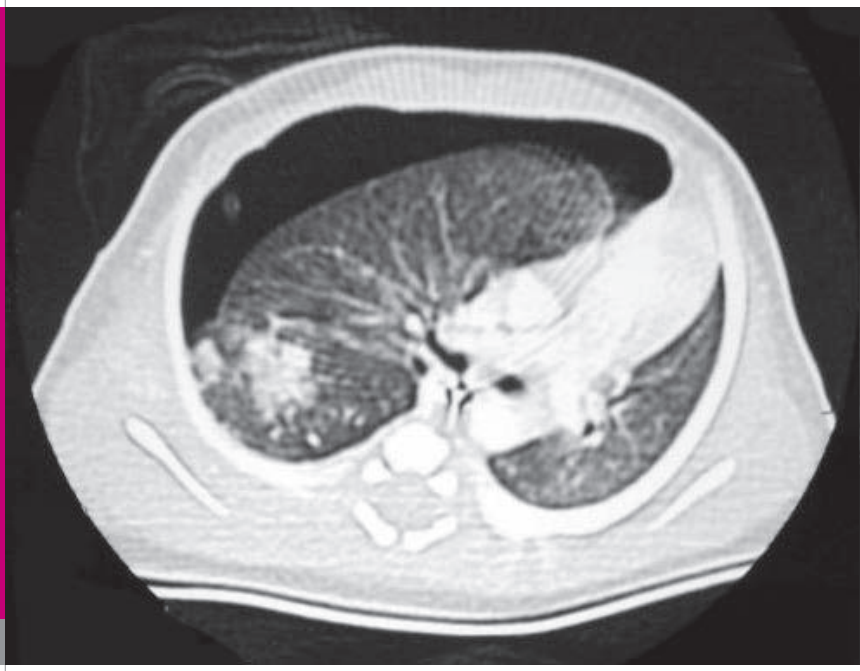

Fig. 4

Chest CT-scan on day 1 of life with right-sided tension pneumothorax.

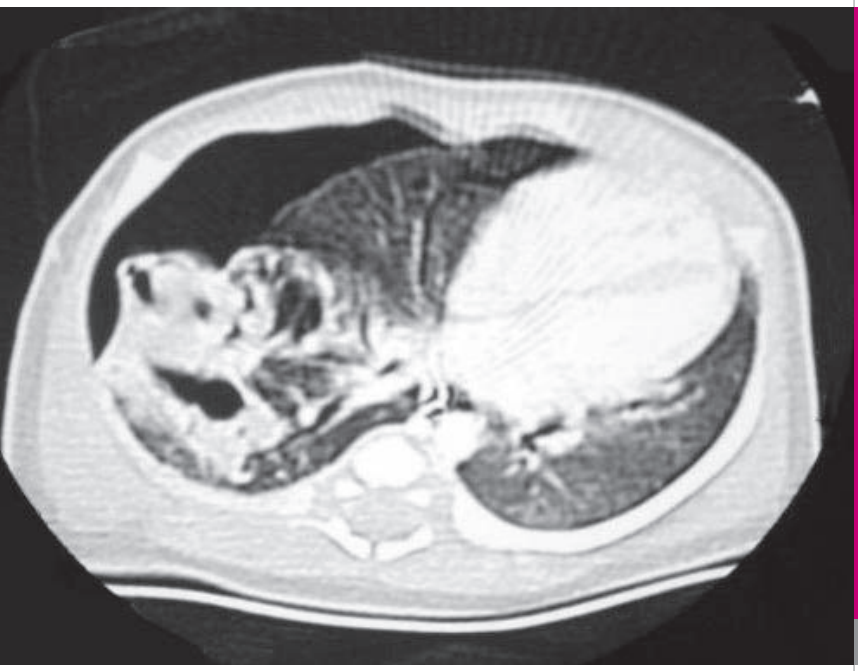

Fig. 5

Chest CT-scan on day 1 of life with abnormal lung parenchyma.

