Congenital pulmonary airway malformation (CPAM) mimicking an spontaneous pneumothorax in a newborn

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ABSTRACT

Introduction. Patients with congenital pulmonary airway malformation (CPAM) are usually asymptomatic, but some may present with respiratory distress. We report a rare presentation of a CPAM as an image compatible with persistent and localized spontaneous pneumothorax.

Case report. A 2-month-old male infant without prenatal diagnosis, postnatal distress or barotrauma, was admitted with acute respiratory symptoms and a right tension pneumothorax on chest X-ray. Despite placement of a chest drain, radiological image persisted. CT confirmed the presence of a CPAM. An open surgical approach was decided and a huge bulla depending from the right upper apex lobe was found and resected. Pathological report disclosed type 4 CPAM.

Discussion/conclusion. Despite the negative prenatal screening, the diagnosis of CPAM should be considered in a patient with sudden respiratory distress and suspicion of an spontaneous pneumothorax. Type 4 CPAM may appear like unique lung cyst mimicking a spontaneous bullae or a massive pneumothorax.

KEY WORDS: Infant; Congenital pulmonary airway malformation; Pneumothorax.

MALFORMACIÓN PULMONAR CONGÉNITA SIMULANDO UN NEUMOTÓRAX ESPONTÁNEO EN UN RECIÉN NACIDO

RESUMEN

Introducción. Los pacientes con malformaciones congénitas pulmonares y de la vía aérea (CPAM en sus siglas inglesas) están habitualmente asintomáticos, aunque algunos pueden presentar dificultad respiratoria. Presentamos un raro caso de CPAM diagnosticado en las pruebas de imagen como un neumotórax aislado y persistente.

Caso clínico. Varón de dos meses de edad sin diagnóstico prenatal alguno, dificultad respiratoria perinatal o barotrauma, que ingresó con síntomas respiratorios agudos y un neumotórax a tensión derecho visible en la radiografía de tórax que persistió a pesar de la colocación de un drenaje torácico. El TC confirmó la presencia de una CPAM. En la toracotomía practicada se observó, dependiendo del ápex del lóbulo superior derecho, una enorme bulla que fue resecada. El informe anatomopatológico fue de CPAM tipo IV.

Discusión/conclusión. Aunque no exista diagnóstico prenatal, el diagnóstico de CPAM debe ser considerado en cualquier paciente con dificultad respiratoria aguda y sospecha de neumotórax espontáneo. La CPAM tipo 4 puede aparecer como un quiste único que asemeje a una bulla espontánea o un neumotórax masivo.

PALABRAS CLAVE: Pediatría; Malformación pulmonar congénita; Neumotórax.

INTRODUCTION

Congenital pulmonary airway malformation (CPAM), previous known as congenital cystic adenomatoid malformation (CCAM), is a pulmonary disease with a reported incidence between 1:25,000 and 1:35,000[1]. However, there is new evidence suggesting a much higher incidence[2].

It results from abnormalities of branching morphogenesis of the lung, with molecular mechanisms still unknown but that may include an imbalance between cell proliferation and apoptosis during morphogenesis[3].

Patients without prenatal diagnosis can present various clinical pictures, from an immediate postnatal respiratory failure to an occasional finding on chest radiography. It is often misdiagnosed as a persistent and localized pneumothorax without any further pathological diagnosis[4]. Malignancy and frequent airway infections are major concerns in these patients[5].

In patients with CPAM that is causing any respiratory symptoms, surgical resection rather than observation is recommended, considering that resected cysts should be carefully examined for evidence of malignancy[6].

In the vast majority of infants with CPAM, however, surgical excision is curative and the prognosis is excellent[7-9].

CPAM rarely presents as an image compatible with persistent and localized spontaneous pneumothorax in the neonatal period. Our objective is to report one of these rare cases.

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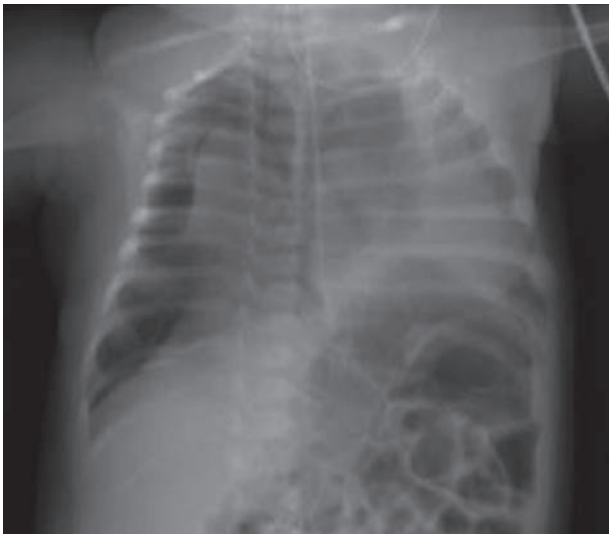

Figure 1. Right tension pneumothorax with associated mass effect of mediastinal shifting.

