

Chronic Eosinophilic Pneumonia: A Diagnostic Challenge



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A previously healthy 38-year-old female nonsmoker presented to hospital with a 10-month history of progressive dry cough, shortness of breath, weight loss, and fevers. She had been diagnosed with recurrent community-acquired pneumonia, although multiple courses of antibiotics had led to minimal improvement. On examination, the patient required supplemental oxygen, and auscultation revealed decreased breath sounds and crackles bilaterally.

Initial bloodwork revealed an elevated white blood cell count of $44.2 \times 10^9/L$, with an absolute eosinophil count of $35.4 \times 10^9/L$, and an elevated IgE level of 3757 UI/mL. Pulmonary function tests (PFTs) were unremarkable. Chest x-ray revealed extensive bilateral mixed interstitial alveolar processes in the mid- and upper-lung zones (Figure 1). High-resolution computed tomography (HRCT) of the chest subsequently demonstrated bilateral patchy opacities in a peripheral distribution in the upper- and mid-lung zones, and relative sparing of lower lung zones (Figure 1). Bronchoscopy with bronchoalveolar lavage (BAL) was significant only for an “abundance of eosinophils”; a differential cell count was obscured by the presence of bronchial cells. Once other causes of eosinophilic lung disease were excluded, a diagnosis of chronic eosinophilic pneumonia (CEP) was made and the patient was started on prednisone, with excellent clinical response.

CHRONIC EOSINOPHILIC PNEUMONIA

In 1969, Carrington et al first described “a most unusual peripheral distribution of the shadows”¹ on chest x-ray, with accompanying eosinophil focus on lung biopsy. CEP has since been defined as an idiopathic chronic condition marked by alveolar filling with mixed inflammatory infiltrate, consisting largely of eosinophils.² CEP characteristically affects patients in their third or fourth decade of life, with an insidious presentation of fever, weight loss, night sweats, chronic dry cough, and progressive shortness of breath over the course of months.³ Peripheral eosinophilia is typically pronounced, and IgE levels may be elevated. Chest imaging with HRCT classically reveals bilateral nonsegmental consolidations in a peripheral distribution

favoring the upper lung zones, described as the photographic negative of pulmonary edema.¹ Nodules, ground-glass opacities, and pleural effusions are uncommon. The diagnosis of CEP should not be made solely on the radiographic pattern, as imaging findings of other respiratory pathology overlap with those of CEP. Most notable among these include cryptogenic organizing pneumonia, drug reactions, other eosinophilic lung disease, and sarcoidosis. When CEP is suspected based on clinical history and investigations, bronchoscopy is usually the key to diagnosis, with BAL eosinophil count virtually always greater than 25%.³ Exclusion of other causes of eosinophilic pneumonias is essential. PFTs bestow limited diagnostic utility, and surgical lung biopsy is generally reserved for cases of diagnostic uncertainty.³

The hallmark of CEP is a profound and rapid response to oral corticosteroids, often within 48 hours. Recurrences commonly occur months to years after the initial event, particularly after cessation of therapy. The patient presented herein had a marked response to therapy, but remains steroid dependent since diagnosis and treatment in 2013.

This case highlights the need to broaden differential diagnoses when a patient's clinical history is incongruent from an expected course, for example, a failure to respond to repeated antibiotics and bronchodilators. The average duration of symptoms in CEP is 7.7 months before a diagnosis is made,³ suggesting that improved recognition of its characteristic clinical and radiographic features may help avoid significant morbidity and potential mortality.

