A Case to Remember.....

Pneumonia is not always what it seems

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Teaching Points
1. Consider chronic eosinophilic pneumonia (CEP) when a patient with apparent pneumonia fails to respond to standard therapy.
2. Peripheral blood eosinophil count may be normal, and radiographic features are often non-specific.
3. Diagnosis of CEP can be made by lung biopsy, with demonstration of large numbers of eosinophils in the alveoli.
4. Corticosteroids usually produce resolution of symptoms, although prolonged treatment may be necessary.

Keywords
Chronic eosinophilic pneumonia, eosinophilia.

Case History

A 33 year old, previously healthy woman was referred to hospital with a 3 month history of cough, 6kg weight loss, dyspnoea and night sweats. She described intermittent expectoration of purulent sputum without haemoptysis. There was no history of recent travel outside UK or contact with tuberculosis. Her general practitioner had treated her with course of amoxycillin followed by ciprofloxacin with no relief.

On examination she had a temperature of 37.4C with a sinus tachycardia of 118 /min. The heart sounds were normal with a blood pressure of 130/80 mmHg. Her lung fields were clear on auscultation.

Initial investigations were as follows: Haemoglobin (Hb). 11.2g/dl, White cell count (WCC) 11.1g/dl, Platelets 949 x 10^9/l, Erythrocyte sedimentation rate (ESR) 135mm/hour, C-reactive protein 179. Differential WCC including eosinophil count was normal. Apart from an alkaline phosphatase of 221 (normal: 40-100), the liver function was normal, as were renal function and arterial blood gas tensions. Chest x-ray showed consolidation of the left midzone with an air bronchogram (Figure 1).

She initially commenced anti-tuberculous treatment, which was discontinued after three sputum samples proved negative for acid fast bacilli, and changed to doxycycline, 200mg initially followed by 100mg daily. Bronchoscopy was normal, and bronchial washing from the left upper lobe failed to grow any pathogenic organisms. Repeat chest x-ray after two weeks showed improvement of the left sided consolidation. Full blood count (FBC) at this time was normal apart from a platelet count of 998 x 10^9/l and ESR remained elevated at 132mm/hr. She was discharged from hospital on doxycycline 100mg daily.

A chest x-ray undertaken two weeks later at outpatient follow-up showed a new focal patch of consolidation in the right mid-zone along with left upper- and mid-zone involvement (Figure 2). Spirometry at this time revealed FEV1 of 1.52 litres

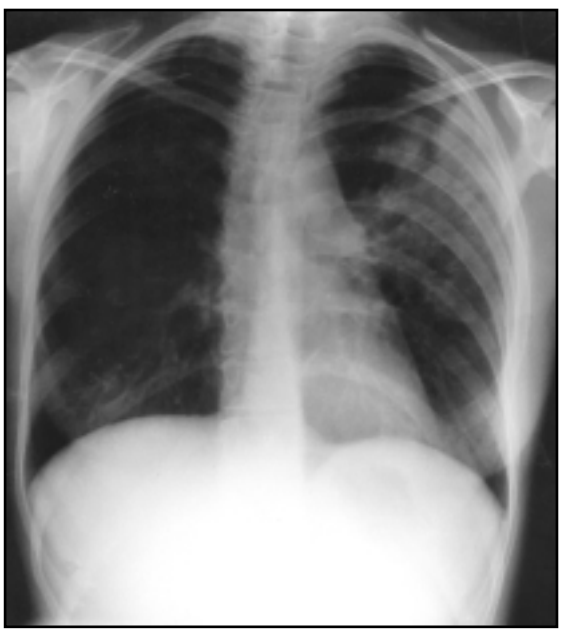

Figure 1. Chest x-ray on admission.

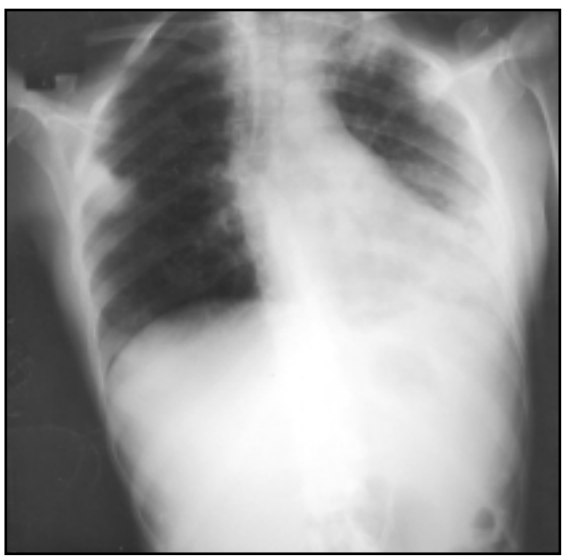

Figure 2. Chest x-ray after 4 weeks' treatment with doxycycline – shadowing is increased in the left mid and lower zones, and there is new peripheral shadowing in the right midzone.

