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Case Report

Hypersensitivity Pneumonitis, a Rare Diagnosis in a Paediatric Patient

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Abstract

Background: Hypersensitivity pneumonitis (HP), also known as extrinsic allergic alveolitis, is a rare immune-mediated inflammatory lung disease, particularly uncommon in pediatric patients.

Case Presentation: A six-year-old girl presented with odynophagia, dry cough, and sudden respiratory distress. Pulmonary auscultation revealed decreased air entry bilaterally associated with crackles. Despite initial treatment, her condition deteriorated, prompting admission for further investigation. Imaging revealed bilateral diffuse micronodules on chest radiography and ground-glass opacities on computed tomography. Bronchoalveolar lavage demonstrated lymphocytic alveolitis with a reversed CD4+/CD8+ ratio, suggestive of HP. Positive avian precipitins supported the diagnosis.

Discussion: Diagnosis of HP in pediatric patients is challenging due to its rarity and nonspecific symptoms. Clinical suspicion, supported by imaging and immunological tests, is crucial for accurate diagnosis.

Conclusion: This case underscores the importance of considering HP in pediatric patients with respiratory symptoms, especially in the context of environmental exposure. Early diagnosis and antigen avoidance are essential for favorable outcomes in HP.

Keyword: *Hypersensitivity Pneumonitis; Interstitial Pneumonitis; Extrinsic Allergic Alveolitis; Paediatric*

Introduction

Hypersensitivity pneumonitis, also known as extrinsic allergic alveolitis, is a rare condition at paediatric age [1]. A report from 2011 estimated a prevalence in Denmark of 4 per 1.000.000 children [2], however, the absence of uniform diagnostic criteria for the disease in children hinders an accurate estimate [3].

HP is an immune-mediated inflammatory response to environmental antigens [4]. In paediatric cases, antigens are usually related to domestic environment or pastime activities [1].

Clinical course is related to type, intensity and duration of exposure in a susceptible host [3]. Due to the overlap between classical forms, practically, they are grouped in acute/subacute and chronic HP [1].

Acute/subacute HP manifests itself with recurrent episodes of cough, dyspnoea or fever, and bronchial obstruction and crackles on pulmonary auscultation [2]. Due to nonspecific signs/symptoms, it's usually misdiagnosed as asthma or an acute lower airways viral infection [5].

Chronic HP presents with progressive respiratory symptoms such as a chronic cough associated to exacerbations with worsening of dyspnoea. On physical examination, crackles are present and, in some cases, clubbing is observed [1].

Chest radiograph is usually nonspecific with reticular, linear or ground-glass opacities, micronodules or a normal exam. Computed tomography (CT), given its sensitivity, is considered key for the evaluation of interstitial lung diseases. In acute/subacute HP, despite the possibility of CT being normal, predominant features are ground-glass opacities or ill-defined small centrilobular nodules. In chronic HP, due to fibrosis progression, CT can reveal reticulation and parenchymal distortion [6].

Laboratory studies are characterized by leukocytosis, elevated erythrocytes sedimentation rate (ESR), C-reactive protein (CPR), and immunoglobulins. Eosinophilia may be present. The presence of specific IgG antibodies in serum can be documented in patients with HP [3]. However, it is not, per se, a marker of the disease or excluded by its absence. Bronchoscopy with bronchoalveolar lavage (BAL) is characterized by lymphocytosis (often>50%) and relative predominance of CD8+ with a decreased CD4+/CD8+ ratio [6].

Diagnosis is suggested by respiratory signs/symptoms, positive clinical history and imagological features. Some laboratory parameters, bronchoscopy with BAL and pulmonary function can give further support to diagnosis [3].

The avoidance of the trigger agent is crucial for a successful treatment [3]. The prognosis of acute HP is usually good with spontaneous resolution after antigen removal. In severe cases, supportive treatment with oxygen or corticosteroids may be needed [6]. In chronic HP, due to prolonged exposure, the disease tends to progress to progressive pulmonary fibrosis [3].

