Ref: Ro J Pediatr. 2017;66(2)
DOI: 10.37897/RJP.2017.2.6

CONGENITAL PULMONARY AIRWAY MALFORMATION – CASE PRESENTATION

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ABSTRACT

Congenital pulmonary airway malformation (CPAM) is a rare cystic lesion, whose prognosis depends on the extent of the lesion, the association of other congenital abnormalities, the associated pulmonary pathology and the possibility of curative surgery. We present the case of a 5-month-old infant diagnosed with bilateral CPAM, based on clinical and imaging investigations, with acute pulmonary manifestations and without other associated congenital anomalies.

Keywords: congenital pulmonary airway malformation, cysts, prognosis

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is a rare anomaly of developing lung of hamartomatos type, which consists in the presence of cysts of varying sizes and locations along airways (1). The lesion was formerly known under the name of congenital cystic adenomatoid malformation or cystic pulmonary malformation, currently having an incidence of 1/10.000 to 35.000 of births (2-4). CPAM represents about 25% of lung congenital malformations, the most cases being encountered in newborns and infants males (5-7). Typical manifestations of MCAP are represented by recurrent infections of the respiratory tract or progressive respiratory distress (2).

Approximately 99% of CPAM can be diagnosed by fetal ultrasound in 18-20 weeks of gestation (8). Postnatal sensitivity of chest X-ray is 60% while the sensitivity of CT or MRI is 100% (8). The lesion is usually unilateral (85%), with no difference in incidence of macrocystic or microcystic forms, but are also reported cases affecting bilateral or an entire lung (7,9).

CASE PRESENTATION

We present the case of C.M. infant, a 5-month-old male, diagnosed with pulmonary malformation, admitted to the Pediatric Clinic of the Emergency Clinical County Hospital Craiova.

The patient was hospitalized for an episode of apnea with cyanosis, spastic cough and dyspnea. *Heredocollateral background* indicated no significant information. *Physiological history*: birth by cesarean delivery at term, birth weight of 3,200 g, Apgar score 9, physiological jaundice, breast diet for 6 weeks and later with milk, rickets prevention done properly, vaccination in maternity. *Pathological history*: at age of 4 months an admission in Pediatric Surgery Clinic for left pneumothorax and subcutaneous thoracic emphysema with laterocervical extension.

Current **physical examination** indicated a weight of 4,800 g, influenced general status, pale skin, anterior normotensive fontanelle of 3/3.5 cm, perioronasal cyanosis, spastic cough, dyspnea, RF (respiratory frecvency) = 50/min, stetacustic lung crepitant and subcrepitant rales in right hemitho-

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rax, VA (ventricular allure) = 120/min, supple abdomen, normal stool.

Paraclinical data: normal values of the transaminases, glycemia, urea, creatinine, serum iron, ionogram. The CBC (complete blood count) indicated an increased number of leukocytes, ESR (erythrocyte sedimentation rate) = 32/55 mm.

Chest X-ray performed on admission indicated the presence of bilateral cystic images with cysts varying in size, ranging between 0,5 and 2,5 cm (Fig. 1A).

Chest computed tomography confirmed the appearance of bilateral pulmonary air cysts, with thin wall, up to 20 mm thickness, with the presence of a giant cyst of 3,8 cm to the right upper lung lobe (Fig. 1B).

The cardiac **ultrasound** and the abdominal ultrasound revealed no abnormalities.

The evolution under the antibiotic and symptomatic therapy was slow favorable, the functional respiratory syndrome was resolved gradually. The control chest radiography after 7 days, revealed the remission of basal and hilar pulmonary opacities. He was discharged with the indication of medical and radiological regular surveillance. Currently, the patient is stable, without any bronchopulmonary acute episodes.

DISCUSSIONS

Congenital cystic lung disease refers to a wide range of entities including cystic adenomatoid malformation, bronchogenic cyst, pulmonary sequestration, congenital lobar emphysema, which are differentiated on the basis of clinical imaging and histopathological aspects (10). In the presented case, the diagnosis of CPAM was supported by the location, size, wall thickness and distribution of cysts, adjacent lung tissue and vasculature aspect and also by associated symptoms. Other entities like congenital diaphragmatic hernia, bronchiectasis, cystic fibrosis, mediastinal masses (teratoma, cystic hygroma, blastoma) and infectious causes, which can sometimes cause problems of differentiation with CPAM, were clinical and imaging excluded (5,11,12).

The cause of CPAM is unknown, but some studies indicate the disturbance of the embryological development between 5th and 22nd week of pregnancy, with the occurrence of some abnormal structires in the terminal bronchioles (5,7). The lesion is similar to hamartoma, with many bronchiolar elements but without the presence of cartilaginous tissue, without obvious alveolar differentiation and mucus production (7,13).

