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Authors

Oh, Scott

Kim, James

Lian, Kuo-Chiang

et al.

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CLINICAL VIGNETTE

Cough, Dyspnea, and Eosinophilia: A Case of Chronic Eosinophilic Pneumonia

Scott Oh DO^a, James Kim MD^b, Kuo-Chiang Lian MD^b, Irawan Susanto MD^a

^aUCLA Division of Pulmonary and Critical Care Medicine, ^bUCLA Department of Internal Medicine

Case Report

An 82-year-old woman with a history of COPD and allergic rhinitis presented to the emergency department with shortness of breath and cough. She experienced similar symptoms one year prior to presentation, which resolved after six months and several courses of antibiotics. She had since been doing well until her dyspnea and cough returned one month ago. Her cough became productive of white, frothy sputum without hemoptysis. Fevers up to 103°F were measured at home in addition to drenching night sweats and diaphoresis for approximately 2 weeks prior to admission despite several outpatient courses of antibiotics including amoxicillin/clavulanate and azithromycin prompting further evaluation. The patient reported some anorexia but no recent change in weight.

Past medical history includes hearing loss, hypertension and GERD. She has no known drug allergies. Home medications included budesonide, lisinopril, esomeprazole, amlodipine, atenolol, montelukast, estradiol and azelastine nasal spray.

She lives alone in Los Angeles and is originally from Iran. She moved to the United Kingdom over 40 years ago then moved to the US about 30 years ago. She is a retired secretary. There is no history of tobacco, ethanol, or illicit drug use. She has no pets or a history of significant exposures. Last travel was to Iran in the fall of 2010.

Vitals revealed a temperature of 37.1°C, pulse of 94, blood pressure of 96/54, respiratory rate of 14, and an oxygen saturation of 97% on room air. She was in no distress and sitting comfortably in a chair. Exam was notable for comfortable breathing with minimal bibasilar expiratory wheezing. The remainder of her exam was unremarkable. Initial laboratories were notable for a leukocytosis of $19.3 \times 10^3/\mu\text{L}$ with 38% eosinophils. A chest film was

notable for borderline cardiomegaly, bronchial wall thickening with ground glass, patchy, confluent and nodular airspace opacities in an upper lobe predominant distribution (Figure 1). Levofloxacin started in the emergency department was continued for community acquired pneumonia. High resolution CT chest (HRCT) without contrast was obtained demonstrating dense diffuse airspace consolidations greatest in the right upper lobe, linear densities in the right upper lobe, and mild mediastinal lymphadenopathy (Figure 2).

Figure 1: Initial chest film

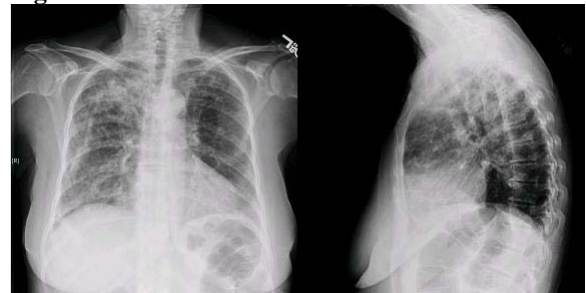


Figure 2: HRCT of the chest



Further work-up including rheumatologic serologies were negative including ANA, c-ANCA, p-ANCA, proteinase-3-antibody, and myeloperoxidase antibody. Infectious work up including blood cultures, stool ova and parasites were also negative as was HIV, histoplasma urine antigen, strongyloides antibody, cryptococcal antigen, aspergillus antigen, and coccidioidomycosis IgG/IgM. QuantiFERON-TB Gold returned positive but sputum AFB stains were

negative. Treatment for latent tuberculosis was started after the third negative AFB.

Pulmonary and Hematology were consulted. A JAK-2 mutational analysis was negative and serum tryptase was within normal limits. Hematology did feel that the patient had a primary hematologic or neoplastic disease.

Bronchoscopy did not reveal significant endobronchial findings. Bronchoalveolar lavage (BAL) and transbronchial biopsies were performed. All microbiologic studies including those for bacterial, fungal, mycobacterial and viral pathogens were negative. BAL cell count and differential was notable for inflammatory cells with 17% eosinophils. Transbronchial biopsies demonstrated an eosinophilic infiltrate of the alveolar parenchyma as well as in some small and medium sized vessels without evidence of a necrotizing vasculitis (Figure 3).

Figure 3: Transbronchial biopsies demonstrating alveolar eosinophilic infiltrate.

