Case Report

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TANAFFOS

Chronic Eosinophilic Pneumonia: a Case Report

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Chronic eosinophilic pneumonia (CEP) is a rare idiopathic interstitial lung disease, predominantly observed in females. Eosinophilia is present in most cases, and alveolar eosinophilia is a diagnostic criterion in more than 40% of bronchoalveolar lavage (BAL) samples.

The current study reported a 27-year-old male patient, non-smoker, with a history of uncontrolled asthma, presented to the emergency room with a complaint of cough, fever, and moderate dyspnea. A 30% eosinophilia was reported in his peripheral blood sample. A chest-X ray examination showed an upper and middle lobe consolidation, especially in the left lung. Broad-spectrum antibiotics were then started with a presumptive diagnosis of pneumonia, but no improvements were evident. The chest computed tomography scan showed air space opacities with septal thickening and predominant involvement of upper and middle lobes. Flexible bronchoscopy was performed, and the BAL sample analysis showed eosinophil infiltration, while negative culture. No parasites were identified. Transbronchial biopsies demonstrated eosinophil accumulation in alveoli and interstitium

Conclusion: Early recognition, diagnosis, and prompt treatment with corticosteroids are the main therapeutic approaches to CEP.

Key words: Chronic Eosinophilic Pneumonia, Eosinophilia, Pneumonia, Interstitial Lung Disease

INTRODUCTION

Chronic eosinophilic pneumonia (CEP) is a rare disorder, accounting for approximately 2.5% of interstitial lung diseases. It is idiopathic and can occur in any age group but is rarely observed in childhood. Up to half of CEP cases have a history of asthma preceding the disease (1,2). Clinical manifestations are nonspecific with subacute to chronic respiratory symptoms as common presentations (3). The presence of peripheral blood eosinophilia and radiographic findings in a patient with pneumonia, failing to resolve with antibiotic therapy raise the suspicion of CEP and other pulmonary infiltrates with eosinophilia-associated syndromes, such as CEP, allergic bronchopulmonary aspergillosis, fungal and parasitic infections, eosinophilic granulomatosis with polyangiitis, and hypereosinophilic syndrome (1-10). CEP has a distinctive radiographic feature, which includes peripheral parenchymal opacities involving the upper lobes (1-3).

CASE SUMMARIES

The present study patient was a 27-year-old male, non-smoker, and cell phone seller, presented to the emergency ward with a complaint of uncontrolled asthma, cough, fever, night sweat, weight loss, and moderate dyspnea started two months before attending the center. He had asthma since six years ago, generally not well-controlled.

He denied any systemic diseases and had never undergone any surgical procedures.

His family history was unremarkable, and the social history was negative for smoking and alcohol consumption. The patient received treatment with fluticasone, salmeterol, and a corticosteroid nasal spray. His vital signs were as follows: blood pressure 110/80 mmHg, respiratory rate 30 per minute, Pulse Rate 110, temperature 38°C, and O2 saturation in ambient air 92%.

He was pale and ill without dyspnea and chest retraction. On auscultation, wheeze and crackles were heard. No clubbing and lymphadenopathy were detected.

The patient was admitted for more evaluations. His white blood cell count was 11.500 cell/µL with 30% eosinophilia in the peripheral blood smear. Moreover, the erythrocyte sedimentation rate (ESR) was 81 mm/hour, and serum C-reactive protein level 12 mg/L. Spirometry revealed mild obstructive pattern with response to bronchodilator (Table 1).

Table 1. Spirometry test results of the patient at first visit

	Pre	%Pre	Post	%Post	%Change
FVC(L)	3.81	88	4.13	96	+8
FEV1(L)	2.37	74	3.25	88	+19
FEV1/FVC(%)	71.7	87	78.7	96	+10
FEF25/75(L/S)	1.99	43	3.39	73	+70

Detailed laboratory test results are illustrated in Table 2. Chest-X ray showed upper and middle lobe consolidations, especially in the left lung. Broad-spectrum antibiotics were therefore started with a presumptive diagnosis of pneumonia. However, no improvement was observed within 48 hours. Chest computed tomography (CT) scan revealed air space opacities with septal thickening and predominant involvement of upper and middle lobes (Figure 1).

The results of echocardiography and electrocardiogram were normal, and did not show any relevant pathology consistent with the patient's symptoms. The abdominal and pelvic ultrasonography result was normal. Endoscopy was done to evaluate eosinophilic esophagitis (a chronic allergic/immune condition of the esophagus in which a large number of eosinophils are found in the inner lining of the esophagus), which the result was normal. Bone scintigraphy and bone marrow aspiration were performed to rule out malignancy; both were negative in terms of malignancy and the FIP1L1-PDGFR gene (FIP1/platelet-derived growth factor). The results of electromyography and nerve conduction velocity were also normal. The results of other laboratory tests, including purified protein derivative, angiotensin-converting enzyme, antinuclear antibody, anti-ds-DNA, peripheral antineutrophil cytoplasmic antibodies, galactomannan, and HIV were negative. Serum protein electrophoresis had a normal pattern. Serum levels of immunoglobulin and different CD markers were also normal.

