Criptogenic Organizing Pneumonia: A Case Report

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INTRODUCTION

Cryptogenic Organizing Pneumonia (COP) is a rare disease in pediatrics. It was described in 1985 as organizing pneumonia and its origin is cryptogenic because it has diverse etiologies; usually its presentation is subacute manifested with tachypnea, cough, dyspnea and hypoxemia. The disease is considered as a diagnosis of exclusion since the tomographic findings are variable. Also, the histopathology is important as it is required to confirm the diagnosis.

CLINICAL CASE

This report presents the case of a six-month-old female patient, product of the first pregnancy of her 18-year-old mother, with complete immunizations up to two months of age. Between the second and the third month of life, the patient stopped gaining weight gain, and presented dyspneic and wheezing events with diaphoresis associated with recurring breastfeeding. Because of this, she is taken to a first-level consultation where she received treatment with antitussives, expectorants, antipyretics, antiviral drugs and antibiotics on three occasions. At the fourth month, the patient is hospitalized due to the presence of wheezing without fever and respiratory difficulty treated with bronchodilators and inhaled steroids. Because the intermittent tachypnea and wheezing persisted, adding cyanotic events, she was taken to urgent care at six months of age. The physical examination showed the following parameters: weight 5 kg, height 60cm, heart rate 170 bpm, respiratory rate 52 bpm, temperature 36.6°C, 87% saturation, peripheral cyanosis, thorax with intercostal retractions, suprasternal retractions, nasal flaring, bilateral bronchoalveolar rales, expiratory wheezing, dynamic precordium with systolic murmur Grade II/VI, hepatomegaly 2x2x1cm, and capillary refill of 2 seconds. Upon her admission a chronic lung disease approach is performed as well as laboratory tests that reported Hemoglobin 15.5 g/dl, Hematocrit 43%, Leukocytes 8800 x10³/ml, Neutrophils 46%, Lymphocytes 25%, Monocytes 11%, Platelets 245,000, IgG 1020 mg/dl, IgM 114 mg/dl, IgA 60mg/dl, PCR 0.34 mg/dl, antinuclear antibodies and negative anticytoplasmic antibodies, HIV-negative. Thorax Radiography (Figure 1).
Electrocardiogram with biventricular growth, echocardiogram with interatrial septum growth of 4mm, pulmonary pressure of 26 mmHg. Gastroesophageal reflux approach with negative pH meter for reflux. The patient persisted with increased work of breathing and wheezing episodes in spite of bronchodilators and inhaled steroids; therefore, a thoracic CAT scan is performed (Figures 2-4).

Because of the persistent clinical findings of poor response and the observed results in the imaging as an approach for chronic lung disease with interstitial condition, it was decided to perform a bronchoscopy and a lung biopsy. These tests reported hypersecretion of mucus, negative cytology of Bronchoalveolar Lavage (BAL) for lipophages, negative bronchial culture, and bronchial brushing with no specific pattern. From the histopathological standpoint, collapsed and over-distended lungs with epithelial hyperplasia were observed as well as regional desquamation of alveolar macrophages with no specific pattern of...
lung damage. The immunohistochemistry in biopsy showed CD68+ in macrophages and CD34+ in vessels. The patient was dispatched to her home being with oxygen therapy, prednisolone 0.5mg/kg and inhaled therapy.

**Discussion**

The COP belongs to the group of interstitial lung diseases characterized by abnormal remodeling of the distal airway that can cause alterations in the ventilation and gas diffusion, it is the idiopathic form of an organizing pneumonia [1]. While the prevalence in pediatrics [2] is unknown, the incidence series of adults has been observed from 2% occupying up to 7% of pulmonary interstitial diseases [3,4].

It can be associated with viral infections that play an etiologic role in lung transplants, radiation, chemotherapy, medications, and autoimmune diseases [5-7]. In this clinical case, the patient presented at two months of age ARIs of undetermined origin, likely viral. There are isolated cases of COP due to broncho aspiration associated with gastroesophageal reflux disease [8], but this was ruled out in our patient because of the negative pH metry for reflux and the absence of lipid-loaded macrophages in the BAL. The increase in the work of breathing is the most common manifestation that may occur in 75-93% of cases. On the other hand, cough, dyspnea and hypoxemia are more common suggestive data in patients with interstitial lung disease, and pulmonary insult [2]. In this case, due to the chronic respiratory symptoms the patient was approached for a chronic lung disease from the interstitial [9] sphere. There are isolated fulminant COP cases described that have come to require ventilation with extracorporeal membrane oxygenation machine as part of the treatment [10]. The BAL usually show an increase in the cellularity with predominance of lymphocytes and a slight elevation of neutrophils [11]. From the histopathological standpoint, it is characterized by myofibroblasts deposits and peribronchial connective tissue involving the small airways, alveolar ducts and chronic inflammation with cellular infiltration of the alveolar wall [12]. The immunohistochemistry reported the expression of CD34+, a cell with a potentialized surface in angiogenesis [13,14] processes and macrophages with expression of CD68+ observed in pediatric patients with severe pneumonias [15] and in macrophages of idiopathic interstitial pneumonias [16]. The Computed Axial Tomography is indispensable and the best diagnostic tool in patients with interstitial disease with increased work of breathing of torpid evolution. There is no specific tomographic pattern; the dominant pattern are multiple areas of consolidation or ground-glass located in the subpleural and peri bronchovascular areas, predominantly in both bases [17] such as in the present case. The tomographic patterns can have migration tendencies and affect other lobe [18] sites. The differential tomographic diagnosis includes lymphoma, eosinophilic lung diseases, vasculitis, and sarcoidosis [3]. There are cases described with spontaneous resolution [19], but immunosuppressive pharmacotherapy has been reported to be useful in interstitial lung diseases in child [2] as in our case.

**Conclusion**

Criptogenic organizing pneumonia is uncommon in pediatrics, its etiology is diverse, being the post infectious origin the most common cause. The clinical case represents the approach for chronic lung condition with interstitial disease, observing the most frequent respiratory interstitial symptoms in infants, and correlating with the CAT and the histopathological report. The utility of steroids in early stages can decrease interstitial damage and improve prognosis. Interstitial diseases should be suspected in healthy infant patients with unfavorable evolution.

**Conflict of Interest**

The authors declare they have no conflict of interest.

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