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CASE REPORT

Malformation lung cystic adenomatoidea

Malformación adenomatoidea quística pulmonar

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ABSTRACT

Introduction: cystic adenomatoid malformation is an alteration of lung development caused by the replacement of the small airway and lung parenchyma by cysts.
Objective: describe a case with a diagnosis of cystic adenomatoid malformation.
Case presentation: 16-year-old male adolescent, product of a pregnancy with low obstetric risk, normal delivery at 38,4 weeks, with a personal pathological history of bronchial asthma, surgery for glaucoma at age 13, surgery for umbilical hernia at 12 years old, no history of previous income. On this occasion he went to the emergency room due to having a dry, sporadic cough that had been going on for approximately 15 days. In addition to sometimes presenting pain in the tip of the side, an X-ray was performed, confirming a radiopaque image in the left lower lobe, deciding to admit him for better study and treatment.
Conclusions: cystic adenomatoid malformation is a rare anomaly, so it is important to keep in mind that any patient with symptoms and signs suggestive of this disease receives complete care to guarantee an early diagnosis. The role of radiological diagnosis is key to fully characterize MCPs. The characterization of the vascular contributions in the radiological report is of great importance for surgical planning.

Keywords: Congenital Bronchopulmonary Malformation; Cystic Adenomatoid Malformation; Teenager.

RESUMEN

Introducción: la malformación adenomatoidea quística, es una alteración del desarrollo pulmonar causado por el reemplazo de la pequeña vía respiratoria y el parénquima pulmonar por quistes.
Objetivo: describir un caso con diagnóstico de malformación adenomatoidea quística.
Presentación del caso: adolescente masculino de 16 años de edad producto de un embarazo bajo riesgo obstétrico, parto eutócico a las 38,4 semanas, con antecedentes patológicos personales de asma bronquial, operado de glaucoma a los 13 años, operado de hernia umbilical a los 12 años, sin historia de ingresos previos. En esta ocasión acude a cuerpo de guardia por presentar tos seca, esporádica, de aproximadamente 15 días de evolución. Además de presentar dolor en punta de costado en ocasiones, por lo que se le realiza un Rx constatándose imagen radiopaca en lóbulo inferior izquierdo decidiéndose su ingreso para un mejor estudio y tratamiento.
Conclusiones: la malformación adenomatoidea quística es una anomalía poco frecuente por lo que es importante tener en cuenta que cualquier paciente con síntomas y signos sugestivos de esta enfermedad reciba atención completa para garantizar un diagnóstico precoz. El papel del diagnóstico radiológico es clave para caracterizar de forma completa las MCP. Es de gran importancia la caracterización de los aportes vasculares en el informe radiológico de cara a la planificación quirúrgica.

Palabras clave: Malformación Congénita Broncopulmonar; Malformación Adenomatoidea Quística; Adolescente.

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INTRODUCTION

Pulmonary malformations correspond to rare developmental anomalies of the respiratory system, including congenital pulmonary airway malformation (CPAM), formerly known as cystic adenomatoid malformation, pulmonary sequestrations, hybrid lesions, and congenital lobar emphysema.[1]

They are a relatively rare finding, occurring approximately 1 in 15 000 live newborns, although their diagnosis is considered to be increasing mainly due to improved ultrasound technology and the widespread use of obstetric ultrasound.[2]

Congenital pulmonary airway malformation (CPAM), formerly called cystic adenomatoid malformation, is a hamartomatous disorder of lung development caused by the replacement of the small airway and lung parenchyma by cysts.[3]

Cystic adenomatoid malformation is a rare anomaly of lung development, with an incidence of approximately 1 in 25 000 to 35 000 pregnancies and 0,15 to 1,7 % of the general population, respectively. It corresponds to the most common congenital pulmonary. Malformation.[4,5]

The clinical expression of CMM is variable and includes asymptomatic patients, rarely fetal or neonatal death, and localized recurrent pulmonary infections. The usual treatment is surgical excision. However, there is discussion in asymptomatic cases with a lesion usually detected prenatally.[6]

Prenatal ultrasound allows an early approach, identifying the malformation in 10 % of cases at birth. In comparison, 14 % are diagnosed in adolescence with a radiological finding or chronic lower airway symptoms.[3]

The presentation in adulthood is infrequent, and it is diagnosed from an imaging finding, such as cavitated lesions in the chest X-ray, or in relation to recurrent pulmonary infections.[7]

Few cases have been reported on the subject in our country, which was the main motivation for conducting this research.

Objective: To describe a case with a diagnosis of cystic adenomatoid malformation.

CASE REPORT

Male adolescent aged 16 years, product of a low obstetric risk pregnancy (LORP), euthyroid delivery at 38,4 weeks, birth weight 8 lbs, without pre-, peri-, or postnatal complications, Apgar 8/9. With a personal pathological history of bronchial asthma, operated for glaucoma at 13 years of age, operated for umbilical hernia at 12 years of age, and no history of previous admissions. On this occasion, he came to the emergency room with a dry, sporadic cough of approximately 15 days. In addition to presenting occasional flank pain, an X-ray was performed, and a radiopaque image was found in the left lower lobe. It was decided that he should be admitted for further study and treatment.