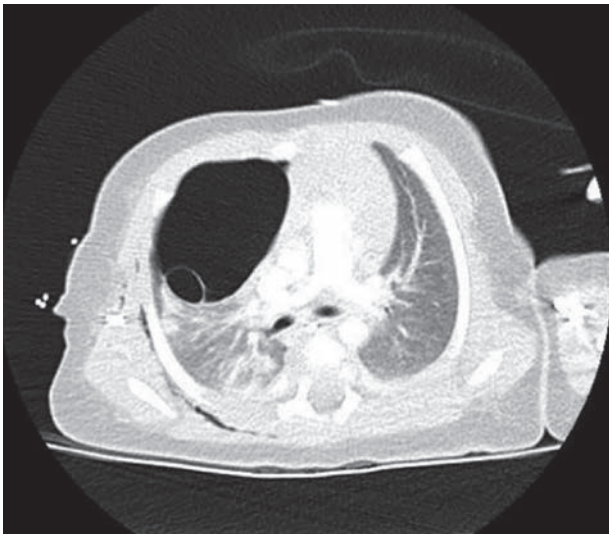

Figure 2. CT scan showing a single large septal bulla in upper-middle right lobe that could be related to congenital pulmonary airway malformation.

CLINICAL REPORT

A 2-month-old male infant was brought to our emergency department with acute respiratory symptoms and suspicion of a right tension pneumothorax on chest X-ray.

The patient was born to a 35-old woman G3/P2/A1 by spontaneous vaginal delivery at 40 + 1 weeks. His birth body weight was 3,375 g and prenatal ultrasonography and postnatal physical examinations showed no abnormalities. Apgar scores were 9 at 1 minute and 10 at 5 minutes. At 2 months of age he suddenly presented a respiratory failure. He was admitted to a secondary hospital, afebrile, with heart rate 160/min, blood pressure 77/54 mmHg, breathing rate 38/min, an oxygen saturation of 86% and a metabolic acidosis with a blood pH of 7.18 (lactic acid 8.8 mmol/L) requiring continuous positive airway pressure (CPAP) for management of acute respiratory failure.

After admission in Intensive Care Unit, a chest radiograph showed an image compatible with right tension pneumothorax (Fig. 1). It was drained by needle aspiration (about 40 ml air was removed), but radiological image of pneumothorax persisted with mediastinal shifting and subcutaneous emphysema. A chest tube drainage was then performed (6 Fr) under aspiration of -10 cmH_2O. The chest tube leaked air constantly and, again, despite chest drain, radiological image did not resolve.

The patient was then transfer to our tertiary hospital with pediatric surgery. Suspecting a pulmonary malformation a CT scan was performed. It showed the correct position of chest tube and a single septal large bulla in the upper-middle right lobe that was related to a congenital pulmonary airway malformation (Fig. 2).

An open surgical approach was decided. The intraoperative findings consisted of a giant bulla in the right upper lobe (Fig. 3). As the rest of the right lobe showed a normal parenchyma, an atypical pulmonary resection was performed.

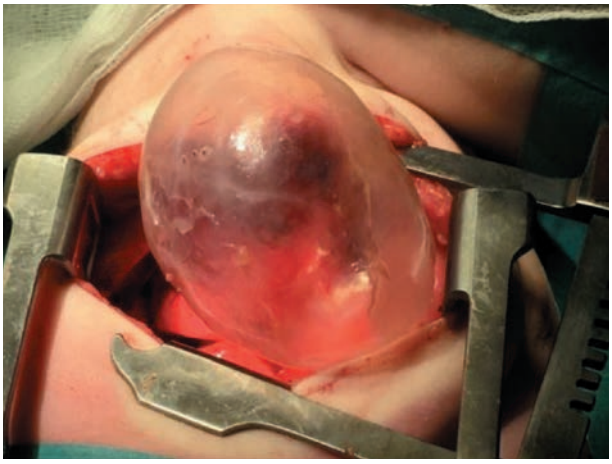

Figure 3. Appearance of the surgical site: bulla 6 x 4 cm in size.