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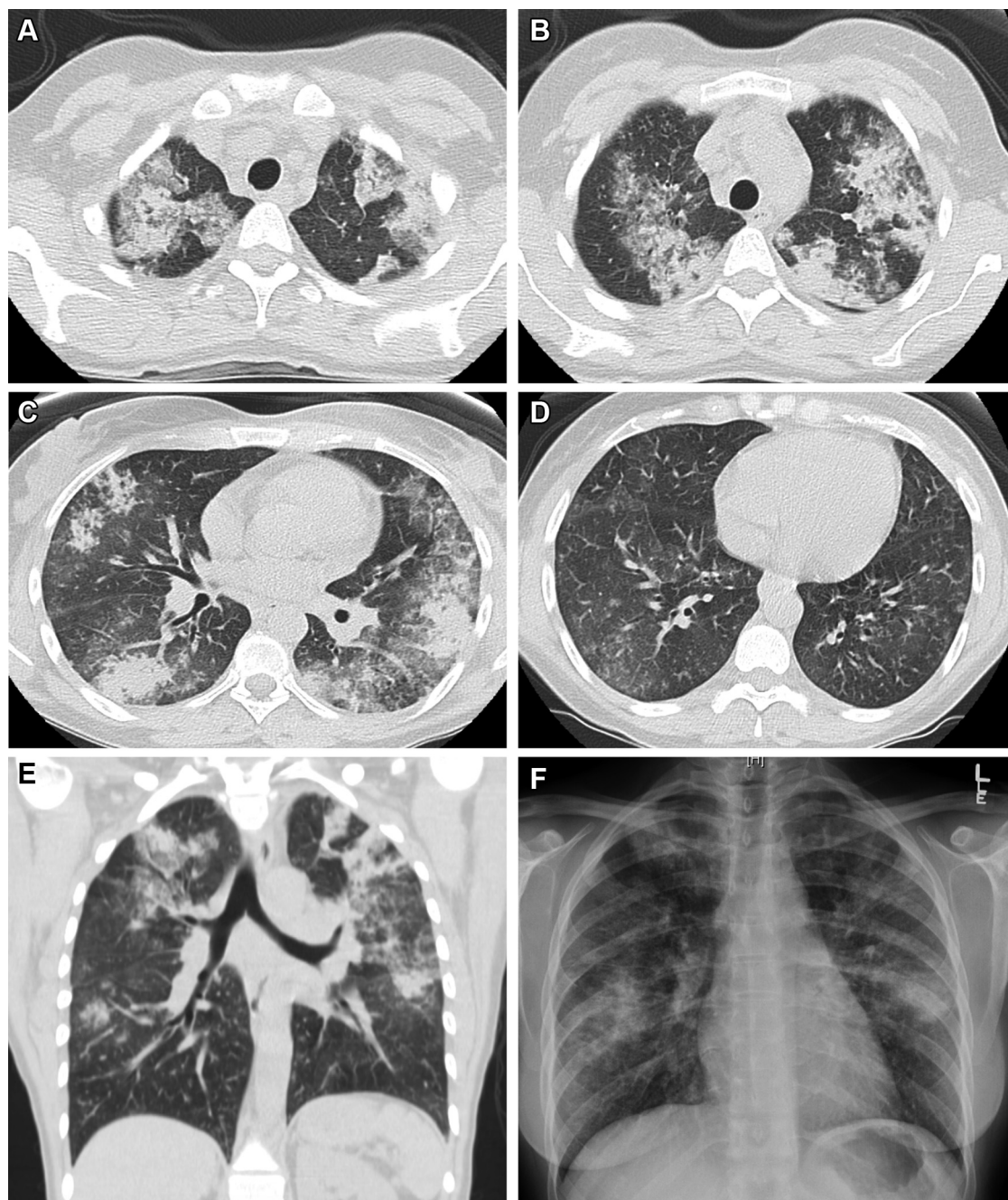


FIGURE 1. A-E, High-resolution computed tomography of the chest demonstrating bilateral patchy peripheral regions of consolidation predominantly involving the upper- and mid-lung zones, with D, relative sparing of lower-lung zones. F, Extensive bilateral mixed interstitial alveolar processes localized to the mid-upper lung zones are seen on x-ray. These radiographic findings are classic for chronic eosinophilic pneumonia. Other leading differential diagnoses should include cryptogenic organizing pneumonia, drug reactions, other eosinophilic lung disease, and sarcoidosis.

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