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Congenital Pulmonary Malformation: About a Case of Congenital Cystic Adenomatoid Malformation

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Abstract

Congenital pulmonary malformations result from developmental abnormalities without known genetic factors with a few exceptions. Congenital cystic adenomatoid malformation (CCAM) is a congenital pulmonary development defect, accounting for approximately 25% of congenital lung lesions; its clinical presentation is highly variable.

The diagnosis is usually suspected on a chest X-ray, but confirmation and identification of the type of malformation requires more invasive investigations.

Keywords: Congenital cystic adenomatoid malformation, newborn, lobectomy

Introduction

Congenital pulmonary malformations result from developmental abnormalities without known genetic factors, with a few exceptions. Currently, most of them are prenatal diagnosis or during early childhood.

Prenatal care will focus on diagnosing the type of lesion, assessing fetal impact, monitoring progress, and determining the place of birth to optimize birth care.

Case report

It is a newborn baby, resulting from a poorly monitored pregnancy, carried to term, vaginal delivery with good adaptation to extra-uterine life, birth weight was 3.5 kg, mother is 27 years old, primiparous, without significant pathological, no notion of consanguinity;

Parents consulted at day 4 of their newborn's life days for respiratory distress noted at day 2 of life. On admission: the newborn was pink, tonic, reactive, hemodynamically stable. Pleuro-pulmonary examination objectified a respiratory distress rated 2/10 according to the Silverman's score made of a marked sub-costal draw, cardiovascular examination did not objectified any breath or added noise.

The abdominal chest X-ray was performed at admission showed a right pneumothorax!

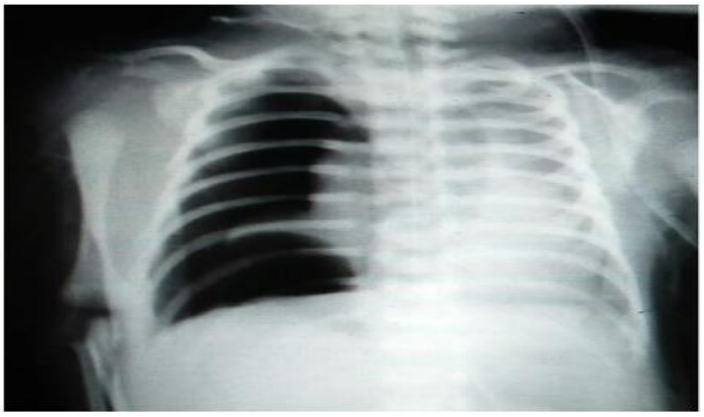

Figure 1: Chest x-ray of the face showing a right pneumothorax

The newborn was hospitalized in a neonatal intensive care unit in a head bag with 100% fio2.

A thoracic CT scan was completed that was in favor of adenomatoid cystic malformation stocker's type I.

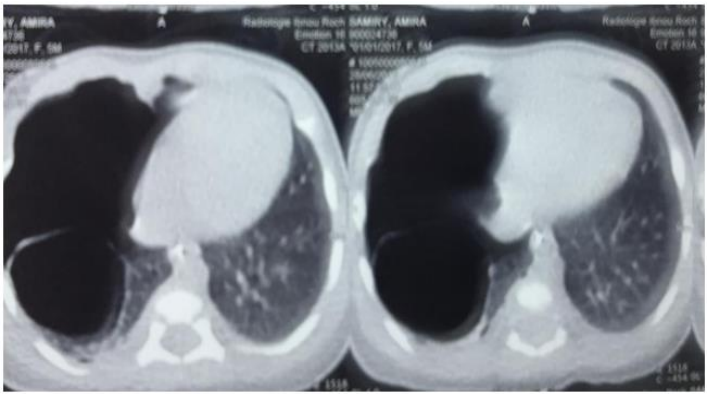

Figure 2: Thoracic CT scan showing adenomatoid cystic malformation stocker's type I.

The assessment was completed with an anatomopathological study of the operating specimen which confirmed the result of the thoracic CT scan.

The postoperative outcomes were simple and the newborn was declared outgoing after 15 days of hospitalization with regular follow-up in consultation.

