RADIOLOGICAL CASE

CLINICS IN DIAGNOSTIC IMAGING (17)

J B K Khoo, K H Pay, W T Poh

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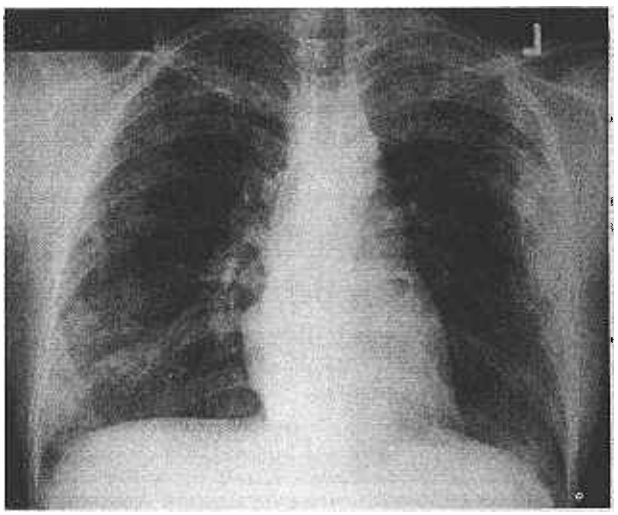
CASE REPORT

A 24-year-old Chinese woman presented with complaints of dry cough, vague chest pain and low grade fever of 1 month's duration. She had been treated by her family physician with a course of Amoxil without improvement. She had also complained of malaise and loss of weight of about 4kg over the past 4 months. No significant past medical or travel history was obtained. There was no family history of pulmonary tuberculosis.

Clinical examination revealed bilateral bronchial breath sounds on auscultation of her lungs. Heart sounds were normal. There was no rash, lymphadenopathy or hepatosplenomegaly. Vital signs were stable. She had intermittent low grade fever of between 37°C to 38°C. Positive blood tests were: raised erythrocyte sedimentation rate (ESR) of 77mm per first hour, haemoglobin 11.6 g/dL and total leucocyte count of $16X10^9$/L. The differential count consisted of polymorphs 47%, lymphocytes 30%, monocytes 3% and eosinophils 20%.

What does the chest radiograph (Fig 1) show? What is the diagnosis?

Fig 1 – Frontal chest radiograph.



Department of Diagnostic Radiology
Singapore General Hospital
Outram Road
Singapore 169608

J B K Khoo, MBBS, FRCR
Senior Registrar

K H Pay, MBBS
Medical Officer

Department of Pathology
Singapore General Hospital

W T Poh, MBBS, FRCPA
Consultant

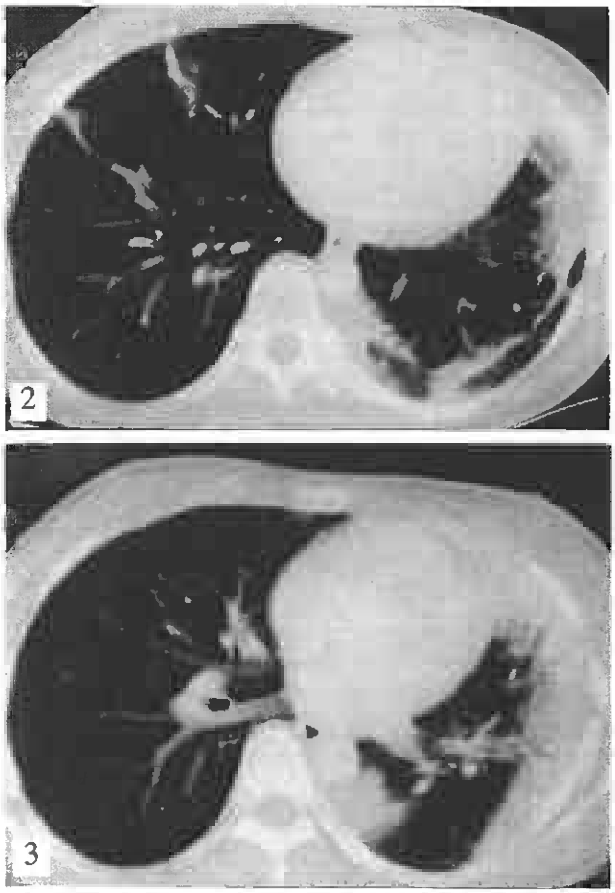
Correspondence to: Dr J B K Khoo

IMAGING INTERPRETATION

The chest radiograph (Fig 1) demonstrates ill-defined coalescent opacities (or air-space consolidation) in the periphery of both lung fields. The pattern of a clear area between the opacities and the central pulmonary vessels resembles a 'photographic negative of pulmonary oedema'. No hilar lymphadenopathy or pleural effusion is seen. The heart size is normal.