Case Report

A six-year-old girl arrived at the emergency room presenting with odynophagia for two days and a dry cough that had worsened with sudden respiratory distress during that day. In addition, she presented anorexia and decreased activity, without fever and expectoration. There was no history of recent travel or contact with sick people including those with COVID-19.

Physical examination revealed an afebrile girl with pallor, exophthalmos and poor peripheral perfusion. Additionally, it revealed tachycardia, tachypnoea, severe and global retraction and hypoxemia needing oxygen. On pulmonary auscultation, decreased air entry bilaterally associated with crackles was noted. No organomegaly or adenopathy was observed.

Regarding her past medical history, reference to an hospital admission the prior week due to viral pneumonia without agent identification associated with hypoxemia with rapid improvement.

At first approach, oxygen therapy, methylprednisolone plus inhaled salbutamol, and ipratropium bromide were started. As a complementary study: chest radiography showed bilateral diffuse micronodules (Figure 1); venous blood gas showed pH of 7.39, pCO_2 of 38 mmHg, pO_2 of 29 mmHg, HCO_3^- of 22.1mEq/L and BE of -2; complete blood count revealed leucocytosis with neutrophilia, thrombocytopenia, elevated ESR of 42 mm/h (N: 0 - 10 mm/h), CPR count of 5.1 mg/L (N: < 5 mg/L) and normal renal function and liver enzymes other than lactate dehydrogenase of 560 U/L (N: 125 - 220 U/L).

Despite optimized therapy she manifested a poor response and needed rising oxygen supplementation (FiO_2 36%), so empiric antibiotic treatment was started with ampicillin and clarithromycin. She was admitted to the Paediatrics ward for close monitoring, treatment and further investigation.

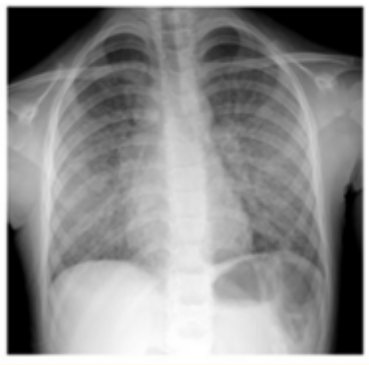

Figure 1: *Admission chest-x-ray showing bilateral diffuse micronodules.*

The clinical history was reviewed, in particular incidence on environmental exposure. Her family had a house cat for several years and recently, in the last month, they acquired two birds (a parakeet and a cockatoo).

An autoimmune work up showed negative antinuclear screen and anti-neutrophil cytoplasmic antibodies, normal complement (C3 and C4) levels, and normal values of immunoglobulins A, E, M, G subclasses. HIV was negative. Furthermore, lymphocyte immunophenotyping showed an increase of CD4+, CD8+ and B cells. Apha-1 antitrypsin was slightly decreased, 88 mg/dl (N: 90 - 200 mg/dl).

To clarify the chest radiography pattern, a CT was performed and showed bilateral and diffuse ground glass densification, compatible with exuberant and nonspecific interstitial pneumonitis (Figure 2). The child underwent flexible bronchoscopy with BAL that showed normal anatomy and BAL revealed lymphocytic inflammatory cell pattern (lymphocyte of 52,37% (N: 0 - 15)) with inversion of the CD4+/CD8+ ratio (0,54), suggestive of lymphocytic alveolitis in the context of Hypersensitivity pneumonitis.

