Un pneumothorax spontané chez un enfant?
Pensez à la malformation adénomatoïde kystique du poumon

Spontaneous pneumothorax in a child?
Think of cystic adenomatoid malformation of the lung

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Résumé :

Introduction : La malformation adénomatoïde kystique est l'anomalie congénitale la plus fréquente du poumon. Le pneumothorax spontané est rarement décrit comme la manifestation initiale de cette malformation.

Objectif : Nous rapportons l'observation d'un enfant dont la malformation adenomatoide kytique du poumon était révélée par un pneumothorax.

Observation : Enfant âgé de dix ans qui avait consulté pour dyspnée et douleur thoracique gauche évoluant depuis sept jours. L'enfant avait à l'examen une tachypnée avec un tympanique à la percussion du thorax et une diminution des murmures vésiculaires au niveau de l'hémithorax gauche. Une radiographie du thorax pratiquée en urgence avait mis en évidence un pneumothorax avec un collapsus pulmonaire gauche. Des lésions kystiques étaient observées dans le poumon gauche. Une exsufflation était pratiquée. Une tomodensitométrie thoracique avait permis d'observer la récidive du pneumothorax et une grande cavité à paroi mince capsulée occupant le lobe inférieur gauche. Une deuxième exsufflation était faite. Une lobectomie était réalisée par la suite. L'étude histologique avait confirmé le diagnostic de malformation congénitale adénomatoïde kystique type I du poumon.

Conclusion: les cliniciens doivent être conscients du faite qu'un pneumothorax spontané peut être la présentation initiale de la malformation congénitale adénomatoïde kystique du poumon chez les enfants. Des examens radiologiques appropriés doivent être demandés pour diagnostiquer cette malformation. En présence d'une grande cavité avec une déviation du médiastin, le drainage de l'épanchement gazeux doit être fait très lentement.

Abstract

Introduction : Congenital cystic adenomatoid malformation is the most common congenital lung lesion. Spontaneous pneumothorax, as the initial manifestation of congenital cystic adenomatoid malformation, is rare and few cases have been described in the literature.
Purpose: We report a case of this disease in a child who presented with features of pneumothorax.

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Case presentation : A ten year old boy had a seven day history of onset of breathlessness and pain in the left side of the chest. The child had tachypnea with a tympanic note on percussion and decreased breath sounds on the left hemithorax. An urgent chest X-ray revealed a large pneumothorax with collapsed left lung. Few cystic lesions were noted in the left lung. A needle aspiration was done. A computed tomography scan of chest showed recurrence of the pneumothorax, cutaneous emphysema and demonstrated a large well defined capsulated and thin walled cavity occupying left lower lobe. A second needle aspiration chest was put to drain the air. A lobectomy was done. The histological study confirmed the diagnosis of type I cystic adenomatoid malformation of the lung.

Conclusion : clinicians should be aware of the possibility that spontaneous pneumothorax may be the initial presentation of CCAM in children. They should undergo appropriate imaging studies to diagnose this malformation

Mots clés : malformation adénomatoïde kystique du poumon,Pneumothorax, enfant

Key words : Congenital cystic adenomatoid malformation; Pneumothorax, child

INTRODUCTION:

Congenital cystic adenomatoid malformation of the lung (CCAM) also known as congenital pulmonary airway malformation (CPAM) is the most common congenital lung lesion. It results from the failure of normal bronchoalveolar development which probably occurs at about the 5th–6th week of gestation with hamartomatous proliferation of terminal respiratory units in a gland-like pattern (adenomatoid) without proper alveolar formation [1]. The pathological mechanisms responsible for this lesion remain unknown.

CCAM is usually considered a fetal or postnatal (infantile) disease that is usually presented as respiratory distress in the neonatal period. On occasion, CCAM may remain asymptomatic and be discovered after becoming complicated. Spontaneous pneumothorax (PTX), as the initial manifestation of CCAM, is rare and few cases have been reported in the literature. We report a case of CCAM in a child who presented with features of pneumothorax.

CASE REPORT

A ten year old boy had a seven day history of onset of breathlessness and pain in the left side of the chest. He had no significant past medical history. The patient had been born at term and had no neonatal problems. At initial presentation, the child had tachypnea with a tympanic note on percussion and decreased breath sounds on the left hemithorax. The oxygen saturation was at 96%. He was normotensive with a heart rate of 150/min.

An urgent chest X-ray (CXR) revealed a large pneumothorax with collapsed left lung. Few cystic lesions were noted in the collapsed portion of the left lung and a right deviation of mediastinum was noted (figure 1).

