

Chronic Eosinophilic Pneumonia in an 11-Year-Old Boy

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Abstract

Idiopathic chronic eosinophilic pneumonia is a rare and serious disease mostly encountered in female asthmatic patients in their fifth decade of life and rarely in children. Herein, we describe an 11-year-old boy presenting with clubbing of fingers and post-exertional cough without asthma and peripheral eosinophilia. He had had a restrictive pattern in his pulmonary function test that showed a dramatic response to corticosteroid therapy.

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Keywords • Chronic eosinophilic pneumonia • children • clubbing

Introduction

Idiopathic chronic eosinophilic pneumonia (CEP) is a rare but serious disease of childhood. Herein, we are going to report an 11-year-old boy who presented with drumstick fingers and interstitial infiltrates on chest radiograph.

Case Report

RJ, an 11-year-old boy visited his physician, because of abdominal pain last year. The presence of drumstick fingers led to further investigation. The only pulmonary symptom of RJ was post-exertional cough without wheezing or dyspnea. On physical examination, he appeared well except for digital clubbing and bronchial breathing with a few inspiratory crackles.

His height was 136.5 cm (within 3rd–10th percentiles) and his weight was 26.1 kg which was below the 3rd percentile.

The medical history was unremarkable except for recurrent episodes of sinusitis and post-exertional cough since four years of age. Chest x-ray revealed pneumonic infiltrations, in perihilar area with peribronchial thickening. High-resolution chest CT scan showed ground glass opacities, small nodules as well as small and large fibrotic bands indicating subacute and active interstitial lung disease. Work-up for tuberculosis including a skin test and gastric washing was negative. Mucociliary clearance and sweat chloride test were normal. Bronchoscopy with broncho-alveolar lavage (BAL) revealed a high proportion of eosinophils (>55% of total cell count). Pulmonary function test using body plethysmography and flow volume curves showed a decrease in vital capacity, FEV₁ and TLC, and an increase in residual volume, hence, diagnosis of a restrictive lung disease with mild obstructive pattern was made.(Table 1).

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Table 1: Pulmonary function test findings by body plethysmography

Measured Parameter	Unit	Normal values (for age)	Patient values	%
Total resistance	kPa s/L	0.36	0.54	150.1
Intrathoracic gas volume	L	1.48	1.18	79.3
Total lung capacity (TLC)	L	3.06	2.07	67.7
Vital capacity (VC)	L	2.28	1.00	43.6
Residual volume (RV)	L	0.75	1.07	142.8
Residual volume%	%TLC	25.47	51.89	203.7
Expiratory reserve volume	L	0.73	0.10	14.3
Vital capacity inspiratory	L	2.28	0.92	40.1
Forced vital capacity (FVC)	L	2.21	1.06	48.0
Forced vital capacity in 1 second (FEV ₁)	L	1.86	1.00	53.8
FEV ₁ %	%VC	84.95	109.18	128.5
Peak expiratory flow rate	L/s	4.25	1.94	45.6
Maximal expiratory flow rate in 75% of VC	L/s	3.83	1.74	45.5
Maximal expiratory flow rate in 50% of VC	L/s	2.70	1.58	58.3
Maximal expiratory flow rate in 25% of VC	L/s	1.38	0.60	43.4
Maximal mid-expiratory flow rate	L/s	2.35	1.26	53.6

Electrocardiography and echocardiography as well as plasma immunoglobulin (IgM, IgG, IgE, and IgA) concentrations, erythrocyte sedimentation rate, C-reactive protein, platelet and total white blood cell count (including eosinophils) were all within the normal range. There was no anemia and other studies including renal and liver function tests were unremarkable. Bronchoscopy and bronchial washing on several occasions revealed ciliate cells, some pigment-laden histiocytes, eosinophils and lymphocytes, and no tumor cells. A transbronchial biopsy revealed chronic inflammatory cell infiltrates within the interstitium and occasional aggregates of fibrin within the alveolar spaces. Scattered neutrophils and eosinophils were also present in alveolar spaces. There was no granuloma formation, vasculitis or any evidence of malignancy. Staining methods specific for *Mycobacteria spp.* as well as Grocott and Gram staining for other organisms were negative. These findings were confirmed by surgical lung biopsy performed in another center. Therefore, the diagnosis of CEP was considered and further investigation was performed to find the underlying cause. Skin prick test was negative for various antigens including dermatophytes, epithelial cells of cat, dog, horse, *Aspergillus spp.*, cladosporium, peanuts and tomato. Rast test for aspergillus was negative. Antigen-specific IgE antibodies revealed a slight elevation for damp water, budgie and canary serum. Serologic studies were negative for leishmania, schistosoma, fasciola, filaria, trichina, toxocaris and echinococcus. Stool exams were negative for protozoa and worms.

Based on these findings, the diagnosis of idiopathic CEP was made for this patient and treatment with oral prednisone was started. Because of a very low vital capacity he had in his pulmonary function test (*i.e.*, 0.45 (19.9%)) high-dose pulse

therapy with methylprednisone was started. He tolerated the treatment very well. Flow volume curves taken three days after treatment showed improved lung function. He had been receiving beclomethasone inhalation and his condition became stable after one year of follow-up.

Discussion

Chronic eosinophilic pneumonia frequently occurs in women in their fifth decade of life rather than in male children. Previous, and often prolonged history of asthma is common.¹⁻³ However, in some reports there has been only a history of atopia without asthmatic attacks.^{4,5} This was true for our case whose clinical picture and pulmonary function tests were not compatible with asthma but there was a history and clinical picture of allergic rhinitis. Altered general status, fever, especially prolonged pyrexia, persistent productive cough, wheezing and night sweating are the cardinal signs and symptoms in typical cases of CEP.¹ Nevertheless, these were not observed in our case. Indeed presentation of CEP with only post-exertional cough, clubbing of fingers and mild cyanosis in a child is the most remarkable feature of this case presentation. Also the absence of peripheral eosinophilia was strange since it is seen in most cases, though its absence does not rule out the diagnosis.^{1,2} Although the so-called negative image of pulmonary edema on chest x-ray is considered diagnostic for CEP, it is noted in only one third of patients.¹⁻⁴ Other features including patchy infiltration in both upper lung fields, pleural effusion, bilateral hilar and mediastinal lymphadenopathy, opacities distributed mainly in the subpleural regions¹⁻⁴, and even a normal chest x-ray has been reported previously.⁶

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Pulmonary function test and particularly BAL and pathologic findings in this case are compatible with typical CEP.^{1,3,4} Moreover, his dramatic response to corticosteroid therapy is another reason for correctness of our diagnosis.

This report emphasizes that CEP should be considered in pediatric age group as a cause for chronic hypoxemia or intractable pulmonary symptom. Although corticosteroid therapy has a dramatic effect on idiopathic CEP, a wide list of differential diagnosis should be considered in each eosinophilic lung disease before labeling patient as idiopathic.

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