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Case Report

Congenital Pulmonary Airway Malformation: A Report of 2 Cases

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Abstract

Congenital lung malformations are a group of infrequent pathologies originating in an alteration during lung morphogenesis. Congenital pulmonary airway malformation (CPAM) is the most frequent of them, previously known as congenital cystic adenomatoid malformation, characterized by an overgrowth and dilatation of bronchial structures. They may be asymptomatic or manifest as fetal hydrops and neonatal respiratory distress. The diagnosis is established in the majority of cases during intrauterine life, allowing prenatal treatment in serious cases in order to improve survival. There is no consensus about postnatal treatment in asymptomatic cases, in which a wait-and-see approach or surgical resection could be decided to avoid complications. In patients with symptoms at birth there is no doubt as to the indication of surgical resection.

Keywords: *Congenital Lung Malformations; Congenital Pulmonary Airway Malformation Type 2; Surgical Resection; Pulmonary Sequestration; Bronchogenic Cyst; Lung Cancer Risk*

Abbreviations

CPAM: Congenital Pulmonary Airway Malformation; CT: Computed Tomography

Introduction

Congenital lung malformations encompass a heterogeneous group of pathologies originated in an alteration of embryologic development of the lungs and airways [1], with a reported incidence between 1:25000 and 1:35000 newborns [2]. Among these malformations we found: congenital pulmonary airway malformation (CPAM), pulmonary sequestration, bronchial atresia, congenital lobar emphysema, bronchogenic cyst. The most frequent of these congenital lung malformations is CPAM, which constitutes 30 - 40% of all pulmonary malformations [3] and is defined as a group of solid lesions, cystic o mixed, not functional, in communication with the bronchial tree and almost always irrigated by pulmonary vascular territory. CPAM are divided into 5 types according to the classification established by Stocker., *et al*. [4], relating to the number and size of the cysts and their anatomopathological origin; type 0: all pulmonary lobules are compromised by cysts < 0.5 cm, it is incompatible with life; type 1: a single cyst o multiple cysts > 2 cm originating from the bronchi o bronchiole, it is the most frequent and with the best prognosis; type 2: single cyst or multiple cysts between 0.5 - 2 cm originating in the bronchiole, it has a high association with pulmonary sequestration and with malformations in other systems; type 3: solid lesion with cysts < 0.5 cm that originate in bronchiole and alveolar ducts, it can compress the contralateral lung resulting in pulmonary hypertension and fetal hydrops : type 4: large cysts > 7 cm of acinar origin.

The advancements in imaging techniques have allowed for the majority of cases of CPAM to be diagnosed prenatally (50 - 85% of cases) [5] and only being diagnosed postnatally in a few, as a debut of respiratory distress at birth, recurrent respiratory infections or a casual finding in a radiography. Clinical manifestations fundamentally depend on the size of the lesion and other associated malformations [3].

Imaging techniques of choice for the prenatal diagnosis are the echography and magnetic resonance imaging, whereas for the postnatal diagnosis radiography and CT scans are the methods of choice [2]. We will now describe 2 cases of type 2 CPAM diagnosed at our center in 2018.

Case Report

Case 1

Female newborn in whom a single cyst of 1.6 cm x 0.9 cm is detected prenatally in the right inferior lobule, being diagnosed as CPAM type 2, without mediastinal displacement, hydrops or other malformations. Medical history: normal pregnancy follow-up, amniocentesis 46XX, amniorrhexis of 1 hour, vaginal delivery, Apgar in the first and fifth minutes 9 and 10 respectively, no needed for resuscitation, adequate weight for gestational age, healthy parents.

The patient is asymptomatic since birth with normal physical examination. Chest radiography is requested without findings. Echographies are performed of heart, brain and abdomen without alterations. A CT scan at 6 months of age is diagnostic of type 2 CPAM in coexistence with intralobar pulmonary sequestration: Area of dysplasic multi-cystic lung tissue of 3.3 cm x 2.6 cm, in the segments 7 and 10 of the inferior right lobule that presents abnormal arterial vascularization from 2 aberrant branches that originate from the abdominal aorta (Figure 1). The patient stays asymptomatic and is referred to the center of Paediatric Surgery where a lobectomy of the inferior right lobe is performed by thoracoscopy at 2 years of age without complications.

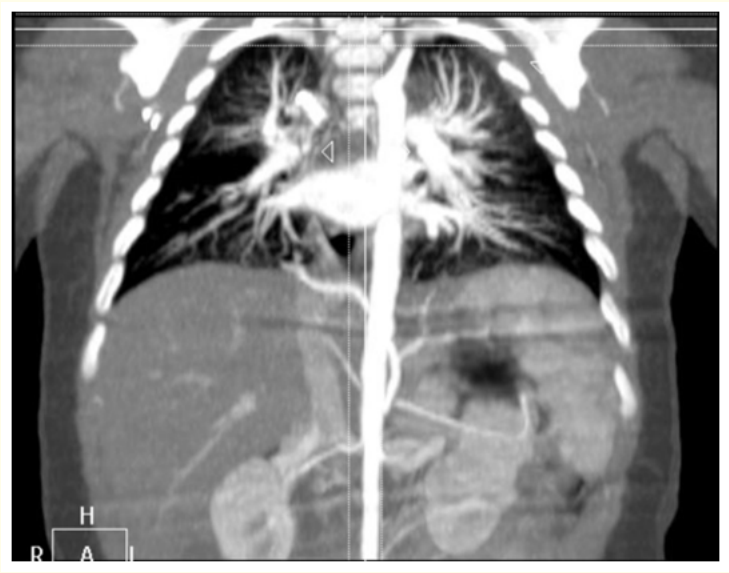

Figure 1: *CT Scan. An ascending branch can be seen from the abdominal aorta that supplies the lung malformation (intralobar pulmonary sequestration).*

Case 2

Male newborn with prenatal diagnosis of a single 1 cm x 1.5 cm cyst in the superior right lobule compatible with CPAM type 2 without other findings. Medical history: normal pregnancy follow-up, amniocentesis 46XY, amniorrhexis 5 hours, vaginal delivery, Apgar at the first and fifth minute 9 and 10 respectively, no needed for resuscitation, adequate weight for gestational age, healthy parents.

