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A case of chronic eosinophilic pneumonia

[1]P Gallipoli, [2]M Leach
[1]*Senior House Officer, Haematology, North Glasgow University Hospitals, Glasgow, Scotland,* [2]*Consultant Haematologist, Stobhill Hospital, Glasgow, Scotland*

ABSTRACT We describe the case of a 58-year-old man who presented with a four-week history of general malaise, sweats, weight loss and dyspnoea together with pulmonary infiltrates on CXR and a peripheral blood eosinophilia. A clinical diagnosis of CEP was made. He was treated with oral steroids with rapid clinical improvement together with resolution of X-ray changes and blood eosinophilia. The clinical presentation, differential diagnosis, radiology, treatment and prognosis of CEP are discussed.

KEYWORDS Eosinophilia, pneumonia

LIST OF ABBREVIATIONS Accident and emergency department (A&E), acute eosinophilic pneumonia (AEP), anti-neutrophil cytoplasmic antibody (ANCA), anti-nuclear antibody (ANA), bronchoalveolar lavage (BAL), chest X-ray (CXR), chronic eosinophilic pneumonia (CEP), computed tomography (CT), full blood count (FBC), white blood cell count (WBC),

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Correspondence to ***P Gallipoli, Department of Haematology, 3rd floor McEwan Building, Glasgow Royal Infirmary, Castle Street, Glasgow, G4 0SF***

tel. ***+44 (0)141 2114 513 / 2114 001***

fax. ***+44 (0)141 2114 931***

e-mail
paolo.gallipoli@northglasgow.scot.nhs.uk

CASE REPORT

A 58-year-old man attended our A&E department with a four-week history of lethargy, night sweats and weight loss. He also reported increasing shortness of breath during the week prior to his presentation. He had a history of mild bronchial asthma and osteoarthritis but used no regular medications. On examination he appeared pale and unwell with a low grade fever. He had bilateral upper lobe crepitations on chest auscultation but the examination was otherwise unremarkable. His FBC showed Hb 96 g/L, WBC 30x10^9/L, platelets 1,202x10^9/L. The eosinophil count was markedly elevated at 17x10^9/L. His CXR showed infiltrates, bilaterally in the upper lobes (see Figure 1) each of which appeared to abut the pleura on CT imaging (see Figure 2). An obvious pleural effusion was also present. At bronchoscopy the bronchial tree appeared normal. Specimens from a BAL failed to grow any organisms. Cytology showed prominent eosinophils, reactive respiratory epithelial cells and necrotic debris. Transbronchial biopsies of the identified parenchymal lesions showed an inflammatory exudate with marked eosinophilic infiltration in the alveoli and interstitium (see Figure 3). Anti-neutrophil cytoplasmic antibody, rheumatoid factor and ANA serology were all negative. Screening of stools and bronchial lavage fluid for parasites were both negative. Strongyloides serology was negative.

A diagnosis of CEP was made on the basis of his constellation of symptoms, peripheral blood eosinophilia, radiological findings and lung histology. He

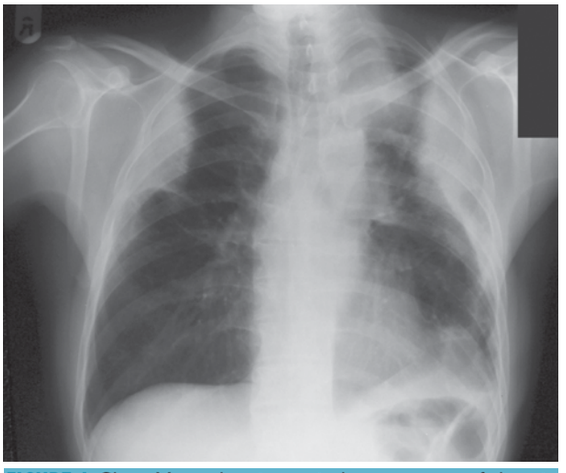

FIGURE 1 Chest X-ray showing typical appearances of chronic eosinophilic pneumonia with bilateral peripheral upper lobe infiltrates.

was commenced on prednisolone 50 mg daily. He responded rapidly with significant improvement of symptoms, fever and eosinophilia within the first 24 hours. The patient's eosinophil count, CXR appearances and clinical symptoms were used to guide early decisions regarding steroid dose reduction. After four weeks of treatment, all symptoms had resolved, along with normalisation of the CXR and FBC. After six weeks on 50 mg daily, the prednisolone dose was reduced to 25 mg daily. Thereafter an empirical stepwise reduction of 5 mg every six weeks was used until steroids were stopped after approximately six months of treatment. He is currently well and off steroids with no evidence of relapse.