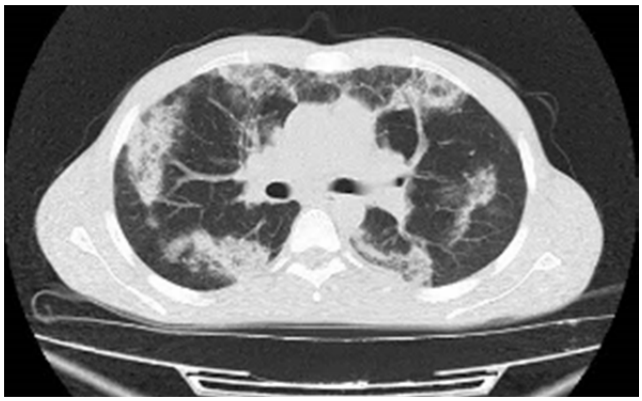

Figure 1. CT scan of the chest demonstrates areas with ground glass opacities

Considering infiltrative changes in chest CT scan, flexible bronchoscopy and lung biopsy were performed. Bronchoalveolar lavage (BAL) sample analysis showed eosinophil infiltration, while negative culture. No parasites were identified. No atypical cell was reported. Transbronchial biopsies demonstrated dense eosinophil accumulation in alveoli and interstitium, consistent with the diagnosis of eosinophilic pneumonia.

Ten days after hospitalization, the diagnosis of CEP was made based on peripheral eosinophilia, a high percentage of eosinophils in BAL specimen, and diagnostic findings on histopathologic examination, as well as ruling out of other causes of eosinophilia. Thereafter, a high dose of oral corticosteroid (50 mg prednisone daily) was started, and antibiotics were discontinued. Rapid and dramatic radiographic and clinical response to treatment was observed, and the patient was discharged on day five after undergoing corticosteroid therapy (Figure 2 a, b).

Table 2. Detailed laboratory data of the patient at admission day

WBC (×10³/mm³)	11500	ESR (mm/hr)	81	Troponin (ng/L)	12.3 (NL)
PMN (%)	49%	CRP (mg/L)	12	ANA	0.67 (NL)
Lymphocyte (%)	17%	BUN (mg/dl)	13	Anti-dsDNA(U/L)	1 (NL)
Monocyte (%)	4%	Cr (mg/dl)	1.2	MPO Ab (U/L)	1 (NL)
Eosinophil (%)	30%	AST (IU/L)	25	ALT (IU/L)	19
Hb (gr/dl)	12.5	Plt (×10³/mm³)	620 ×10³	LDH (u/l)	502

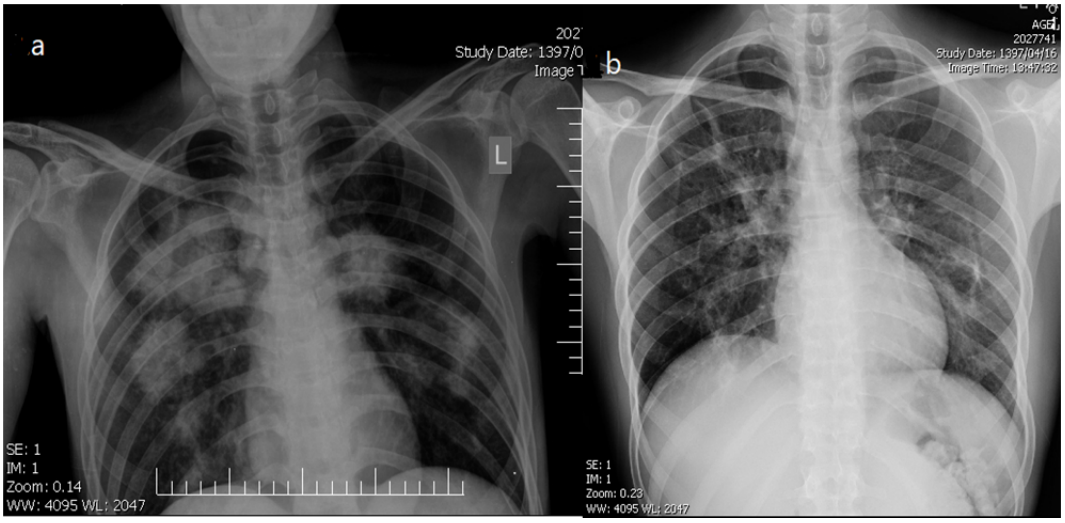

Figure 2. Chest radiographs of the patient before starting corticosteroid therapy (a) and 72 hours after starting medication (b) revealing dramatic regression of interstitial opacities in response to the treatment.

