

Chronic Eosinophilic Pneumonia With Mucous Plugs in a Child

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Summary. Chronic eosinophilic pneumonia is a rare cause of chronic lung disease in children. A 7-year-old girl who attended our clinics with cough and sputum lasting for 5 years, has been evaluated for bilateral alveolar infiltration and ground-glass opacities. Peripheral eosinophilia was detected in total cell blood count. Flexible bronchoscopy showed mucous plugs. Bronchoalveolar lavage fluid and cell block of mucous plugs determined hypereosinophilia. Chronic eosinophilic pneumonia was confirmed after the elimination of other eosinophilic lung diseases and the case was accepted to be idiopathic. She showed a dramatic response to oral corticosteroids. This is the first reported case of chronic eosinophilic pneumonia presenting with mucous plugs in children described to date in the literature. *Pediatr Pulmonol.* 2010; 45:1040–1042. © 2010 Wiley-Liss, Inc.

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INTRODUCTION

Eosinophilic lung diseases are a diverse group of pulmonary disorders associated with peripheral or tissue eosinophilia. Asthma (chronic eosinophilic bronchitis), allergic bronchopulmonary aspergillosis, simple pulmonary eosinophilia (parasitic infections and drug reactions), eosinophilic vasculitis, and eosinophilic pneumonia (acute, chronic, and drug induced) are the most common pulmonary eosinophilic syndromes.^{1–6} Chronic eosinophilic pneumonia (CEP) is an extremely rare disease in childhood.^{7–9} We report a CEP with eosinophilic mucous plugs in a 7-year-old girl who presented with respiratory symptoms and typical radiological findings.

CASE REPORT

A 7-year-old girl was admitted to our hospital with a history of persistent cough and sputum for 5 years. The patient has been treated elsewhere for asthma with fluticasone propionate, salmeterol and montelukast during the past 5 years. She had no history of drug use, chronic diarrhea, vomiting, and chronic infection rather than respiratory system. There was no history of family asthma, atopy, and allergic diseases. Her respiratory rate was 30 breaths/min with no fever. She had wheezing and crackles bilaterally on chest auscultation. Lung function tests showed a vital capacity of 53% of predicted values, a forced expiratory volume of 57% of predicted values.

The total blood cell count was 10,500/mm³ with 21% eosinophils on differential. There was no atypical cell. Chest radiography and high-resolution chest computed tomography showed bilateral alveolar infiltration and ground-glass opacities (Fig. 1). Flexible bronchoscopy (Olympus®, BF 3C160, 2.8.) detected whitish mucous plugs (Fig. 2).

Cell block and slides were prepared from bronchoalveolar lavage (BAL) fluid and stained with Hematoxylin & Eosin. Microscopic examination revealed 4.1×10^5 cells/ml with a differential count of 60% eosinophils, 20% macrophages, 5% plasma cell, 57% lymphocytes,

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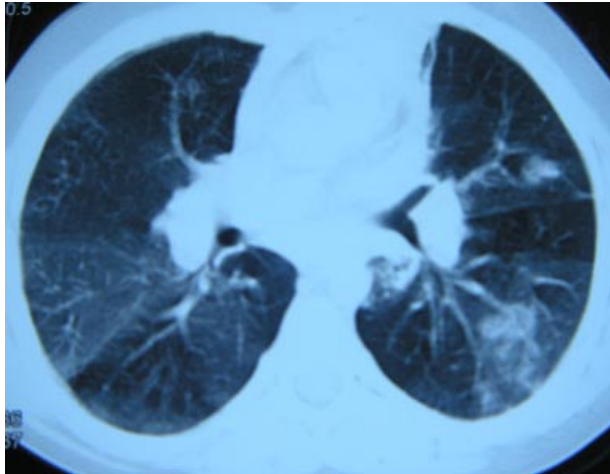


Fig. 1. High-resolution chest computed tomography showing bilateral alveolar infiltration and ground-glass opacities.

10% neutrophils with prominent numbers of Charcot-Leyden crystals (Fig. 3). There was no evidence of bacterial, fungal, tuberculosis or parasitic infection in BAL fluid. Lung function tests showed a vital capacity of 53% of predicted values, a forced expiratory volume of 57% of predicted values.

Differential diagnosis was made for eosinophilic lung diseases. Biochemical analysis, urine analysis and ultrasonography of the urinary system were normal. Serum IgE was 940 IU/ml. Allergen prick test was negative including aspergillus antigen. Aspergillus IgE and IgG, antinuclear antibody, anti double-stranded DNA and antineutrophil cytoplasmic antibodies were negative.



Fig. 2. Macroscopic bronchoscopy finding: Whitish mucous plug obstructing anterobasal segment of left lower lobe.

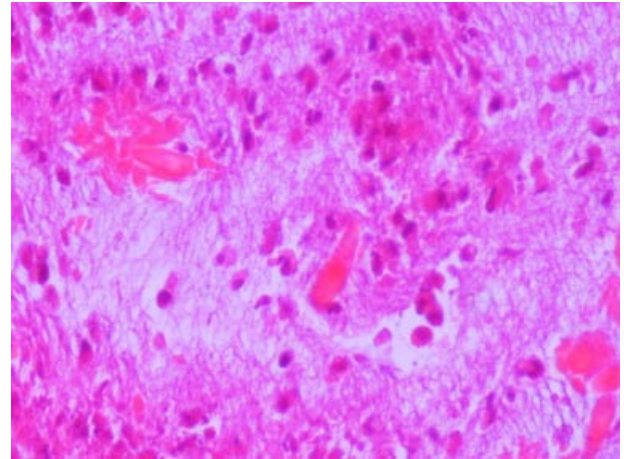


Fig. 3. Eosinophils and Charcot-Leyden crystals detected in the bronchoalveolar lavage fluid stained with Hematoxylin & Eosin.