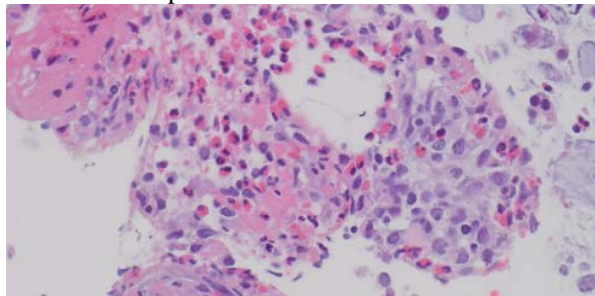
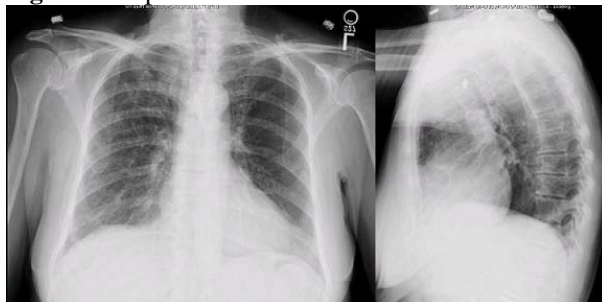


Figure 4: Repeat chest film after treatment



The patient was discharged on hospital day number 10 on prednisone 40mg daily and has been doing well. Prednisone was eventually able to be titrated down to 5mg. However, five months after discharge she experienced a relapse and prednisone was increased back to 40mg daily.

Discussion

Chronic eosinophilic pneumonia is an idiopathic disorder characterized by the accumulation of eosinophils in the lung. It was first described as a clinical syndrome in 1969¹. Published case series ranging from 12 to 62 cases have further characterized the syndrome²⁻⁴. Presenting symptoms are typically subacute and non-specific including cough, dyspnea, wheezing, fever, night sweats and weight loss²⁻⁴. It occurs more frequently in women and smoking is thought to exert a protective effect as the disease is seen predominantly in non-smokers²⁻⁴. Peripheral eosinophilia of >5% was noted in 88% of 111 cases based on a literature review². This may be a key initial finding on routine evaluation prompting further evaluation, as other findings are often non-specific.

Results of chest imaging are non-specific and consistent with pneumonia. Findings include air space disease with an alveolar pattern in a peripheral distribution in the majority of cases^{1-3, 5-8}. These findings have been described to be migratory in nearly 25% of cases³. The classic and almost pathognomonic "photonegative" pattern of distribution, described as the reverse of pulmonary edema, is seen only in about 25% of cases²⁻⁷. These densities are thought to develop into band like opacities, as seen in this case⁶. Pleural effusions are uncommon. Small pleural effusions were noted in 10% of cases on HRCT of the chest¹⁰. Cavitation is unusual and should prompt consideration of an alternate diagnosis.

Cell count with differential on bronchoalveolar lavage samples obtained via bronchoscopy consistently demonstrates significant alveolar eosinophilia often >50%^{3, 9}. Clinicians should be aware this can quickly drop within days of treatment with steroids¹⁰. Histopathologic findings include eosinophils and histiocytes in the interstitial and alveolar spaces, as seen in this case². Fibrotic changes are minimal but a component of organizing pneumonia is often seen².

The disease consistently responds to corticosteroids but most require more than 6 months of treatment. More than 75% experience a relapse while titrating down steroids or after discontinuation², as was seen in this case. Symptomatic improvement is often seen within 48 hours, as was the case with our patient³. Radiographic abnormalities also respond quickly with reports of resolution of chest film findings within 1 week in more than half of cases³. The optimal dose of corticosteroids is not well established but typical doses range from 20-60mg of prednisone

per day. Again, relapses are common and prolonged courses of greater than 6 months are often required. In patients who are unable to be weaned off oral corticosteroids, inhaled corticosteroids may help reduce the required oral dose¹¹.

Conclusion

Chronic eosinophilic pneumonia is an atypical cause of pneumonia. Initial presentation and findings are often non-specific and require a high index of suspicion for diagnosis. Findings, which should prompt consideration include peripheral eosinophilia, recurrent pneumonias, radiographic findings including a classic "photonegative" pattern which may be migratory, and band-like densities. The diagnosis can be confirmed with bronchoscopy revealing marked eosinophilia on bronchoalveolar lavage prior to the initiation of steroids. The disease is consistently steroid responsive but relapses are common requiring prolonged courses of > 6 months. The addition of inhaled corticosteroids may reduce the required oral dose in those unable to be weaned off systemic steroids.

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