Unusual Presentation of Thoracic *Pneumocystis carinii* Infection in a Patient with Acquired Immunodeficiency Syndrome

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Pleura-based masses and hilar adenopathy were seen on a chest radiograph of a patient with acquired immunodeficiency syndrome who had a history of *Pneumocystis carinii* infection. The differential diagnosis of such a presentation is discussed in light of atypical and extrapulmonary manifestations of *P. carinii* infection in a patient receiving prophylaxis with dapsone.

We report an unusual case of *Pneumocystis carinii* infection that illustrates the myriad ways that this infection can manifest in an HIV-infected patient. This case is also noteworthy because the patient was receiving systemic prophylaxis for *P. carinii* pneumonia (PCP).

**Case report.** A 43-year-old woman presented in February 1997 with a 4-month history of productive cough, dyspnea on exertion, fevers, chills, weight loss, and fatigue. HIV infection had been diagnosed in 1988, and she had an episode of PCP in 1994. Since 1994, she had received maintenance PCP prophylaxis with dapsone (100 mg/d) because of a history of pruritic reaction to trimethoprim-sulfamethoxazole (Bactrim; Roche). She was enrolled in a trial of protease inhibitor therapy in July 1995. However, she discontinued antiretroviral therapy in August 1996 after the study was completed.

A chest radiograph done at the time of presentation showed bilateral hilar and subcarinal adenopathy, basilar linear and nodular opacities, and pleura-based masses (figure 1). A CT scan of the chest showed, in addition to the above, multiple parenchymal nodules and extensive multifocal pleural disease (figure 2, top and bottom). Fine-needle aspiration of a left pleura-based lesion showed foamy alveolar casts consistent with *P. carinii*. Results of a gram stain and stains for acid-fast bacilli (AFB) and fungi were negative.

An outpatient follow-up chest radiograph showed the increasing size of the right pleura-based opacity and new bibasilar opacities. Subsequently, bronchoalveolar lavage showed numerous foamy alveolar casts suggestive of *P. carinii*. Special stains confirmed this diagnosis. Results of stains and cultures were negative for cytomegalic viral inclusions, fungi, and AFB. The patient was admitted because of worsening respiratory status after the bronchoscopy.

There was no history of tuberculosis, and the result of a recent PPD test was negative. There was a 5-year history of residence in Arizona, but the patient denied any history of coccidioidomycosis.

Examination on admission showed the patient to be in mild respiratory distress, with an oxygen saturation of 88% on 4 L/min of supplemental oxygen. She had bibasilar crackles on
Figure 2. CT scans of the chest (top and bottom) at presentation show pleura-based opacities and pulmonary nodules as well as subcarinal adenopathy.

Figure 3. Chest radiography 1 year later showed resolution of adenopathy and pleural and parenchymal opacities (figure 3).

Discussion. The commonly seen radiographic picture of pulmonary pneumocystosis is that of bilateral perihilar “ground glass” opacities [1]. However, no appearance is pathognomonic of \textit{P. carinii} infection [1]. Our patient presented with an unusual pattern of thoracic involvement due to this infection that manifested as peripheral pleura-based masses and intrathoracic adenopathy, a presentation that has not been reported previously.

Pleural involvement due to \textit{P. carinii} has been described in reports of extrapulmonary pneumocystosis [2], of patients with AIDS who have spontaneous pneumothoraces [3], and of pleural effusions seen with PCP [4]. Spontaneous pneumothoraces

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have been reported to occur in 4%-5% of patients with PCP, and patients receiving pentamidine prophylaxis are at higher risk for this complication [4]. Thoracoscopic in some of these patients has revealed pleural involvement in the form of visceral and parietal nodules, of which a biopsy reveals the presence of *P. carinii* [3]. Although the incidence of pleural effusions in PCP ranges from 1.7% to 18.3% in different reports, pleural effusions due to *P. carinii* infection are rare, occurring in close to 2% of all patients with PCP [4].