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(44% predicted) and FVC of 1.80 litres (45% predicted), indicating a restrictive defect (FEV1 / FVC 84%). FBC was repeated, and indicated Hb 10.3g/dl, WCC 12.5g/dl, and platelets 1023 x 10^9/l. The differential white cell count revealed an eosinophilia of 14.2% (normal 0-8%).

Lung biopsy was undertaken. This showed a dense infiltrate of eosinophils filling the alveolar spaces (Figure 3), indicating a diagnosis of chronic eosinophilic pneumonia. She was started on oral prednisolone.

At outpatient follow up her chest x-ray had returned to normal (Figure 4) and her symptoms and spirometry had also improved, with FEV1 2.52 (74% predicted) and FVC 3.03 (77% predicted). Two years later she remains on a daily dose of 10 mg of prednisolone. An attempt to reduce the dose of prednisolone has resulted in worsening dyspnoea and a rise in her eosinophil count.

Discussion

Chronic Eosinophilic Pneumonia was first described by Christoforidis and Molnar in 1960.[1] Although reported in children, the peak incidence occurs in the fifth decade and females predominate 2:1.[2] The onset is insidious with an average symptom duration of over 7 months before diagnosis.[2] The most common symptoms are cough, fever, dyspnoea and weight loss.[2] Haemoptysis is rare. Peripheral blood eosinophilia occurs in most patients, but may be absent at presentation as in this case. The elevated ESR and thrombocytosis seen in this case are well recognised features. The classical radiographic finding of dense, extensive, bilateral peripheral infiltrates, often described as a "negative image of pulmonary oedema" is often considered diagnostic[3] but is seen only in 25% of the patients.[2] The appearance of patchy, peripheral infiltrates is a more common appearance, and may mimic the appearance of bronchopneumonia, leading to initial misdiagnosis. Pulmonary function tests can be normal in mild cases but may show restrictive defects with a reduced transfer factor.

Diagnosis is usually confirmed by histological examination of the lung, which demonstrates eosinophil accumulation in the alveoli. Interstitial fibrosis is noted in about 50% of cases.

Other causes of pulmonary abnormalities with peripheral eosinophilia include simple pulmonary eosinophilia, idiopathic hypereosinophilic syndrome, allergic bronchopulmonary asperginosis Churg-Strauss syndrome, parasitic infections and drug reactions.

Prednisolone in a dose of 30-40 mg/day will cause dramatic resolution of symptoms within 24-48 hours[4] and of chest x-ray abnormalities, within 10 days.[5] Relapse may occur if steroids are withdrawn in the first 6 months. Most patients will be able to discontinue corticosteroids without further relapse; however, a minority will require steroids indefinitely.

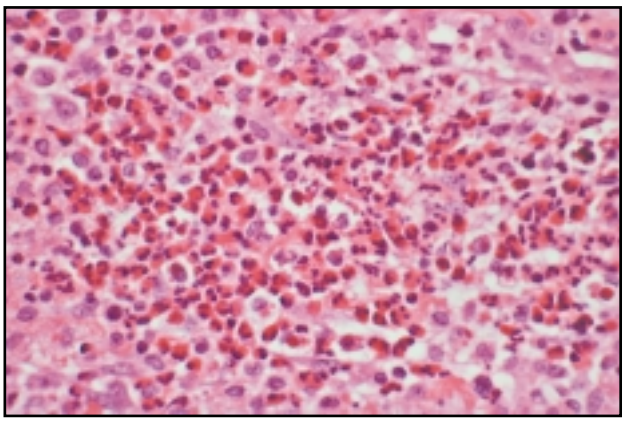

Figure 3. Lung biopsy showing extensive infiltration with eosinophils.

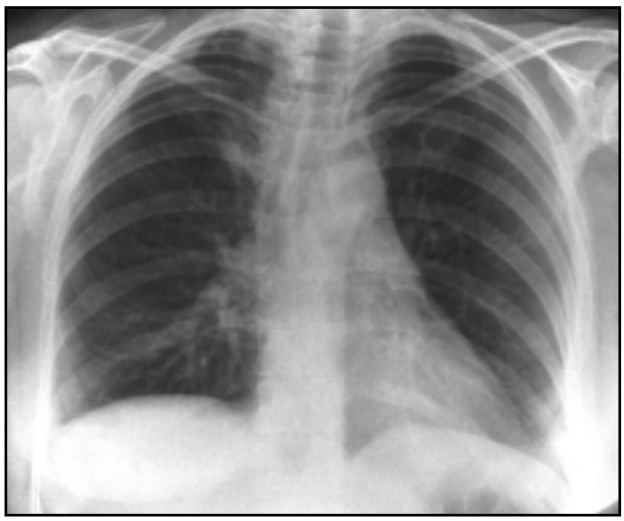

Figure 4. Chest x-ray after commencing treatment with corticosteroids.

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