A gross histological examination showed a large bulla measuring 6 x 4 cm in size with thin walls compressing the normal pulmonary parenchyma (Fig. 4). Microscopically, a type 4 CPAM lesion composed of large cysts with thin walls and a flattened alveolar-type epithelial lining. There were few cuboidal and mesenchymal cells. There were no malignant proliferation or other immature tissues.

The post-surgical condition of the patient was uneventful, a subsequent chest radiography showed complete resolution of the original image. No more respiratory episodes were recorded during two years of follow-up.

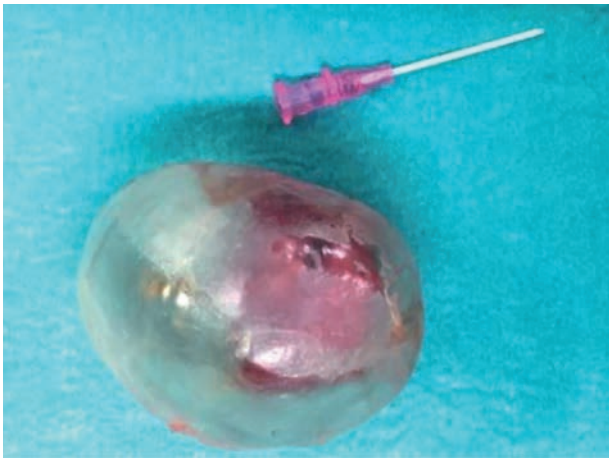

Figure 4. Macroscopic appearance of the specimen after resection.

DISCUSSION

Congenital lung disease in children is rare with various clinical presentations. Pulmonary sequestration, congenital pulmonary airway malformation (CPAM), congenital lobar emphysema, and bronchogenic cysts are the four major congenital cystic lesions founds in the lungs[4].

CPAM is histopathologically characterized by the lack of normal alveoli and an excessive proliferation and cystic dilatation of terminal respiratory bronchioles with various types of epithelial lining[10].

The precise etiology is unknown. It may present as a non-hereditary lesion or arise in association with certain genetic syndromes. The family history should be explored in detail for cancers and cystic lesions that might suggest familial pleuropulmonary blastoma syndrome including renal cystic disease, small bowel polyps, childhood cancers or dysplasia and a history of spontaneous pneumothorax[11]. In present case, the patient is a healthy 2-month-old male infant without prenatal diagnosis or family history.

The accepted classification is the one described by an american military pathologist, J.T. Stocker, based on the size of the cyst noted in resected lung specimens and at autopsy. He initially described three subtypes in 1977 and later included two further subtypes in 2002 (types 0 and 4). The most common lesions are type 1[12].

Type 4 CPAM lesions comprise 5 to 10 percent of CPAMs[13] are large peripheral thin-walled cysts, often an asymptomatic incidental finding or, as in the present case, as sudden respiratory distress from spontaneous pneumothorax[10]. Type 4 lesions are histopathological similar to cystic pleuropulmonary blastoma and sometimes may result in misdiagnosis[14]. Pathological report of our patient didn't show malignant cells, however it is important to emphasize that there should be a strong suspicion for malignancy in any infant presenting with pneumothorax and CPAM[3,15,16].

Apart from a chest radiograph, symptomatic patients should be further evaluated with advanced thoracic imaging (CT or MRI) to further define the lesion and distinguish it from other developmental anomalies of the lung, as part of preoperative planning[17]. In this case, despite placement of a chest drain, radiological image of right tension pneumothorax persisted. The sudden enlargement of the air cyst caused respiratory distress and changes similar to pneumothorax in the images. Not achieving the decompression of the pneumothorax reveals the need for distinguish giant congenital lung cysts of the pneumothorax. In addition, there is evidence that transthoracic catheter drainage can be an effective interim management for symptomatic newborn infants who require emergency surgical resection of large CPAM[18].

The postnatal management of CPAM depends on whether the patient has respiratory distress or is asymptomatic. In symptomatic patients, CPAM is treated by surgical resection. For infants and children who remain completely asymptomatic, the decision between surgical management and observation is controversial[19,20]. The main rationale for surgery is that it eliminates the risks of future infection and the potential for malignant transformation. However, the magnitude of these risks is poorly established. The risk of malignancy is extremely low except in type 4 CPAMs[5]. In this patient, the radiological image confirming a large symptomatic bulla probably related to a CPAM prompted us to an early surgical approach.

CONCLUSION

In summary, CPAM should be considered in a patient with sudden respiratory distress and image compatible with spontaneous pneumothorax. Type 4 CPAM may appear like unique lung cyst mimicking a spontaneous bulla or a massive pneumothorax, so we should consider this entity in these patients so prompt recognition can facilitates management and counseling.

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