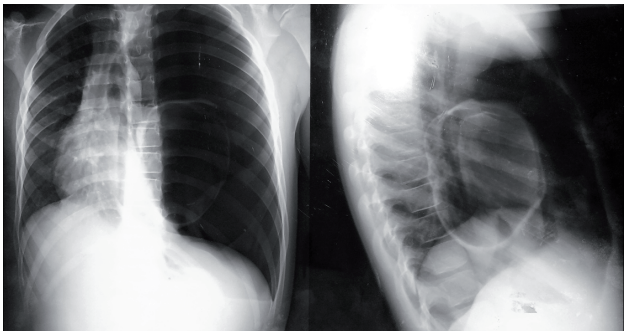

FIGURE 1 : (a, b) : Chest radiograph AP view1a and left lateral view1b: shows large left sided pneumothorax with compression of the collapsed lung and shift of the mediastinum to the right . Also seen a large thin-walled cystic lesion in the collapsed left lung.

A needle aspiration was done slowly to allow left lung reexpansion and pleural drainage of 700 ml of air. The procedure relieved the child's symptoms. On follow up radiograph there was complete opacification of the left hemithorax. The cystic lesion persists on the left side (fig 2, 3).

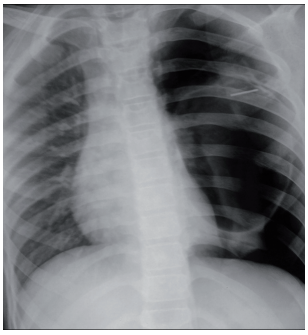

FIGURE 2 : Chest radiograph taken at the time of putting a needle into the left hemithorax for initial drainage of air

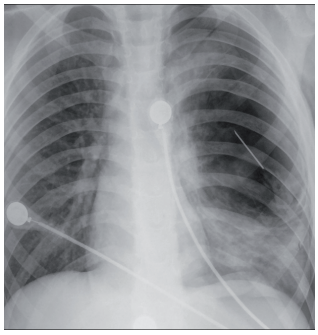

FIGURE 3 : Following needle aspiration Still well defined cystic lesion is seen in left zone

The next day, the patient was sent for a computed tomography (CT) scan of chest. This showed recurrence of the pneumothorax, cutaneous emphysema and demonstrated a large well defined capsulated and thin walled cavity occupying left lower lobe. This cystic lesion measured 100x85x 65 mm and contains incomplet thin septations (figure 4).

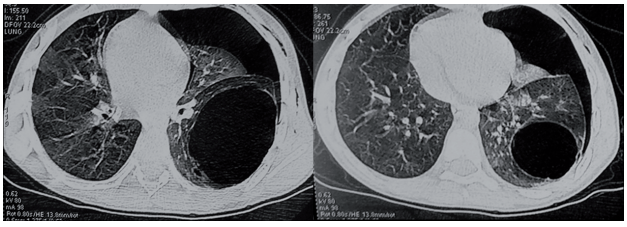

FIGURE 4 : (a, b) : CT scan in lung window shows a cutaneous emphysema, a large pneumothorax in the left side with well defined capsulated and thin walled cavity occupying left lower lobe(a) , a marked shift of mediastinum to theright side

are re-demonstrated (b). rax with compression of the collapsed lung and shift of the mediastinum to the right . Also seen a large thin-walled cystic lesion in the collapsed left lung.

The coronal and sagital reformat showed no obvious communication with lower lobe bronchus. The CT examination was reported as congenital cystic adenomatoid malformation.

Since there was a recurrence, the insertion of a chest drain was discussed but it was decided to put a second needle aspiration chest to drain the air. The patient condition improved significantly and he was referred to surgery service. The child underwent open surgical exploration through a posterolateral thoracotomy. The exploration found a CCAM depending on the left lower lobe. A lobectomy was done without any complications. The histological study confirmed the diagnosis of type I cystic adenomatoid malformation of the lung.
Two months after surgery clinical examinations of the child were normal. The CXR showed a good expansion of the left lung.

DISCUSSION:

CCAM represents a rare anomaly of the lung. With the advancement in antenatal sonography, most CCAM are nowadays detected antenatally and its frequency is higher than previously reported. Data from large population registries suggest an incidence of 1 in 10,000 pregnancies [2]. In developing country, the antenatal diagnosis of CCAM is still uncommon and the diagnosis occurs once the child becomes symptomatic.

The term ''congenital cystic adenomatoid malformation'' was coined by Ch'in and Tang in 1949 [3]. Stocker JT et al [4] published a classification of CCAM into three major groups (Type I-III) which later revised in 2002 [2]. Two additional types (Type 0 and IV) have been added to make up the five current group CPAM classifications. Type 0 CCAM is an extremely rare and lethal condition in which the respiratory bronchioles, alveolar ducts and alveoli fail to develop. Type I CCAM is the most common CPAM and accounts for 60 to 70 percent of all cases. It is composed of one or several large cysts greater than 2 cm in diameter lined by a ciliated pseudostratified columnar epithelium. Our case fits into this group. Type II CCAM consists in multiple small cysts, usually less than 2 cm in diameter whereas type III is a solid adenomatoid malformation with minor cystic components. Finally, type IV CCAM is characterized by large peripheral thin-walled cysts lined by pneumocytes.