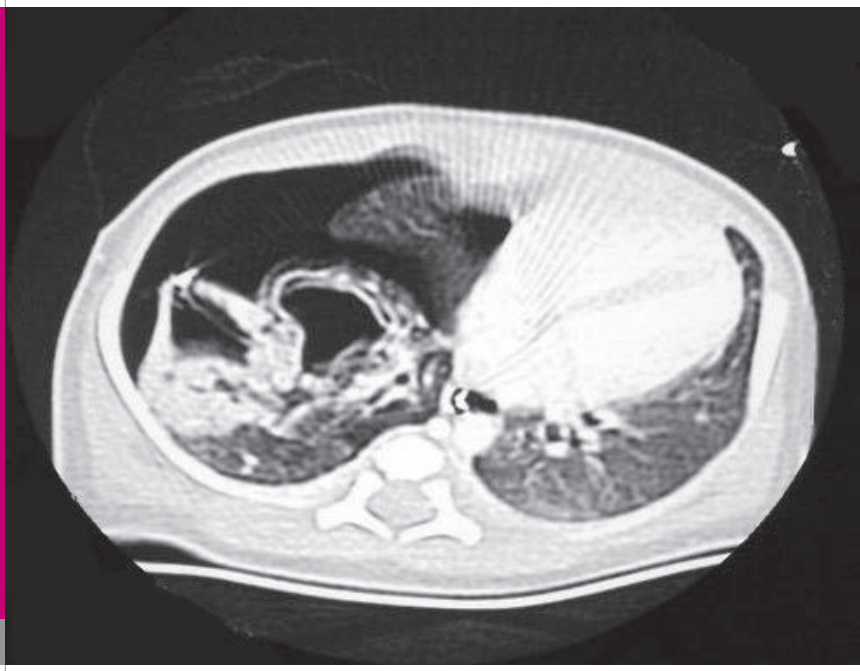

Fig. 6

Chest CT-scan on day 1 of life with abnormal lung parenchyma.

This case illustrates many aspects of congenital cystic adenomatoid malformation (CCAM). The prenatal diagnosis confirmed by a fetal MRI allowed early detection in utero of a potentially fatal complication (severe mediastinal shift with compression of adjacent and controlateral lung parenchyma). For this reason, drainage of the largest cyst with a pigtail device was performed in utero a couple of weeks before birth.

At birth, respiratory distress developed rapidly and required admission to an intensive care unit. The pigtail catheter was not removed initially, nor was it occluded. We were concerned by the possibility of a pneumothorax because of an air leak between the drained cyst and the pleural space; therefore, we decided initially to keep this pigtail in place. The pneumothorax which was obvious 12 hours after birth can be due to usual mechanisms, but inspection of the skin around the drain suggests that air could have entered the pleural space by flowing around the drain. However, it is also possible that the insertion of this pigtail resulted in a bronchopleural fistula.

CCAM was described for the first time by Ch'in and Tang in 1949. Since then, approximately 500 cases have been reported. The incidence is 1/25'000 to 1/35'000 pregnancies. It is the most frequent congenital anomaly of the lung and represents 25% of these conditions. Generally, only one pulmonary lobe is

abnormal, and most frequently CCAM is located on the right side. However, diffuse CCAM of both lungs have been described.

CCAM results from an anomaly of lung development occurring in the 6th to 7th week of gestation. Several pathogenic hypotheses are plausible (impairment of lung vascularization, infection, bronchial obstruction) but no single mechanism has been clearly identified.

Several classifications of CCAM have been proposed, based on Stocker's work. In Stocker's classification, type I represents 60 to 70 % of all CCAMs and is characterized by large cysts over 2 cm in diameter and lined by a pseudo-stratified epithelium with mucus cells. In type II, cysts are smaller (< 1 cm in diameter) and lined by a cuboid epithelium. Type III represents only 10% of all CCAMs and looks macroscopically like a solid mass, with small microscopic cysts.

Other congenital anomalies can be associated with CCAM, for example bilateral renal agenesis, sirenomelia, trisomy 18, congenital heart disease, anomalies of the digestive tract (labio-maxillo-palatine defects, esophageal cyst, intestinal malrotation, imperforate anus, Meckel's diverticulum), and vertebral anomalies.

The differential diagnosis of CCAM includes diaphragmatic hernia, lung sequestration, congenital lobar

emphysema, atresia of airways, bronchogenic cyst, mediastinal teratoma or pericardial cyst. The possible consequences of the development of CCAM in utero are pulmonary hypoplasia, polyhydramnios, foetal hydrops or fetal ascites.

In CCAM, risk factors for poor prognosis are type III, size of the lesion, involvement of both lungs, associated pulmonary hypoplasia, fetal hydrops, polyhydramnios or shift of the mediastinum. The natural history of CCAM depends on the size of the cysts, on their pathophysiological effects and on the size of the normal lung parenchyma. Overall, mortality in CCAM is reported to be 10%. Respiratory distress or pneumothorax in the newborn or recurrent pulmonary infections in the older child can be the presenting signs. Some CCAMs are fortuitously diagnosed on chest X-rays later in life.

ACKNOWLEDGEMENT

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