Diagnostic impression: left lung tumor vs. community-acquired pneumonia with atelectasis component in the left lower lobe.

Laboratory tests: Laboratory: Hemogram: Ht 0,45, Leukogram 12,5 x 109, Platelets 205 x 109, Erythrocyte sedimentation rate 5 mm/L.

Rx findings: Non-homogeneous opacity in the left basal hilum region of well-defined, lobulated contours that does not blur the cardiac contour (figure 1).

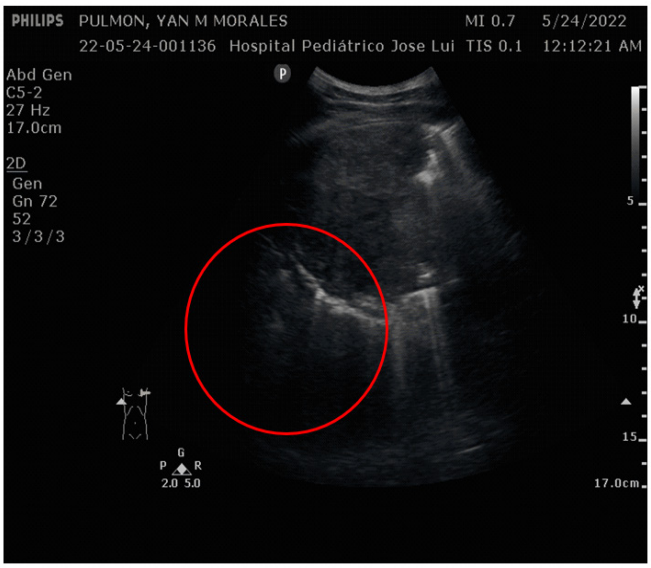


Figure 1. PA view of the thorax

Results of the US of the left pulmonary base: Echogenic, homogeneous mass with well-defined contours, without calcifications or areas of cavitation in its interior, accompanied by a small pleural effusion that enters the large fissure (figure 2).

CT (Computed Axial Tomography with EV contrast) results: It shows a thick hyperdense image, with density ranging between [40 and 70uh] of well-defined contours, polylobed, measuring in axial section 70x100mm and 150mm of caudal skull diameter in coronal MPR reconstruction (figure 3).

Pathology findings: left pulmonary pneumectomy showing at the level of the lower lobe and part of the upper lobe, chronic

inflammatory process, rich in histiocytes, presence of hyperplasia of bronchiolar elements, smooth muscle and type II pneumocytes, some cysts, the largest of 2 cm with detritus. There is no evidence of a neoplastic process. The described features correspond to a congenital cystic adenomatous pulmonary malformation.

Management: The patient was scheduled for surgery, approached by left posterolateral thoracotomy, with anatomical lobectomy of the left lower lobe of approximately 7 cm × 4 cm × 2 cm. He continued with a follow-up study by the Department of Surgery, ruling out any other type of alteration; he evolved satisfactorily and was discharged after four weeks. Currently, he is asymptomatic, and in his outpatient follow-up, there are no respiratory symptoms related to any complication.