Based on the classification of Stocker ST et al. and taking into account the histopathological micro- and macroscopic criteria and clinical aspects are described five types of CPAM (1,4,11). Type 0 has origin tracheobronchial and is incompatible with life (10,14).

Type I is the most common (50-70%), is composed of single or multiple large cysts (> 2 cm), usually affects only one lobe and is not associated with other malformations, with a good prognosis (10,12). Type II (15-30%) has lesions consisting of multiple small cysts (<2 cm), sometimes associated with other anomalies incompatible with life, such as pulmonary hypoplasia, renal agenesis, atresia of the esophagus (4). Type III is rare (5-10%) and may affect a lobe, an entire lung or, less commonly, both lungs, with cysts less than 0.2 cm and frequently

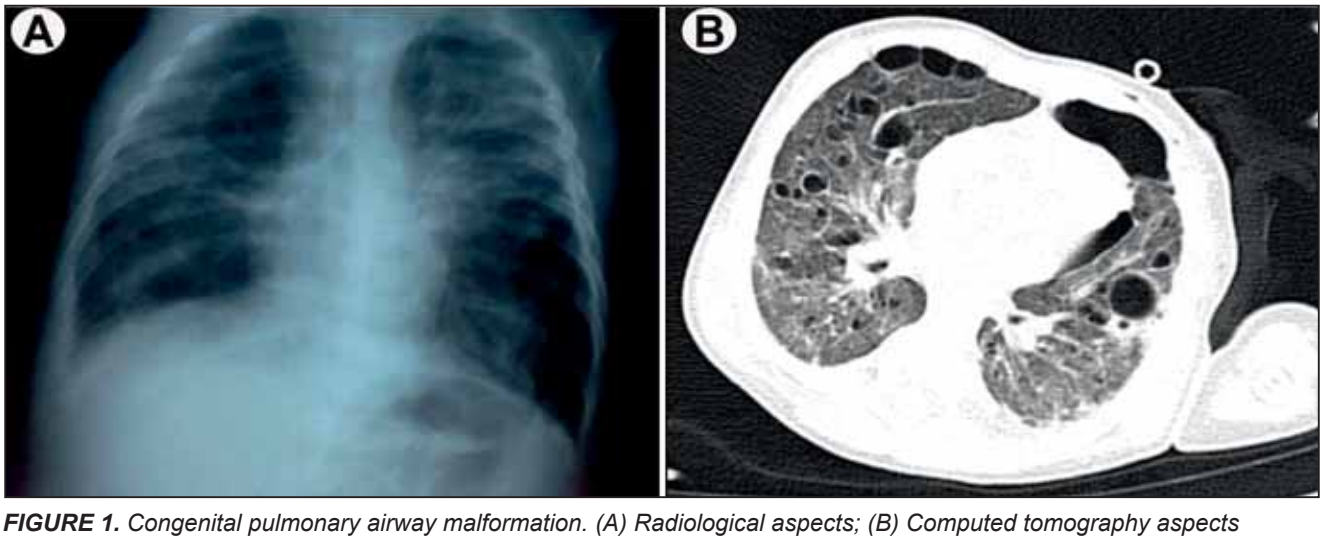

FIGURE 1. *Congenital pulmonary airway malformation. (A) Radiological aspects; (B) Computed tomography aspects*

associated with fetal hydrops (15). Type 4 (10%) is macrochistic and is commonly associated with malignant tumors (13).

Based on imaging aspects and the absence of other congenital anomalies, but in the absence of histopathological investigations, the presented case may be associated with the type 1 of CPAM. Although this is often located usually unilateral also bilateral cases have been reported (16).

CPAM prognosis is given by the extent of the lesion, the presence of associated congenital anomalies and intercurrent infections (15). Generally, in symptomatic CPAM, the affected tissue lobectomy is indicated and for asymptomatic cases, clinical and imaging follow-up of patients, may be an option, as only 10% of them will develop in the future inflammatory or neoplastic complications (7). Although Type 1 of CPAM usually associate a good prognosis, in the presented case, the surgical intervention was not an option to ensure a favorable evolution, given the diffuse appearance of cysts in both pulmonary areas. In addition, the development of lung parenchyma until the age of 8 years, and reporting of cases of type 1 CPAM in young adults (16), may be an argument for the therapeutic expectation attitude.

CONCLUSIONS

The particularity of the presented case is given by the diagnosis of CPAM, with a poor prognosis due to bilateral diffuse location, in the context of clinical and imaging favorable evolution and the absence of other congenital abnormalities.

Preventing and combating of acute pulmonary infectious or non-infectious episodes and the periodical imaging follow-up of patients may be an effective conservative therapeutic attitude, especially for bilateral CPAM.

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