Variable presentations of CCAM have been described in the literature. Cases may be indentified prenatally by routine ultrasonography screening. The disease is most commonly found in the neonatal period and up to 90% of diagnoses are made within the first two years of life [5].Affected newborns typically present with symptoms of respiratory distress soon after birth, including tachypnoea, grunting, retractions and cyanosis. More commonly, children remain asymptomatic.

Complications can include recurrent infections, pulmonary hemorrhage or PTX.
Our patient presented with PTX at the age of 10 years old. There was no history of respiratory complications at or soon after birth.
CCAM is a rare condition that may be difficult to diagnose, especially in patients who do not present in infancy with the classic respiratory distress.
CCAM has been shown to present as PTX in 14 % of cases in some series [6].

The CXR was once the most efficient method of diagnosing pulmonary surgical lesions. However, the normal CXR does not exclude the presence of CCAM. CT scan is more accurate than CXR for the detection and follow-up of CCAM. CT not only delineates more accurately the location and extent of the lesion, but also identifies the vascularization (arterial and venous) of the malformation and rules out if there is a systemic supply to the lesion or if there are other associated pulmonary congenital anomalies.

The diagnostic method of choice is high-resolution CT, which helps clarify the diagnosis by allowing excellent visualization of the cysts to determine their distribution, size, site, and wall thickness. Ultrasound or MRI also may be a powerful tool in diagnosis of CCAM.

Distinguishing between CCAM, especially type 1, which has a large cyst, cystic bronchiectasis, bronchogenic cysts and congenital lobar emphysema, is not always possible as in our case.

We would recommend CT scan for all children with large, first-time and recurrent spontaneous PTX.
There are a few publications describing cases of spontaneous PTX as the initial presentation of CCAM in children [7-9].Tension pneumothorax is an uncommon disease with a malignant course leading to death if untreated. This case presented an acute management problem to the staff.

A chest tube was inserted in the majority of published cases presented with PTX.
Chest tube insertion should be avoided in CCAM because that may worsening the respiratory distress and increasing the rate of complications like PTX. Prabhu SM et al recommended chest tube insertion only in extreme cases with severe respiratory distress and significant mediastinal shift which may decompress a potential pneumothorax, tension pneumatocele, or a large CCAM [10].

On the other hand, Ganguly T et al [9] reported a case of type 1 CCAM in a child complicated by a rapid hemodynamic deterioration immediately after drainage tube insertion. In presence of large cavity and mediastinal shift pleura, the authors suggest needle thoracostomy as a mean to controlled decompression to prevent catastrophe.

In our case, once the diagnosis of pneumothorax had been made, a decision was made to perform a needle decompression. In retrospect, it was felt that the best treatment option had been taken by inserting a needle into the chest to decompresse slowly.

Surgery is the only definitive diagnosis and treatment. There is agreement between surgeons regarding the treatment of symptomatic patients, but controversy exists about the management of asymptomatic neonates and infants with CCAM. Some authors advocate early intervention. Advantages of early resection include prevention of possible recurrent infections and later Malignancy and the low rate of complications in elective versus emergency surgery.

Lobectomy clearly remains the procedure of choice, to prevent residual disease and recurrence in the remaining lobe. The initial approach may be by video-assisted thoracoscopy, as this allows complete resection of the lesions while avoiding the drawbacks of thoracotomy. In our patient, we did not use video-assisted thoracoscopy but rather direct posterolateral thoracotomy to assess the extent the parenchyma to be resected by palpation and meticulous inspection. The prognosis is excellent after surgical resection in Type 1 CPAM.

Conclusion

CCAM is a rare congenital malformation of the lungs. This abnormality usually considered a fetal or postnatal (infantile) disease. CCAM may present in the older child an incidental finding or secondary to complication. Clinicians should be aware of the possibility that spontaneous pneumothorax may be the initial presentation of CCAM in children. They should undergo appropriate imaging studies to diagnose this malformation. In presence of large cavity and mediastinal shift pleura should be decompressed slowly. Surgery is the only definitive diagnosis and treatment. Although rare, recognition of CCAM pre- or post-natally is important to undertake surgery early and prevent the complications.

Abréviations :

CCAM: Congenital cystic adenomatoid malformation of the lung
CPAM : Congenital pulmonary airway malformation, CT: Computed tomography , CXR: Chest X-ray , PTX : Pneumothorax

Conflict of interest

Authors declare no conflict of interest.

Ethical approval :

This article does not contain any studies with human participants or animals performed by any of the authors.

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