Serologic tests for parasites were negative and stool microscopy was normal. Thus, CEP was confirmed after the elimination of other eosinophilic lung diseases and the case was accepted to be idiopathic. The patient was administered with 2 mg/kg/day of methylprednisolone therapy. The symptoms improved within 2 weeks of treatment. Peripheral eosinophilia and total serum IgE declined to 3.7% and 108 IU/ml respectively and there was complete resolution of the infiltrates in the lungs on computerized tomography after two months of therapy. Lung function tests improved to a vital capacity of 95% of predicted values, a forced expiratory volume of 98% of predicted values. Control bronchoscopy was performed and no mucous plugs were observed. Methylprednisolone was tapered after 2 months and discontinued at the end of 6 months. The patient was free of symptoms and signs under inhaler corticosteroid therapy for 6 months.

DISCUSSION

Eosinophilic lung diseases comprise a heterogeneous group of disorders. CEP is characterized by an abnormal accumulation of eosinophils in the lung.^{1,2,4,10,11} The diagnosis of CEP is generally made on the patient history and clinical presentation, association with peripheral eosinophilia, a high percentage of eosinophils in BAL cytology, and suggestive radiographic findings.^{2,3,11,12} In our patient, the diagnosis of chronic eosinophilic pneumonia was established by the presence of chronic pulmonary infiltrates, peripheral eosinophilia and the histopathologic evidence of BAL cytology with eosinophils.

CEP is a rare disorder, the exact prevalence of which remains unknown. CEP may affect every age group, with a mean age at presentation of 45 years. It is extremely rare in childhood.⁷⁻⁹ This is the first reported case of chronic

eosinophilic pneumonia presenting with mucous plugs in children described to date in the literature. Associated symptoms are usually dry cough, fever, sputum production and dyspnea. Clinical examination may show wheezing, crackles, and rhonchi. All of these symptoms are non-specific and contribute to a delay in diagnosis.^{3,10} Our patient also had history of persistent cough and sputum for 5 years. Peripheral eosinophilia is often above 1,000/mm³ in blood and BAL fluid always demonstrates abnormally high levels of eosinophils and Charcot-Leyden crystals.^{3,10,11} The most common radiological presentation of CEP is peripheral alveolar consolidation and ground glass opacities on chest radiography and computerized tomography.^{10,12} Our patient had peripheral alveolar consolidation and ground glass opacities either and complete resolution was established after steroid therapy.

The other causes of eosinophilic lung diseases should be eliminated for differential diagnosis. The most common causes of pulmonary eosinophilia in childhood are parasitic infections and drug use. *Ascaris*, *Strongyloides*, *Ancylostoma*, *Toxocara*, *Schistosoma*, *Trichinella*, *Distomatosis*, and *Anuillulosis* are associated with eosinophilia and pulmonary infiltrates. The diagnosis relies on serologic tests and stool examination. The symptoms are usually more transient than in CEP.^{2,3,5} Several drugs may cause eosinophilic pneumonia. The most frequently involved drugs are ethambutol, non-steroidal anti-inflammatory agents, including salicylates and antibiotics, such as minocycline and cotrimoxazole. A detailed history with respect to drug consumption is always needed to exclude any potential pharmacological cause of eosinophilic pneumonia.^{3,6,10} Both allergic bronchopulmonary aspergillosis and CEP may present with pulmonary infiltrates and high total IgE levels. Although relatively rare, Churg–Strauss syndrome must be considered in differential diagnosis. Churg–Strauss syndrome is defined as airway obstruction, peripheral eosinophilia, and eosinophilic vasculitis of multiple organs. Anti-neutrophil cytoplasmic antibodies are frequently present in Churg–Strauss syndrome.^{2,3,10} Acute eosinophilic pneumonia results from acute hyper sensitivity to inhaled unidentified antigens. Patients present with acute respiratory insufficiency. The number of eosinophils in peripheral blood is usually normal.¹³

The treatment of CEP is based on oral corticosteroids. Respiratory symptoms, blood and BAL hypereosinophilia, and radiographic findings regress within a few days or a few weeks. There is no consensus on dosage and duration of therapy. High doses of corticosteroids are required initially and should be followed by a very progressive tapering. Most patients are treated for more

than six months, and often for 12 months. Survival is excellent, but some patients become steroid-dependant with multiple relapses and severe bronchial hyper-responsiveness. Inhaled steroids may be efficacious in some cases.^{3,10,11,14,15} The findings of our patient regressed in two weeks. Complete remission was achieved in two months and the treatment lasted six months. The patient is still under inhaled corticosteroid therapy for 6 months with no relapse. In conclusion, although CEP is rare in childhood, it must be included in the differential diagnosis of unexplained persistent cough with hyper-eosinophilia and alveolar consolidations.

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