Case study

Congenital cystic adenomatoid malformation: A case report

Abstract:

The case report is being done to increase the knowledge of Congenital Cystic Adenomatoid Malformation (CCAM) along with to better understand about the disease and its management & raise awareness. A female infant (aged 9 months and birth weight 2500 gm) was selected as participant. After physical examination, the state of the participant was good (fever:37.6ºC, heart rate: 120/min, respiratory rate: 40/min). Holding a good health certificate, a cystic image was identified at pulmonary artery in chest X-ray report. The histology findings were consistent with CPAM Type I. CPAM surgery, generally favorable, results in low postoperative morbidity and mortality. CPAM is a rare developmental malformation of lung that causes pulmonary compression and hypoplasia leading to respiratory distress but the postnatal management of asymptomatic CPAM remains controversial.

Key words: CCAM, Cystic image, Morbidity, Mortality, Congenital abnormality, carcinoma.

Introduction:

Congenital cystic adenomatoid malformation (CCAM) is a congenital abnormality of lung development caused by an overgrowth of abnormal lung tissue that forms cysts [1]. In the past few years, CCAM was renamed as congenital pulmonary airway malformation (CPAM) and classified into five types by Stocker based on clinical and pathological features [2]. The clinical presentation can range from respiratory distress to lung infection. Some cases may remain asymptomatic throughout life [3]. Surgical removal of the affected lobe of the lung is the best treatment for CPAM. It prevents complications such as repeated infection of the lungs, hemo-pneumothorax, hemoptysis, and potential risk of malignant transformation [4].
We aim through this case, to better understand the disease and its management and raise awareness about its seriousness.

Case presentation:

A Female infant, 9 months old, from a consanguineous marriage of 2nd degree, from a well-attended pregnancy carried to term, birth weight: 2500 gm, no neonatal suffering. In history two hospitalizations for pulmonary infection which were successfully treated with antibiotics. 3

weeks before her hospitalization, she presented a cough then a respiratory discomfort with no fever. On physical examination, medical state was good, fever: 37.6ºC, heart rate: 120/min, respiratory rate: 40/min, no anomalies in the pulmonary auscultation. Other system examinations were normal. The blood parameters were within normal limits. The chest X-ray showed a mediastinal mass with suspicion of the pulmonary artery dilatation, completed by a cardiac ultrasound with a cystic image in the mediastinal area.

The CT scan examination with contrast was performed on this patient, revealing the presence of a mass in the middle and upper left mediastinum of liquid density, well limited, with regular contours, with homogeneous content. Given these elements, we suspected a bronchogenic cyst, a thymic cyst or cystic lymphangioma. The patient underwent a resection of the mass, the anatomopathological findings confirmed the existence of CPAM type I.

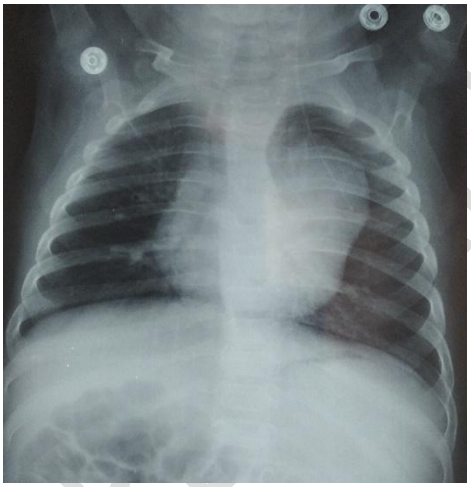

Fig 1: Chest X-ray showing a mediastinal mass with suspicion of the pulmonary artery dilatation.

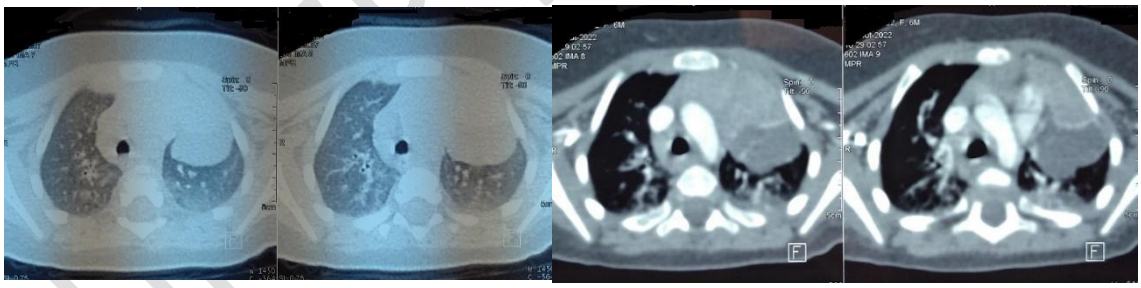

Fig 2: CT scan examination revealing the presence of a mass in the middle and upper left mediastinum of liquid density, well limited, with regular contours, with homogeneous content. Wall enhancement after injection of the contrast medium.

