

PHOTOCLINIC

Figure 1

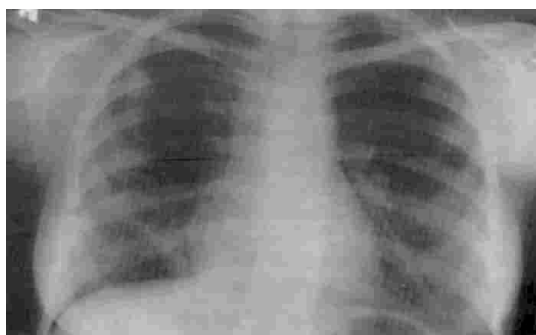
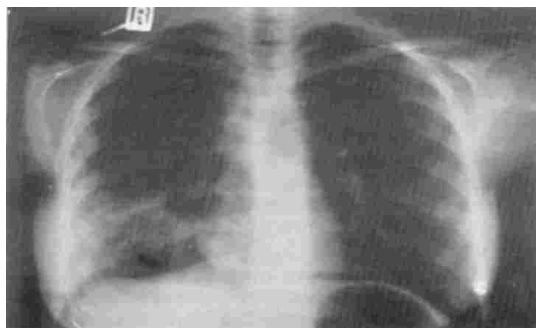


Figure 3

A 19-year-old lactating woman was referred to our hospital with cough, scanty sputum, increasing dyspnea, fever, night sweats, lassitude, and loss of weight for 8 months. Her illness started during pregnancy. Drug history was negative and she denied any personal or family history of tuberculosis, asthma or atopy. In physical examination, she seemed to be wasted. No adenopathy or organomegaly was documented and her heart and lungs were normal. Chest X-ray showed a patchy non-homogenous infiltrate in the lower part of the right lung (Figure 1) which was increased in size over time and involved the peripheral portion of the right hemi-

Figure 2

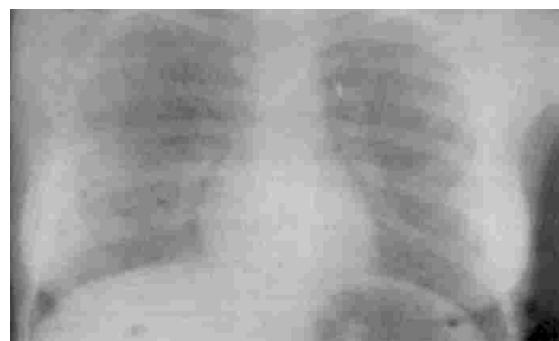
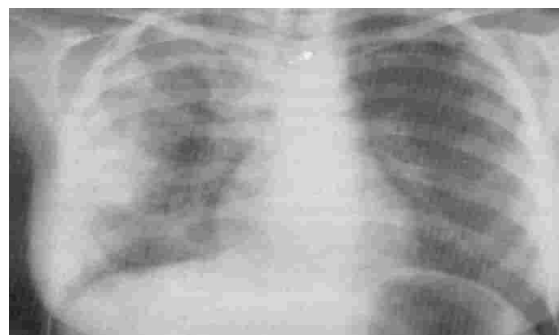


Figure 4

thorax with no evidence of significant lymphadenopathy in 3 weeks (Figure 2).

Liver function tests and routine chemistry were normal; Hb: 13g/dL, WBC: 18,400/ μ L with 30% PMN, 3% monocytes, 47% eosinophils, and 20% lymphocytes; platelets: 320,000/ μ L; ESR: 90 and 115 at the end of first and second hours respectively. Three consecutive stool examinations were negative for occult blood and parasites. Pulmonary function tests showed a severe restrictive pattern with FVC: 0.83/L (24% of normal), FEV₁: 0.75/L (24% of normal), and EFV₁/FVC=0.9.

Your Diagnosis?

See page 157 for diagnosis

Photoclinic Diagnosis: Chronic (Cryptogenic) Eosinophilic Pneumonia (CEP)

The patient's prominent constitutional symptoms, accompanied by peripheral pulmonary opacities associated with high eosinophilia and high sedimentation rate are in favor of CEP. Allergic bronchopulmonary aspergillosis and Churg-Strauss syndrome were ruled out due to absence of asthma, atopy and vasculitis. Loeffler's syndrome was excluded because of protracted course of the illness. Lack of any drug history made eosinophilic drug reaction unlikely. There was no finding in favor of parasitic infestation or hypereosinophilic syndrome. The patient was treated with steroids. Clinical improvement was noticed after the first dose of steroid. CXR showed dramatic improvement after 7 days (Figure 3) with complete resolution after one month (Figure 4). Three weeks after steroid therapy, great improvement in the pulmonary function tests and hematologic indices were also achieved as follows: FVC: 2.73l (77% of normal), FEV₁: 2.34 (75% of normal), Hb: 12.5g/dL, WBC: 10,800/μL with 70% PMN, 21% lymphocytes, 4% monocytes and 5% eosinophils.

CEP is an uncommon disease of unknown etiology. Activation of eosinophils plays a major role in its pathogenesis. Activated eosinophils with eosinophil-derived granule proteins (EDGP) has been detected in the lung tissue and bronchoalveolar lavage (BAL) fluid of the patients.¹ It has female predominancy and may begin in pregnancy. The disease is more frequent in the fourth decade of life and 1/3 to 1/2 of patients have asthma or atopy which may precede or concur with CEP. The chronic clinical course and the presence of constitutional symptoms render the disease similar to tuberculosis or fungal infections. CXR findings also may be confused with chronic lung infections, sarcoidosis and cryptogenic organizing pneumonia. Peripheral distribution of infiltrate, sometimes bilateral and dense, defined as "photographically negative pulmonary edema" is the pathognomonic sign of this condition.^{1,2}

Pulmonary function tests in CEP show a restrictive pattern with varying intensity. Patients with an asthmatic component may also have obstructive ventilatory defects.^{1,2} Providing adequate clinical, radiologic, and laboratory data, pathologic examination is not necessary, but in suspicious cases, study of lung tissue or bronchoalveolar lavage (BAL) fluid is necessary to rule out other possibilities. The characteristic pathological feature in CEP is the infiltration of the alveolar spaces and walls by eosinophils. In some cases, microabscesses and obliterative bronchiolitis have also been observed.^{1,2,3} Corticosteroids are the mainstay of therapy. Moderate doses are sufficient and dramatic response to these drugs is one of the diagnostic tests of the CEP. Complete recovery is usually seen in a few weeks, but steroids should be tapered very slowly to prevent relapse. Long-term use of steroids as low-dose, daily, or alternate day regimens, is usually necessary in the treatment of the majority of patients.^{1,3}

References

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2. Fraser RG, Pare JAP, Pare PD, Fraser RS, Genereux GP. *Diagnosis of Diseases of the Chest*. 3rd ed. Philadelphia: WB Saunders; 1989: 1290-8.
3. Taylor AN. Pulmonary eosinophilia: the eosinophilic pneumonia. In: Du Bois RM, Olivieri D, eds. *Interstitial Lung Disease. European Respiratory Monograph*. Vol 5. Sheffield, UK: European Respiratory Journals; 2000: 206-225.

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