CRYPTOGENIC PULMONARY EOSINOPHILIA - A CASE REPORT

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ABSTRACT

A 44-year-old Indian housewife presented with prolonged cough, intermittent fever, loss of weight and shortness of breath on exertion. Physical examination revealed a thin lady with fine crackles on both lungs. Marked eosinophilia and elevated erythrocyte sedimentation rate were noted on peripheral blood. Chest radiograph showed characteristic bilateral infiltrates affecting the periphery of both lungs. Treatment with corticosteroid resulted in dramatic improvement in symptoms, signs and radiographic changes within a few days.

Keywords: Cryptogenic pulmonary eosinophilia, corticosteroid

SINGAPORE MED J 1990; Vol 31: 629-631

INTRODUCTION

Since the description of a syndrome called pulmonary eosinophilia by Loffler in 1932, several different entities have been described. Fungi especially Aspergillus fumigatus, parasites and various drugs have been identified as causing the syndrome (1).

The diagnosis of cryptogenic pulmonary eosinophilia is often made after excluding these various causative agents. However, certain features are recognised and peculiar to the condition. These are illustrated in this case report and in the discussion that follows.

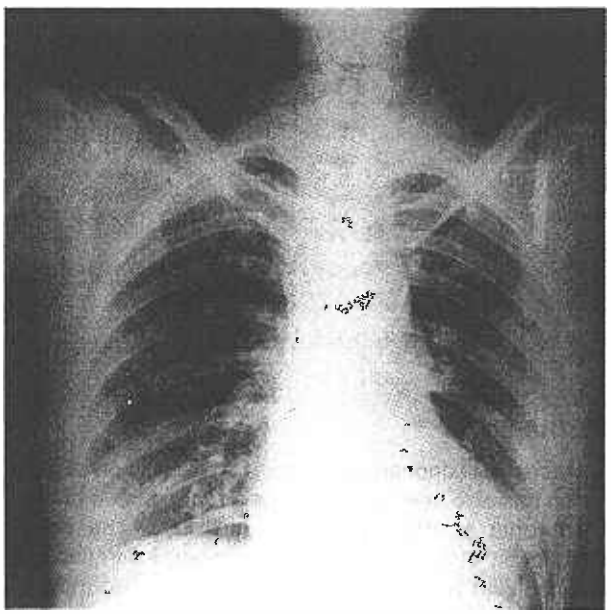
CASE REPORT

A 44-year-old Indian housewife presented to General Hospital, Kuala Lumpur in late 1989 with a 3-year history of recurrent cough, intermittent fever, night sweats and weight loss. She also had progressive shortness of breath on exertion. Prior to this she had been in good health. There was no history of allergy, asthma or pulmonary tuberculosis in herself or her family. She had not been on any prolonged drug treatment and did not keep birds at home.

On examination, she was febrile with a temperature of 38°C but appeared quite comfortable. She was thin and pale but not cyanosed or clubbed. Chest expansion was bilaterally diminished and fine late inspiratory crackles were heard over mid and lower zones of both lungs. Abdominal and cardio-vascular examinations were unremarkable. Investigations showed haemoglobin of 7.9 gm%, total white count of 8.7 x 10^9/l with eosinophilia of 1700/c.mm (19%). Erythrocyte sedimentation rate was 140 mm per hour. Chest radiograph showed bilateral pulmonary infiltrates in the periphery of both mid and lower zones (Fig 1). A provisional diagnosis of pulmonary tuberculosis was made. Sputum smear for acid fast bacilli were repeatedly negative, and Mantoux test was non-reactive. Serum creatinine, electrolytes, liver enzymes and urine analysis were normal. She was referred to the Respiratory Unit for opinion and further management.

Fig 1

Chest radiograph of the patient taken a few days after admission showing homogenous opacity on right lower zone and left mid zone located peripherally.



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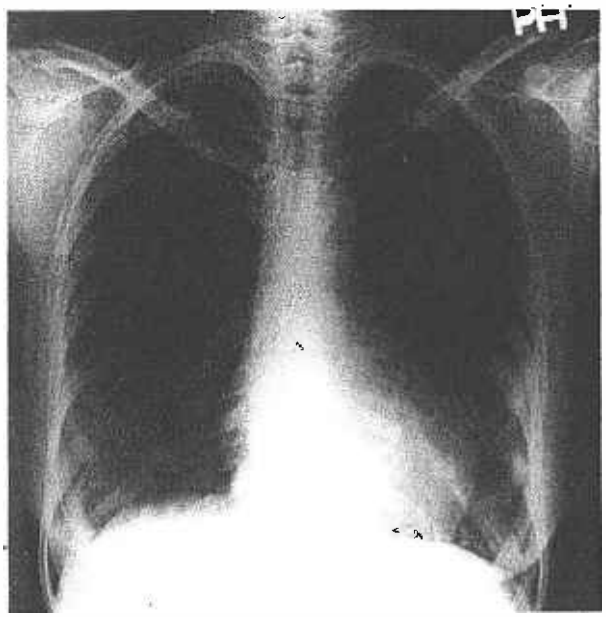
Further investigations were done including stools for ova and parasites, skin sensitivity test for Aspergillus fumigatus, collagen screening and filarial antibody test which were all negative. Lung function test showed forced expiratory volume in 1 second (FEV_1) and forced vital capacity (FVC) of 1.6 and 2.3 litres respectively. These were both reduced to 61% and 72% of the predicted values. Carbon monoxide transfer factor was also decreased (4.86 ml CO/min/mmHg). A bronchoscopy was performed which showed normal bronchi. A diagnosis of cryptogenic pulmonary eosinophilia was made and prednisolone 20 mg daily was started. Within 24 hours the temperature returned to normal and her dyspnoea improved. Lung crackles were not audible anymore. Repeat spirometry 3 days after the commencement of corticosteroid showed a rise in FEV_1 and FVC to 2.3 and 2.65 litres respectively. Carbon monoxide transfer factor done a week later showed a normal value (29.03 ml CO/min/mmHg).