Discussion:

CPAM was first described in 1897 by Stoerk [5] and individualized by Ch'in and Tang in 1949 [6]. They are the most common form of congenital parenchymal lung malformations (25% [7]) and the incidence ranges from 1/10,000 to 1/35,000 children [8]. Most of the cases are diagnosed

within first 2 years of life. It is usually a unilateral condition and restricted to one lobe, but can involve whole of one lung, or both the lungs [9,10].

Currently, thanks to advances in prenatal imaging, pulmonary tract defects can be detected during pregnancy or at birth, and allows for perinatal care planning [11, 12]. In Morocco, there is no prenatal diagnosis, pregnancies are not well attended, and there is a lack of experienced practitioners. The prenatal outcome of CPAM is variable, it may result in hydrops fetalis in 40% of cases or regress completely in 15% of cases [13]. The mode of revelation may be acute respiratory distress. In rare cases, it may remain asymptomatic and be of incidental radiological finding.

In our case, the histology findings were consistent with CPAM Type I. It is the most common subtype, corresponding to 65% of the cases. Type I consists of single or multiple large cysts (>2 cm), filled with air or fluid, and are communicated with surrounding bronchial tube and pulmonary parenchyma, so it is difficult to be regarded as true cyst. In some cyst wall, there are mucous secreting cells. There are no cartilages on the walls and no other associated anomalies. CPAM type I has good prognosis. [14, 2]

The chest X-ray may show single or multiple unilateral lesions, hence the interest of the CT scan. The differential diagnosis of CPAM includes pulmonary sequestration, pneumatocele, congenital lobar emphysema, cystic hygroma, thymic cycst and bronchogenic cysts. Pulmonary sequestration can be distinguished by the presence of abnormal artery. In pneumatocele, complex epithelial and stromal components are not present as in CPAM. There are no alveoli at the distance between the cysts in the lobar emphysema. Bronchogenic cyst can be distinguished by its placement in the hilar region, solitary formation, and the presence of cartilage in the wall [15, 16]. Cystic hygromas are thought to develop when lymphatic tissue becomes isolated from lymphatic sacs during the formation of lymphatico-venous sacs. These isolated tissues do not establish communication with the rest of the lymphatic or venous system. Subsequently, the sequestered lymphatic tissues expand, leading to the formation of cystic structures characteristic of these lesions. Thymic cysts arise from the incomplete closure of the thymopharyngeal duct. These ducts should disappear during development but may persist, forming cystic structures. Thymic cysts are found in the anterior mediastinum and are filled with fluid or other tissues. In countries where tuberculosis such as our country is common, pulmonary tuberculosis should be considered in differential diagnosis. The most frequent malignant complication associated with CPAM is pleuropulmonary blastoma, in CPAM type 4. There is an association between CPAM type 1 and bronchoalveolar carcinoma [17].

When diagnosed prenatally, clinical and ultrasound obstetrical surveillance is required, CPAM can be managed before delivery if there is a risk of hydrops fetalis. In the postnatal period, if the infant is asymptomatic, the majority of authors recommend surgery especially in early childhood to prevent complications, mainly recurrent infections and pneumothorax, and to benefit from the lung growth potential of the first four years of life [18, 19]. Others recommend abstention as they believe that the risks of CPAM are insignificant compared to the morbidity of surgical treatment [20]. However, abstention has the inconvenience of a very long period of regular clinical and annual scannographic surveillance. For symptomatic cases, lobectomy is the

treatment of choice [19]. The outcome of CPAM surgery is generally favorable. It results in low postoperative morbidity and mortality, shorter hospital stay and mainly reduces the risk of recurrence [7, 11, 17].

Conclusion:

CPAM is a rare developmental malformation of lung that causes pulmonary compression and hypoplasia leading to respiratory distress. In cases of prolonged lung infection that do not respond to treatment, CCAM should be considered and histopathologic study should be performed to establish a definitive diagnosis. Surgery remains the cornerstone treatment of symptomatic lesions, but the postnatal management of asymptomatic CPAM remains controversial.

Consent:

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

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