DISCUSSION

Eosinophilic lung diseases are characterized by the accumulation of eosinophils in the pulmonary interstitium and airspace. There are many different reasons for the disease (1,2) listed in Table 3. CEP was first described by Carrington CB in 1969 (3). The eosinophilic types of pneumonia are a heterogeneous group of diseases characterized by the accumulation of eosinophils in lung tissue based on the BAL samples evaluation. In addition, CEP is a rare cause of eosinophilic pneumonia with an estimated incidence of 0.23 cases per 100,000 general population. It has a 2:1 female/male ratio with an unknown etiologic cause. Most patients are middle-aged, and about half of them have a history of atopic diseases, particularly asthma (4).

The disease usually presents with nonspecific symptoms, including productive cough, fever, dyspnea, weight loss, and nocturnal sweat. On physical examination, wheezing or crackles might be detected, but most patients have mild abnormalities on auscultation. The vast majority of CEP cases have either obstructive or restrictive impairment in spirometry tests. It is a reliable diagnostic tool to document the severity of respiratory impairment and evaluate the response to the treatment. Chest imaging usually demonstrates pathologic changes that are not diagnostic. Chest CT classically reveals bilateral upper and peripheral opacities only present in 28% of the cases.

The diagnosis is typically made based on clinical presentations after ruling out other causes of the eosinophilic types of pneumonia. A history of asthma and

atopic diseases, in addition to insidious onset of symptoms, can guide toward making the accurate diagnosis (4,7,8). Most patients show eosinophilia in the complete blood cell count test and examination of peripheral blood smear. ESR and IgE levels elevate in most patients. The increased number of eosinophils and lymphocytes are observed in sputum and BAL specimens (9,10). Histopathologic examination of the specimens shows moderate to extensive consolidation of alveoli, flooded with a proteinaceous exudate, and predominately eosinophilic infiltration into the alveoli and interstitium. A dramatic response to corticosteroid therapy with rapid resolution of radiographic involvement of the chest further confirms the diagnosis (11). The mainstay of the treatment is corticosteroids and most patients require a relatively prolonged therapy for about six months with transient tapering of medication to reduce relapses (10,12).

It is noteworthy that the diagnosis of CEP is made after excluding other eosinophilic lung diseases, such as parasitic and fungal infections, medication-induced eosinophilic pneumonia, malignancy (both solid and hematologic malignancies), hypereosinophilic syndrome, and other causes mentioned in Table 3. In cases with no response to treatment, bone marrow biopsy and FIP1L1-PDGFR test should be taken to evaluate bone marrow diseases, such as myeloproliferative neoplasms and hypereosinophilic syndromes (2). In the present case report, the patient had a weight gain, and his signs and symptoms were resolved rapidly after corticosteroid therapy. During tapering corticosteroid therapy, the patient experienced an episode of disease relapse. Corticosteroid was discontinued more slowly within nine months. Currently, he is in remission and does not have any symptoms.

Table 3. Causes of eosinophilic lung diseases

Helminthic infections	Interstitial lung diseases	Drug-induced eosinophilic lung disease
Ascaris lumbricoides	Cryptogenic organizing pneumonia	Nitrofurantoin
Hookworm	Hypersensitivity pneumonitis	Minocycline/tetracyclines
Strongyloides	Idiopathic pulmonary fibrosis	Sulfonamides
Paragonimiasis	Langerhans cell histiocytosis	Ampicillin
Trichinellosis	Sarcoidosis	Daptomycin
Cutaneous and visceral larva		Phenytoin
Schistosomiasis		Cutaneous and
Wuchereria bancrofti		L-Tryptophan
Brugia malayi		Methotrexate
Non-helminthic infections	Idiopathic	NSAIDs
Coccidiomycosis	Acute eosinophilic pneumonia	Anti-malarials
Aspergillosis	Chronic eosinophilic pneumonia	(dapsone, pyrimethamine)
	Idiopathic hypereosinophilic syndrome	Amiodarone
		ACE inhibitors
		H2-receptor antagonists
Malignancy	Associated with systemic vasculitis	Allergen-related eosinophilic lung disease
Leukemia	Eosinophilic granulomatosis with polyangiitis (EGPA)	Allergic bronchopulmonary aspergillosis (ABPA)
lymphoma	Granulomatosis with polyangiitis	
Primary lung cancer		

CONCLUSION

It was a rare case of CEP presenting with prominent B symptoms (fever, night sweat, and weight loss). Similar to other cases of CEP, early recognition, diagnosis, and prompt treatment with corticosteroids are the main therapeutic approaches. In other words, a strong clinical suspicion, a thorough history, accurate diagnosis, and timely treatment with corticosteroids are the cornerstone of therapy.

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