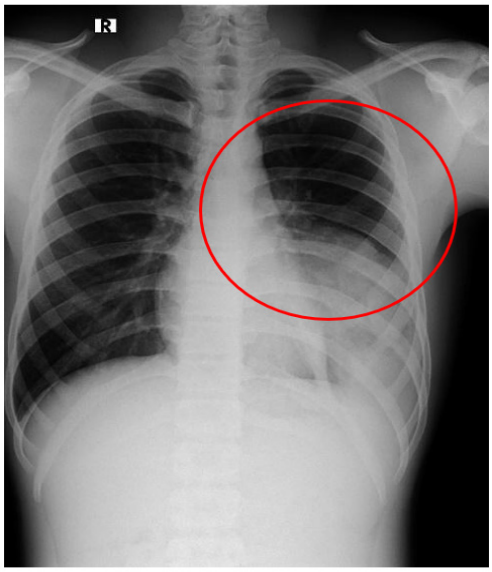


Figure 2. US of left pulmonary base

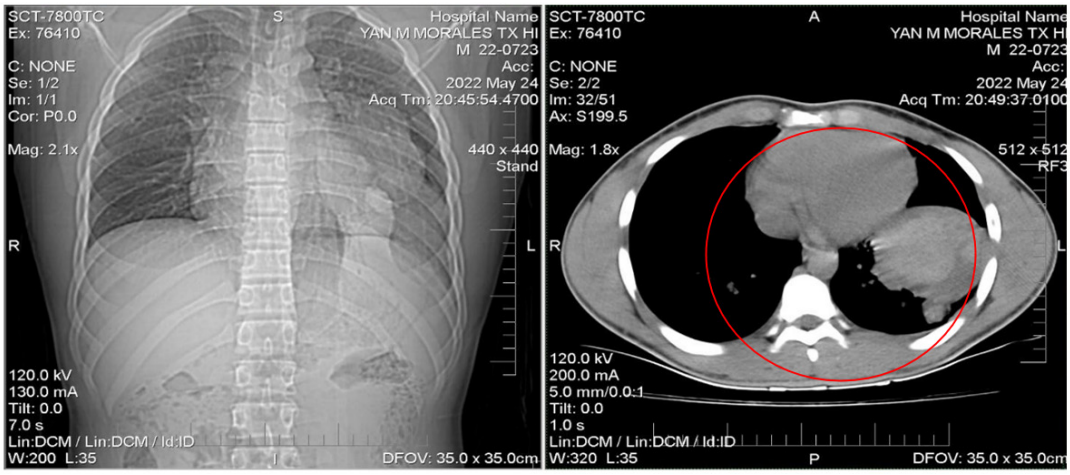


Figure 3. Chest CT scan

DISCUSSION

Congenital malformations of the pulmonary airway, particularly the so-called cystic adenomatoid malformation (CCAM) or Congenital Cystic Adenomatoid Malformation, are the most common intrathoracic tumor observed by prenatal ultrasound. It develops during the first six weeks of gestation. It represents 10 to 25 % of congenital pulmonary alterations and 95 % of cystic lung diseases. It is a sporadic non-hereditary disease with a certain predisposition for male sex, as in the case presented, also associated with some genetic syndromes such as trisomy 18 and hereditary renal dysplasia.[4,8]

It is characterized by adenomatous proliferation of the respiratory bronchioles, cysts intercommunicating with the bronchial tree, and a defect in the maturation and formation of terminal alveoli.[4]

Lesions vary widely in size and may affect an entire lobe or segment or involve an entire lung. They may be bilateral, although they occur more frequently in the lower lobes. Eighty-five to 95 % of cases occur in a single lobe.[1] In the case presented, the multicystic lesion encompasses the lower lobe and part of the upper lobe of the left lung.

According to Stocker in 2002, the five different types of MCVAP occur at different tracheobronchial tree levels and at different lung development times. Type 1 is the most frequent, representing 65 %, and is associated with the best overall prognosis, remaining undetected until after birth. Type 2 is the second most frequent, representing up to 25 %, and the other types represent the remaining

15 %. The differential diagnosis includes intralobar pulmonary sequestration, bronchogenic cysts, and congenital lobar emphysema.[4]

Clinically, it is considered a late presentation if the malformation debuts after 6 months. In infancy it presents as respiratory failure, recurrent pulmonary infections or growth retardation. CMM occurs less frequently in adolescents and adults (17 %); the patient remains asymptomatic until diagnosed incidentally by chest X-ray detecting cystic lesions, or as recurrent pulmonary infections, spontaneous pneumothorax, hemoptysis or chest pain.[7] The case presented here is a male adolescent who was diagnosed by a radiological finding whose clinical picture was characterized by dry cough and flank tip pain at times.

Some studies speak of conservative management, especially for patients who remain asymptomatic. In these patients, serial controls of the lesions are performed using tomography to determine possible malignant degeneration. Surgical treatment is reserved for patients with symptomatology or postnatal complications. The surgical technique depends on the size and location of the lesion, with lobectomy and segmentectomy being the most frequently used.[4] In the case presented, it was resolved with pneumonectomy of the left lung.

The possibility of malignancy of congenital cystic lung lesions explains the tendency of many pediatricians and pediatric surgeons to take an active surgical approach even in the absence of symptoms. Two types of pulmonary neoplasms have been described in MCVAP: Pleuropulmonary Blastoma (BBP) and Bronchioloalveolar Carcinoma (BAC). BBP is a rare dysembryogenic neoplasm (0,35-0,65/100 000 live newborns), described in 1988, which can arise as a malignancy of a type 4 MCVAP. Ninety-four percent of cases have been described in children under 6 years of age, 25 % of them having a familial genetic predisposition. CBA is much rarer, only 21 cases have been described in association with type 1 MCVAP at a mean age of 20 years. Most of them have been diagnosed by chance when the cystic lesion was resected. CBA can progress to invasive mucinous adenocarcinoma. In the case presented, there was no evidence of a neoplastic process.

CONCLUSIONS

Cystic adenomatoid malformation is a rare anomaly, so it is essential to ensure that any patient with symptoms and signs suggestive of this disease receives comprehensive care to provide an early diagnosis. An adequate reading and interpretation of the thorax's radiological and tomographic images will guide the MCVAP diagnosis. Of great importance is the characterization of the vascular contributions in the radiological report, which is of great importance for surgical planning.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

AUTHORSHIP CONTRIBUTION

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Writing - revision and editing: Dania María García-Rodríguez, Dionis Ruiz-Reyes, Diana García-Rodríguez, Adriel Herrero Díaz, Ileana Beatriz Quiroga-López.

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