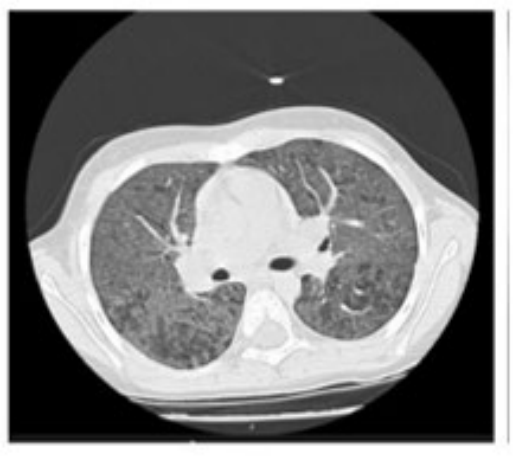

Figure 2: *CT scan showing bilateral and diffuse ground glass densification, compatible with an exuberant and nonspecific interstitial pneumonitis, no other pleuroparenchymal alterations were founded as well as no alterations in the mediastinum, adenopathy, pleural or pericardial effusion.*

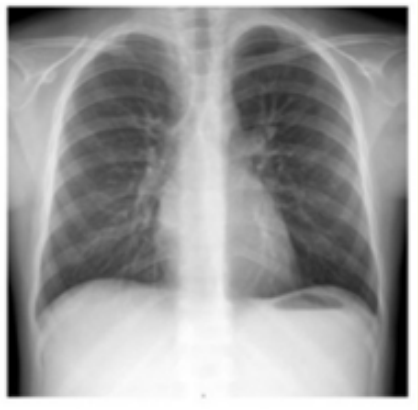

Figure 3: *Post-acute chest-x-ray showing resolutions of interstitial alterations, without evidence of micronodules.*

Additionally, mycological, bacteriological, mycobacteriological, as well as viral and *Pneumocystis jirovecii* tests were performed and came back negative. Specific IgG antibodies were investigated and avian precipitins (feathers, parakeet, canary, cockatiel, parrot, chaffinch) were positive 27.4 mg/L (N < 15).

The remaining exams were negative, including respiratory virus by polymerase chain reaction, SARS-CoV-2, serology for *Mycoplasma pneumoniae, Chlamydia pneumoniae, Chlamydophila psittaci*, *Legionella* antigen urinary, *Legionella pneumophila, Aspergillus fumigatus* and tuberculosis - interferon gamma release assay test.

Due to respiratory symptoms, temporal relationship with antigen exposure, features on chest radiography and CT, BAL suggestive of lymphocytic alveolitis, positive avian precipitins and absence of other causes, the diagnosis of HP was assumed.

During admission, she maintained treatment with ampicillin, clarithromycin, methylprednisolone plus inhaled salbutamol and ipratropium bromide. Oxygen therapy was suspended on the second day with clinical improvement, without cough or respiratory distress.

After hospital discharge, she performed avoidance of the causative allergen and maintained corticotherapy with prednisolone. For five weeks on a weaning schedule, then she started fluticasone propionate.

During follow-up, respiratory function tests were performed and were normal. One year after, CT scan was repeated and showed no significant alterations.

Currently she is not under medication and maintains antigen eviction, without recurrence of the disease.

Discussion

Our clinical patient´s presentation, on the first instance, was interpreted as viral pneumonia. Another exacerbation in a short period and the fast improvement, made us review the history and look for possible triggers, in this case, the birds present at her home.

In this clinical case, CT revealed a ground-glass pattern highly suggestive of interstitial disease. Laboratory studies showed almost all the cited typical changes except eosinophilia. Considering the clinical manifestations, time of evolution, and imaging features, we concluded that our patient presented an acute/subacute HP.

In concern to the prognosis expected in these case, being an acute/subacute HP, it was expected that, with the avoidance of the trigger antigen, the symptoms and pulmonary alterations would recede just like it happened.

Conclusion

Interstitial lung diseases like HP, despite being rare in paediatric age, should be considered in cases where clinical response to treatment and evolution is not as expected for the diagnosis presumed.

In this particular disease, the earlier diagnosis and shorter time of exposure to the causal antigen, would confer greater probability of symptoms and lung alteration regression and lower the potential of evolution to chronic form.

Antigen avoidance is key to successful treatment, however, some patients might benefit from corticosteroid therapy in the face of severe manifestations of the disease.

Conflict of Interest

The authors declare no conflicts of interest in conducting this work.

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