Computed tomography (CT) of her thorax confirmed the peripheral distribution of the air-space consolidation. (Figs 2 and 3).

Fig 2 and 3 – Bilateral patchy peripheral air-space consolidation on CT of the thorax. Air bronchogram effect is seen in Fig. 3.



DIAGNOSIS

Chronic eosinophilic pneumonia (CEP)

CLINICAL COURSE

The diagnosis of CEP was evident from the constellation of symptoms and the highly characteristic chest radiographic signs together with blood eosinophilia. This was supported by the findings of the transbronchial lung biopsy which showed infiltration of the alveolar spaces and interalveolar septa by histiocytes and eosinophils (Fig 4). Immunoperoxidase stain for S100 antigen was negative indicating that these were not Langerhan's histiocytes. Bronchial aspirate was negative for acid-fast bacilli and pneumocystis carinii.

The other eosinophilic lung diseases such as Loeffler's syndrome and tropical pulmonary eosinophilia were less likely in the absence of parasites and ova in the stool and filarial antibody, all of which were tested for in this patient.

The patient was treated with oral corticosteroids. Complete resolution of the consolidation was seen on the chest film after 2 weeks of treatment (Fig 5). She is presently maintained on low dose corticosteroids and is clinically well at the latest follow-up.

Fig 4 – Photomicrograph of the lung tissue showing patchy consolidation of the alveolar spaces by infiltrates of histiocytes (arrow head) and eosinophils (long arrow). H + E x 400.

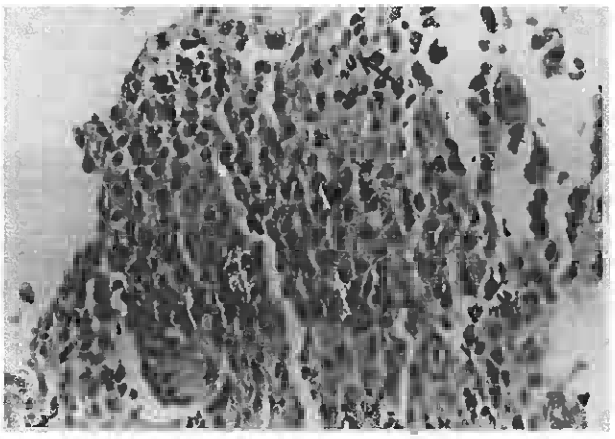
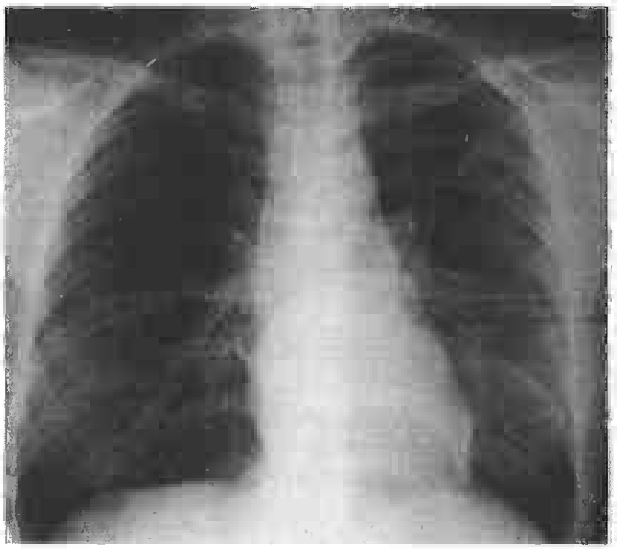


Fig 5 – Repeat chest radiograph shows complete resolution of the peripheral air-space opacities after 2 weeks of oral corticosteroid treatment. Residual linear scar is seen in the left upper lobe.



DISCUSSION

Air-space consolidation is seen on the frontal chest radiograph as ill-defined coalescent opacities. Presence of air bronchogram is an additional confirmatory sign of air-space consolidation. The cause of air-space consolidation depends on the material that fills the pulmonary acini, examples of which include water in pulmonary oedema, blood in pulmonary haemorrhage, inflammatory material in pneumonia, and tumour in alveolar cell carcinoma or lymphoma. Although air-space consolidation is a non-specific radiological sign, one can often arrive at the correct diagnosis by studying the distribution of the consolidation and associated signs together with a review of the clinical presentation. In alveolar pulmonary oedema, there is air-space consolidation in the perihilar distribution producing a 'batswing' pattern. To date, there has been no satisfactory explanation for this distribution.

When the air-space consolidation is in the lung periphery or is subpleural in location, hence sparing the perihilar region, the

term 'photographic negative of pulmonary oedema' pattern has been coined. This pattern is highly characteristic of CEP in the appropriate clinical setting in a patient with eosinophilia. The chest radiographic appearance by itself can resemble Loeffler's syndrome and bronchiolitis obliterans organising pneumonia (BOOP) (also known as cryptogenic organising pneumonia).