Chest radiograph done 10 days later showed remarkable resolution of the pulmonary infiltrates (Fig 2). Meanwhile, the result of transbronchial biopsy was available. It showed scattered inflammatory cells including eosinophils in the bronchial wall. However, no lung tissue was included in the biopsy. The corticosteriod was continued on discharge. She was reviewed a month later at our outpatient clinic. She remained well and asymptomatic and had gained 3 kg of weight. The corticosteroid was continued at the same dose. A repeat chest radiograph showed clear lung fields.

Fig 2

Repeat chest radiograph after 10 days of corticosteroid treatment showing resolution of pulmonary infiltrates.



DISCUSSION

Cryptogenic pulmonary eosinophilia is a respiratory disease which has also been called as eosinophilic pneumonia (2) or chronic eosinophilic pneumonia (3). The earlier description of PIE syndrome (pulmonary infiltration with eosinophilia) by Reeder and Goodrich (4) also includes patients with features similar to cryptogenic pulmonary eosinophilia. Most reported cases have a few features in common: 1) typical radiographic changes, 2) a remarkable and rapid response to corticosteroid treatment and 3) a characteristic histology of lung biopsy.

The typical chest radiographic features of dense opacities with ill defined margins and without lobar or segmental distribution arranged peripherally is virtually diagnostic. Gaensler and Carrington (5) reviewed 29 cases of cryptogenic pulmonary eosinophilia and found 24 with typical radiographic changes. The remaining 5 cases had radiographic changes not typical of crytogenic pulmonary eosinophilia infiltration of the interstitium and air space. The radiographic feature is sometimes described as photographic negative or reverse butterfly pattern of pulmonary oedema.

Response to corticosteroid is usually so dramatic that most patients improve within 24 hours of therapy (2 -6). This was well illustrated in our case where the temperature returned to normal and the dyspnoea improved. Lung function parameters also improved remarkably within 72 hours and definite resolution of pulmonary shadowing was observed after 10 days. Apart from bronchial asthma and allergic bronchopulmonary aspergillosis which may respond fairly rapidly to corticosteroid, it is unusual for other conditions to respond in this manner. Prednisolone of 20 mg daily may be sufficient (6) and should be maintained for several months to prevent relapses.

Although peripheral blood eosinophilia with typical radiographic features are strongly diagnostic, the exclusion of other causes of pulmonary eosinophilia is often necessary. Worm infestation and filariasis are two conditions which are endemic in our country and warrant investigations for the exclusion. Pulmonary eosinophilia secondary to worm infestation is often unnoticed or transient and unlikely to continue for months. The absence of ova and parasite in the stool excludes the diagnosis. Tropical pulmonary eosinophilia due to microfilaria may occasionally cause prolonged symptoms, but dramatic response to corticosteroid is unlikely. Positive serology and good response to diethylcarbamazine are diagnostic criteria (7).

The initial diagnosis of pulmonary tuberculosis as in this case is not unexpected in a person who presents with chronic cough with fever, sweats and marked loss of weight. In fact in many reported series, a significant proportion of patients was initially diagnosed and treated with antituberculous drugs but deteriorated while on treatment (2,4). Pulmonary tuberculosis is certainly an important differential diagnosis to be considered especially in an endemic country. In about half of the cases of cryptogenic pulmonary eosinophilia, bronchial asthma may co-exist (3,6). Radiological changes, eosinophilia and clinical course are similar between asthmatics and non asthmatics.

We feel the diagnosis of cryptogenic pulmonary eosinophilia can be reasonably made in a patient with chronic respiratory symptoms, typical chest radiographic features and peripheral blood eosinophilia. A trial of corticosteroid should be given after excluding other causes of pulmonary eosinophilia and chronic pulmonary infection like tuberculosis. Lung biopsy may be needed in a small proportion of patients with less typical presentation.

ACKNOWLEDGEMENT

We wish to thank the Department of Medical Education, Universiti Kebangsaan Malaysia for preparing the photographs and Mrs Khairun Nor Zabaai for typing the manuscript with great care.

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