The patient is asymptomatic since birth with normal physical examination. Chest radiography performed at birth shows radiolucent image of round morphology in the superior right lobule which causes discrete mass-effect in adjacent parenchyma. Echographies are performed of heart, brain and abdomen without findings. The patient does not exhibit breathing difficulties or other symptoms and is discharged from Hospital after 24 hours of monitoring.

CT scan is performed at 6 months of age, being diagnostic of CPAM type 2 in association with bronchogenic cyst: CT shows an area of 4.6 cm x 3.3 cm in the segment 2 of the superior right lobule, composed of coalescent multiple cysts less than 1cm and associated to a 10 mm cystic lesion in the main right bronchi. The patient is referred to the center of Paediatric Surgery where a lobectomy of the superior right lobe is performed by thoracoscopy at 14 months of age without complications.

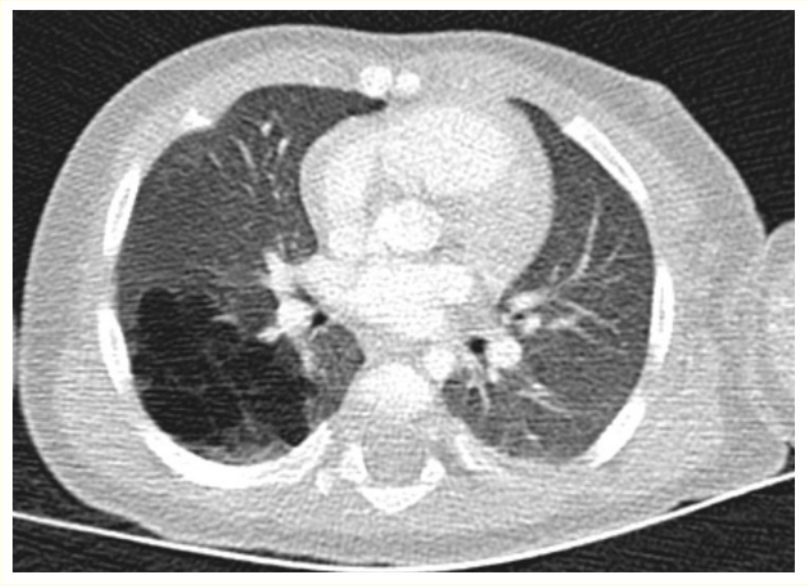

Figure 2: *CT Scan. Confluent multicystic zone in superior right lobule.*

Discussion

CPAM are caused by an alteration in pulmonary maturation characterized by the presence of adenomatoid-cystic tissue, whose prognosis depends on the size of the malformation, the subtype and the presence of other anomalies. CPAM types 1, 2 and 4 have a favorable prognosis, whereas types 0 and 3 do not. Although the risk of malignancy is low, there is an association between type 1 CPAM and bronchioalveolar carcinoma, and type 4 CPAM and pleuropulmonary blastoma [2].

Clinical manifestations can be very varied but usually there are 3 clinical features according to age at debut: serious prenatal presentation with hydrops; in the neonatal period as respiratory distress; later as bacterial infection or as a radiological casual finding [2]. They can also display as pneumothorax, hemothorax and hemoptysis, although up to 90% remain asymptomatic in neonatal period [6]. Fetal hydrops is the most relevant factor for the prognosis as it is and indicator for fetal intervention [7]. Cavoretto P., *et al.* have reported a mortality rate less than 5% in CPAM without hydrops, while mortality increases to 95% in cases associated with hydrops in which an expectant attitude is maintained [8]. Prenatal treatment includes the administration of maternal steroids, percutaneous aspiration of cysts, thoracoamniotic derivation or surgery during labor by the EXIT technique [9]. With respect to postnatal management, the treatment of

choice in symptomatic patients with CPAM is surgery, but there is controversy over the management of asymptomatic patients in which some authors defend the wait-and-see approach and others advocate for a surgical procedure to avoid the risk of malignancy or other complications. In case of elective surgery in asymptomatic patients, it should be performed within the first year of life, preferably through thoracoscopy for its lower morbidity [10]. In our patients, despite them not showing symptoms, surgical excision was performed to avoid complications.

It is possible to find an association between various congenital lung malformations, one of our patients present CPAM type 2 and an intralobar pulmonary sequestration and the other patient CPAM type 2 in coexistence with bronchogenic cyst. Also, up to 50% of CPAM type 2 show extrapulmonary anomalies: cardiovascular, diaphragmatic herniation, extralobar sequestration and renal agenesis. Despite its frequency none of our patients showed any extrapulmonary malformation.

Conclusion

CPAM is the most common congenital lung malformation. It can be severe in some cases, therefore a prenatal diagnosis and a multidisciplinary management is required to establish the proper course of action. A close follow-up is necessary for these patients for the possibility to show other malformations as well as complications as pneumothorax or malignancy. Although there is controversy over the treatment of asymptomatic patients it seems that there is a tendency towards elective surgery in the first year of life to avoid posterior complications.

Conflict of Interest

None to be declare.

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