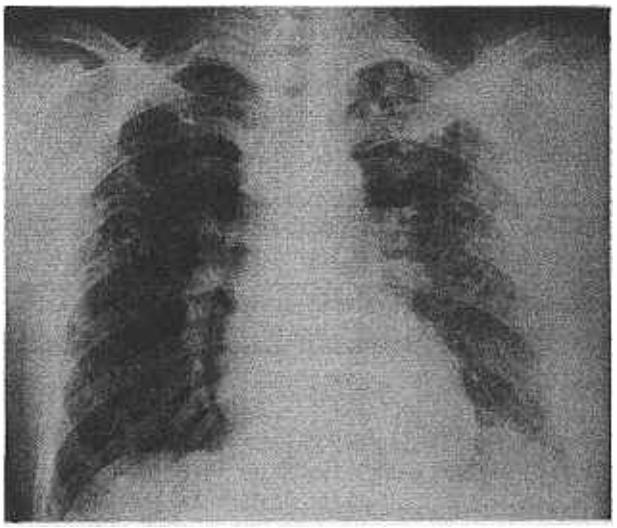
Patients with Loeffler's syndrome or simple pulmonary eosinophilia have mild blood eosinophilia and bilateral fleeting peripheral air-space opacities on the chest radiograph which does not usually last more than 2 weeks. It is a mild, self-limiting illness that usually resolves spontaneously within a month and is associated with ascaris worm infestation. The pulmonary opacities are believed to be the result of an immunological reaction in the lungs rather than actual migration of ascaris through the lung[1]. BOOP is the result of incomplete resolution of inflammation in the distal lung structures. It is most commonly seen as a chronic sequel to unresolved bacterial pneumonia but it has also been linked with collagen vascular disease, ulcerative colitis. radiation injury and drugs such as acebutolol and amiodarone[2]. Despite a raised ESR, the peripheral white blood count is normal and this distinguishes it from CEP.

More than 20 years ago, Carrington et al[3] coined the term 'chronic eosinophilic pneumonia' (CEP) to describe a condition in which there was coexistence of blood eosinophilia with pulmonary eosinophilic infiltration for which there was no obvious cause. The condition occurs most commonly in middle-aged women, but can occur at any age and in either sex. Cough either without or productive of mucoid sputum, breathlessness, weight loss, fever, and night sweats are the main initial symptoms. These symptoms may persist for weeks or months if adequate therapy is not given[1].

CEP classically produces non-segmental homogeneous consolidation in the periphery of the lungs, described as 'photographic negative of pulmonary oedema[4] pattern on plain chest radiographs. The peripheral air-space opacities are more elegantly demonstrated on CT scans. Therefore in instances where the distribution of the pulmonary opacities are not readily apparent on the plain chest radiograph, CT may be helpful in establishing the diagnosis[5].

Although an unusual condition, the diagnosis of CEP rarely presents a problem because of the highly typical combination of clinical symptoms, radiographic signs and blood eosinophilia. Occasionally where the diagnosis is doubtful, lung biopsy may be needed. The predominant histological feature of chronic eosinophilic pneumonia is filling of the alveolar spaces by eosinophils and histiocytes. Histologically, CEP can resemble cosinophilic granuloma (EG) but it differs from EG in that the infiltrates are in the alveolar spaces and not in the pulmonary interstitium in the case of the latter condition. In difficult cases, performing an immunoperoxidase stain for S100 antigen should answer the question, since it is present in the histiocytes of EG but not the macrophages of eosinophilic pneumonia[6]. An easier

Fig 6 – Another patient with diagnosis of eosinophilic granuloma (EG). Diffuse reticulonodular shadowing is present involving both lungs.



alternative is to review the chest radiograph. The two conditions have contrasting appearances. In EG, there is typically diffuse reticulonodular shadowing in the lungs, progressing to a honeycomb appearance in the later stages of the disease (Fig 6). CEP responds promptly to oral corticosteroid treatment. The long-term prognosis is excellent but the majority of the patients will require long-term low-dose oral corticosteroid therapy in order to prevent relapse[7].

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ABSTRACT

A 24-year-old Chinese woman presented with cough, chest pain, weight loss, low grade fever and bronchial breath sounds on auscultation. The diagnosis of chronic eosinophilic pneumonia was made on characteristic systemic and pulmonary clinical manifestations, blood eosinophilia and the striking chest radiographic appearance. This rare, idiopathic but benign condition responds well to corticosteroid treatment and the long term prognosis is excellent. The typical chest radiographic pattern of 'photographic negative of pulmonary oedema' in this condition is emphasised.

Keywords: *chronic eosinophilic pneumonia, air-space consolidation, lung diseases*