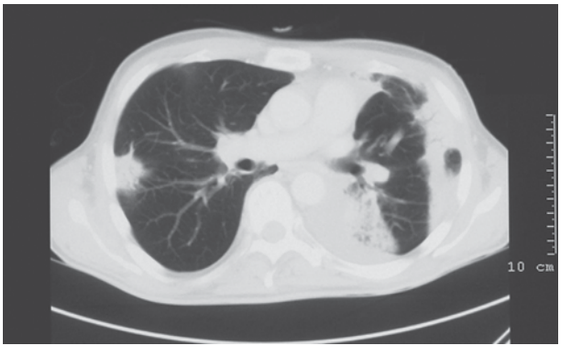

FIGURE 2 Computed tomography (lung windows) showing typical bilateral infiltrates directly abutting the pleura.

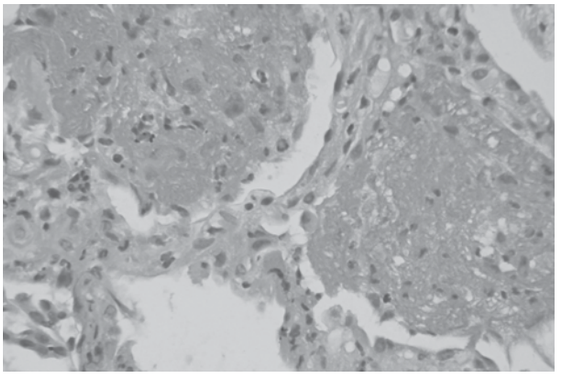

FIGURE 3 Transbronchial biopsy showing intra-alveolar inflammatory exudates with marked eosinophilic infiltration.

DISCUSSION

Eosinophilic lung diseases are a heterogeneous group of clinical entities characterised by an accumulation of eosinophils in airways or lung parenchyma with or without peripheral blood eosinophilia. A diagnosis of eosinophilic lung disease can be made when respiratory and systemic symptoms and pulmonary opacities on X-ray are present together with either peripheral blood eosinophilia or tissue eosinophilia confirmed in BAL or at lung biopsy. They can be associated with other known disease entities (such as interstitial lung disease, drug reactions, pulmonary vasculitis, parasitic disease and malignancy) or be idiopathic. According to the pathogenetic mechanism, eosinophils can play different roles in these diseases as they can cause direct tissue damage, be just part of the inflammatory process, or may even have a protective role.[1–5]

Chronic eosinophilic pneumonia is one of the idiopathic forms of eosinophilic lung disease together with simple pulmonary eosinophilia (Loeffler syndrome) and acute eosinophilic pneumonia. It is not known to be triggered by reactions to drugs or infection and its cause is unknown. Eosinophil accumulation in the lungs in CEP seems to be due to selective migration of Th2 cells to the lungs in response to an unknown trigger. Th2 cells release increased amounts of IL-5 and other eosinophil active cytokines resulting in eosinophil accumulation in the alveolar space and interstitium.[2]

Chronic eosinophilic pneumonia tends to present with a several week history of marked systemic upset (night sweats, weight loss, general malaise, fever). Chest symptoms like cough and dyspnoea are frequent but can be mild. These presenting features allow to differentiate CEP from AEP as the latter tends to present with an acute illness of usually less than five days' duration and significant respiratory impairment that can be so severe to require ventilatory support. Chronic eosinophilic pneumonia is more frequent in middle-aged women but can affect both sexes and all ages. It may be associated with a previous history of asthma or atopy but can affect previously fit individuals. Extrapulmonary involvement is rare and should prompt a consideration of alternative diagnoses.[6–7] The diagnosis, however, is one of exclusion. Chest X-ray appearances are characteristic with bilateral pleural based, wedge shaped infiltrates that can be migratory. Computed tomography will show typical nonsegmental areas of airspace consolidation with peripheral predominance usually in the upper lobes with ground glass texture. The absence of bronchiectasis on CT helps to differentiate CEP from allergic bronchopulmonary aspergillosis, another common cause of eosinophilic lung diasease. Pleural effusion is observed in less than 10% of cases.[4] Inflammatory markers are normally elevated and, except for increased serum IgE levels, other serological tests are usually negative. Abnormal pulmonary function tests, both restrictive and obstructive defects, have been described in association with CEP although they are not always present.[6–7] Serum IgE levels and pulmonary function tests were not performed in the case described. In the presence of a suggestive clinical picture and once alternative causes have been excluded, CT appearances in conjunction with alveolar eosinophilia or peripheral blood eosinophilia is enough to establish diagnosis in most cases without resorting to lung biopsy.[4]

Spontaneous resolution has been described in less than 10% of patients, but CEP tends to respond dramatically to corticosteroid therapy (0·5 to 1 mg/kg daily). However, this condition has a high tendency to relapse when steroids are withdrawn. This means that corticosteroid therapy will often need to be tapered over several months, and in some instances it is not possible to wean patients off the steroids completely.[7]

CONCLUSION

Chronic eosinophilic pneumonia appears to be a distinct clinical entity with a currently unknown aetiology. The

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condition should be considered in any patient presenting with a slow-onset history of dyspnoea, night sweats, lethargy or weight loss in association with pleural based pulmonary infiltrates and a peripheral blood eosinophilia. Patients respond promptly to oral corticosteroid therapy and the prognosis is good, though relapses do occur if steroids are withdrawn too quickly.

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