Pleural disease due to *P. carinii* appears to occur in patients with significant parenchymal lung disease, with subpleural necrosis and cyst formation predisposing to the occurrence of pneumothoraces [4]. Multiple peripheral pleura-based masses, like those we saw in our patient, have never been reported. A solitary peripheral pleura–based mass due to *P. carinii* has been reported once before by investigators documenting the diagnostic yield of cytological techniques for patients with HIV disease [5]. It is noteworthy that our patient developed a spontaneous pneumothorax on the right side, which was likely secondary to extensive subpleural lung involvement due to *P. carinii* [4].

Lymph node involvement is the most common extrapulmonary site of pneumocystis infection in HIV-infected patients [2]. A small series of patients with noncalcified mediastinal adenopathy due to *P. carinii* infection has been described [6]. Intrathoracic adenopathy is seen on chest CT scans of 18% of patients with PCP, although this is infrequently seen on chest radiographs [1]. Few reports, however, have systematically assessed the importance of intrathoracic adenopathy in patients with advanced HIV disease [7]. Intrathoracic adenopathy usually indicates serious disease and does not represent the benign reactive hyperplasia of diffuse, persistent lymphadenopathy syndrome [8].

Recent studies have found that infectious causes, predominantly *Mycobacterium tuberculosis* and *Mycobacterium avium* complex, account for 50%-60% of thoracic adenopathy [7]. Malignancy, mainly lymphoma, bronchogenic carcinoma, and Kaposi’s sarcoma, accounts for the rest [7]. Nonspecific inflammation resulting in thoracic lymphadenopathy has also been reported in a few instances, although the long-term outcome for these patients has not been discussed [7]. Because of our patient’s unstable condition, we did not perform a mediastinoscopy. On a follow-up chest radiograph, our patient showed complete resolution of thoracic adenopathy following specific treatment for PCP, indicating that the thoracic adenopathy most likely occurred because of *P. carinii* infection. Another possible explanation for the thoracic adenopathy is nonspecific/reactive inflammation that might have responded to steroid therapy, which was administered for 3 weeks. Although the administration of highly active antiretroviral therapy (HAART) alone can result in resolution of opportunistic infections in patients with AIDS, the duration of antiretroviral therapy for our patient was too brief (2 weeks) to have likely resulted in sustained immune-modulating effects.

The combination of pleura-based masses and intrathoracic adenopathy in HIV-infected patients has been most commonly seen with AIDS-related non-Hodgkin’s lymphoma [9]. Although AIDS-related Kaposi’s sarcoma involving the lung can involve the pleura and the intrathoracic nodes, the findings of peripheral pleura–based masses and adenopathy are very unusual [10]. Coccidioidomycosis was considered as a diagnosis for our patient because of a history of residence in an area where it is endemic. The latter consideration and the unusual presentation of *P. carinii* infection delayed the initiation of systemic steroid therapy for our patient.

Atypical and extrapulmonary pneumocystosis have been reported with increasing frequency since the onset of the AIDS epidemic [2]. An increased frequency of extrapulmonary pneumocystosis has been noted among users of aerosolized pentamidine [2]. It has been suggested that pentamidine in an aerosolized form distributes locally in the lung and does not afford protection against the systemic spread of *P. carinii* infection. The lack of penetration of aerosolized pentamidine into distal lung parenchyma may also account for the increased occurrence of spontaneous pneumothoraces in patients with PCP [3, 4]. Only 2 cases of extrapulmonary pneumocystosis involving AIDS patients receiving systemic prophylaxis for PCP have been reported [11, 12]. Both of these patients were receiving prophylaxis with dapsone (100 mg/d) and pyrimethamine (25 mg/d).

Our patient also had extrapulmonary spread of *P. carinii* infection to the pleura and probably to the intrathoracic lymph nodes. Like previously reported patients, she was also receiving dapsone prophylaxis at a dosage of 100 mg/d. Again, similar to prior reports, our patient was not receiving antiretroviral therapy. The rarity of extrapulmonary disease in reported studies of dapsone prophylaxis may be due to the frequent use of antiretroviral therapy [13]. Recent studies have shown that HAART significantly reduces pneumocystis infections, even in the absence of any specific prophylactic therapy for PCP [14].

The propensity for *P. carinii* infection in HIV-infected immunodeficient patients is well known. In an era of changing systemic prophylaxis for PCP, this report reminds clinicians that *P. carinii* can present with atypical manifestations in HIV-infected patients, especially in those not receiving antiretroviral inhibitors.

References


