Upper zone cystic lung disease in HIV related Pneumocystis carinii pneumonia

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Abstract
Although unusual, focal upper zone air space opacification and cyst formation in Pneumocystis carinii pneumonia and HIV infection can occur and may mimic "classical" tuberculosis.

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Pneumocystis carinii pneumonia (PCP) is the most common HIV related pulmonary infection in both Great Britain and the USA. We report a patient with upper zone opacification and cavitory disease caused by PCP in HIV disease mimicking "classical" tuberculosis. This is clinically important as tuberculosis is a major infectious pulmonary complication of HIV infection. Because of large overlap of the clinical and radiological signs in HIV related conditions, it is important to reach a definitive diagnosis in HIV infection.

Case report
A 37 year old man presented with a four month history of a non-productive cough, progressive exertional dyspnoea, and night sweats. Recent travel abroad had included a four month visit to Florida, USA. The patient looked well. Small lymph nodes were palpable in the left axilla. The presentation chest radiograph showed bilateral diffuse upper zone airspace opacification with widespread cavitation. Both hila were elevated but there was no hilar lymphadenopathy or pleural effusion (fig 1). A preliminary diagnosis of tuberculosis was made and antituberculosis chemotherapy was started. Results of further investigations at presentation were: Hb 10.9 g/dl, WBC 6.3 × 10⁹/l, granulocytes 4.7 × 10⁹/l, lymphocytes 1.3 × 10⁹/l, platelets 347 × 10⁹/l. Urea and electrolyte levels and liver function test results were normal. The CD4 count was 123/mm³ (410–1540/mm³), and the CD8 count 728/mm³ (230–1100/mm³). The patient was HIV antibody positive. A bronchoscopy was performed but no acid fast bacilli, P carinii or other organisms were identified in bronchial washings.

Despite antituberculosis chemotherapy the condition of the patient deteriorated rapidly over the following five days. He developed a high swinging pyrexia, tachycardia and tachypnoea, with marked central cyanosis and hypoxia (SaO₂ on air 66%). The chest radiograph now showed an increase in the density and extent of the airspace opacification involving the upper and mid zones of both lungs. A further bronchoscopy with bronchoalveolar lavage was performed. The patient was considered too ill for transbronchial biopsy. The endobronchial mucosa appeared normal and no organisms were identified in the bronchial washings. An open lung biopsy was therefore performed and histological examination showed an interstitial pneumonitis in which alveolar septa were extensively infiltrated by mononuclear and plasma cells. In the smaller airways P carinii organisms were seen with Grocott staining. No fungi or acid fast bacilli were identified. Intravenous co-trimoxazole and hydrocortisone were commenced and the antituberculosis chemotherapy stopped. The patient made a gradual recovery over a period of three weeks and was discharged from hospital feeling well.

High resolution computed tomographic examination of the thorax performed after treatment showed the size and distribution of the lung cysts (fig 2).

Discussion
PCP is the most common HIV related pulmonary infection in Great Britain and the USA, occurring in 60–80% of patients with AIDS. Typically it presents as bilateral perilobular and/or basal reticulonodular or ground glass opacities which progress in 2–7 days to diffuse airspace opacification, initially sparing the apices but finally progressing to complete consolidation of the entire lung.

Atypical patterns are reported in less than 5% of patients in most series and include unilateral focal consolidation, cystic disease, predominant upper lobe involvement, localised nodular densities, solitary pulmonary nodules, hilar and mediastinal lymphadenopathy, and pleural effusions. These atypical findings should initially suggest an alternative diagnosis. For example, hilar and mediastinal
lymphadenopathy in HIV seropositive patients is most commonly caused by mycobacterial or fungal infection, Kaposi's sarcoma and, rarely, lymphoma or metastatic spread from a bronchogenic carcinoma.3

In a review of 100 consecutive patients with AIDS with PCP, pulmonary air filled cysts or pneumatoceles were identified in 10%.4 There are, however, very few reports elsewhere in the literature which recognise PCP as a cause of this radiographic pattern. Computed tomography is much more sensitive than plain radiography in detecting cysts, well shown by one series in which 40% of patients with cystic abnormalities were only identified by computed tomography even when the plain radiographs were reviewed retrospectively.3 Initially the cysts appear as small foci in areas of parenchymal consolidation, typically within the upper lobes and superior segments of the lower lobes. They may coalesce to form bizarre shaped, thick walled cystic lesions that frequently appear to have septa. With therapy these lesions eventually regress and may completely resolve.1

Upper zone consolidation caused by PCP in HIV disease is rare and may mimic "classical" tuberculosis.6 This is clinically important since tuberculosis is a major infectious pulmonary complication of HIV infection.7 An association between inhaled pentamidine prophylaxis and predominant or isolated distribution of PCP in the upper zones has been reported,6 but the mechanism is unclear. A possible explanation is that aerosolised pentamidine fails to reach the upper lobes in sufficient concentration to prevent the development of upper lobe PCP.

Fibroepithelial bronchoscopy is the procedure of choice for diagnosing pulmonary opportunistic infections in HIV seropositive patients. The relative sensitivity of bronchoalveolar lavage and transbronchial biopsy for detecting P carinii varies between series; however, bronchoalveolar lavage has been reported to be the more effective method because at least 50 cm³ of lung is sampled compared with less than 1 cm³ by transbronchial biopsy,9 and the organisms lie almost entirely within the alveolar space. Fibroepithelial bronchoscopy and transbronchial biopsy have a higher complication rate in HIV seropositive patients than in the general population.10 Complications are most commonly associated with transbronchial biopsy and include haemorrhage and pneumothorax, particularly in patients whose lungs are infected with P carinii.10 Because of the rapid deterioration of our patient, the relatively low yield of additional diagnoses produced by transbronchial biopsy, and the risk of complications, transbronchial biopsy was not performed before open lung biopsy. An increased sensitivity and specificity of the bronchoalveolar lavage fluid for P carinii might have been achieved by the use of indirect fluorescent antibodies or DNA amplification with ethidium bromide staining, but at the time these techniques were not available.10

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