Discussion

The formation of the lungs begins in the embryonic period with the formation of the trachea and bronchial tubes. Then, it continues until birth with the formation of acing and the differentiation of the cells of the distal lung. This differentiation will continue until the age of two years.

Congenital cystic adenomatoid malformation (CCAM) was described by Ch'in and Tang in 1949, and is defined as an adenomatoid proliferation of terminal respiratory structures, manifested by cysts bordered by a cylindrical or cubic epithelium [1].

Usually, it is unilateral and secondary to a cessation of maturation of the conduction pathways in the bronchioles without the formation of alveolar tissue.

Its incidence is estimated to be between 1/11,000 and 1/35,000 live births [2]

It is the most frequently diagnosed broncho-pulmonary malformation in antenatal. It represents 25% of broncho pulmonary malformations, most often carried in the second trimester during the 22-week morphological ultrasound [3].

The ultrasound aspect is not always conclusive, lesions appearing hyper echoic can be distinguished, most often corresponding to a pulmonary cystic adenomatoid malformation (CCAM) but can also correspond to a sequestration or a bronchogenic cyst. On the other hand, a non-cystic hyperechoic aspect can correspond to a CCAM, sequestration, bronchial atresia, or congenital lobar emphysema.

The evolution is most often spontaneously favorable, and more than 30% of lesions regress in antenatal, but only 70% of these children will actually have no visible lesions on the CT scan after birth [4].

In addition, they can be asymptomatic, discovered during a standard or symptomatic chest X- ray.

Its clinical presentation is highly variable, it is essentially correlated to the size of the malformation [5]. Almost half of newborns are asymptomatic [6, 7-8]. Large malformations can cause signs of respiratory distress and should be promptly operated because of the risk of degeneration.

Diagnosis is usually suspected on a standard X-ray, but confirmation and identification of the type of malformation requires more invasive investigations, namely a thoracic CT scan, which generally identifies the type of pulmonary malformation by objectifying multiple aerial cysts.

In our case, since the pregnancy was poorly monitored, the diagnosis was made only to j4 of life before the installation of respiratory distress secondary to a complication of CCAM: Pneumothorax.

There are classically three types according to Stocker's histological classification: type I, the most frequent (65%): contains macro cysts whose size varies between 2 and 10 cm,

type II, observed in 20 to 35% of patients, consisting of multiple cysts of 0.5 to 2 cm, and type III (10%) which defines micro-cysts whose size is less than 0.5cm[1].

Magnetic resonance imaging has little interest in confirming this diagnosis.

Only the anatomopathological study confirms the diagnosis and eliminates other pulmonary cystic malformations, It shows cysts with aerial or fluid content, limited by a wall deprived cartilage and without inflammatory signs, lined by an epithelium composed of pseudo- stratified ciliated cells and mucus cells [9].

CCAMs are rarely associated with other malformations and the associations described are mainly cardiac malformations and renal agenesis [10]. Hence the interest of completing the malformative assessment, namely a cardiac ultrasound and abdominal-renal ultrasound.

Therapeutic management is based in all cases on surgery. Lobectomy is preferred to segmental resection since it avoids prolonged air leakage and recurrent postoperative infections secondary to incomplete resection of the lesion, is indicated urgently if the newborn is symptomatic, and must be cold programmed if the patient is asymptomatic given the risk of degeneration.

The prognosis depends on the anatomopathological aspects, the infection, the size of the lesion and its impact on the lungs [9]. Indeed, type I of Stocker has a good prognosis, while types II and III are pejorative and are associated with a high incidence of associations with other congenital malformations, particularly renal and gastrointestinal, and with the risk of malignant degeneration.

Conclusion

Congenital cystic adenomatoid malformation is the most frequently diagnosed antenatal broncho pulmonary mal formation; its clinical presentation is highly variable,

The diagnosis is usually suspected on a chest X-ray, but confirmation and identification of the type of malformation requires more invasive investigations,

Therapeutic management is based in all cases on surgery, it is indicated urgently if the newborn is symptomatic.

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