Pneumocystis Pneumonia

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Epidemiology

Pneumocystis pneumonia (PCP) is caused by *Pneumocystis jirovecii*, a ubiquitous fungus. The taxonomy of the organism has been changed; *Pneumocystis carinii* now refers only to the *Pneumocystis* that infects rats, and *P. jirovecii* refers to the distinct species that infects humans. However, the abbreviation PCP is still the preferred acronym to designate the clinical syndrome of *Pneumocystis* pneumonia,[1] although PJP is commonly used. Initial infection with *P. jirovecii* usually occurs in early childhood; two-thirds of healthy children have antibodies to *P. jirovecii* by age 2 years to 4 years.[2]

Rodent studies and case clusters in immunosuppressed patients suggest that *Pneumocystis* spreads by the airborne route. Disease probably occurs by both new acquisition of infection and by reactivation of latent infection.[3-12] Before the widespread use of PCP prophylaxis and antiretroviral therapy (ART), PCP occurred in 70% to 80% of people with advanced HIV,[13] with a 20% to 40% mortality rate in individuals despite anti-*Pneumocystis* therapy. Approximately 90% of PCP cases occur in people with HIV with CD4 T lymphocyte (CD4) cell counts <200 cells/mm^3.

The incidence of PCP has declined substantially with widespread use of PCP prophylaxis and ART; incidence among people with HIV in Western Europe and the United States is <1 case per 100 person-years.[14-16] Most cases of PCP now occur in people with HIV who are unaware of their HIV status or are not receiving ongoing care for HIV,[17] and in those with advanced immunosuppression (i.e., CD4 counts <100 cells/mm^3).[18]

Clinical Manifestations

In people with HIV, the most common manifestations of PCP are subacute onset of progressive dyspnea, fever, non-productive cough, and chest discomfort that worsens within days to weeks. The fulminant pneumonia observed in people who do not have HIV is less common among people with HIV. A more fulminant course can occur particularly after initiation of therapy.[19-21]

In mild cases, pulmonary examination while the patient is at rest usually is normal. With exertion, tachypnea, tachycardia, and diffuse dry (cellophane) rales may be observed.[20] Fever is present in most cases and may be the predominant symptom in some people. Pneumonia limited to the apices and extrapulmonary disease, which can occur in any organ, are rare and have been associated with use of aerosolized pentamidine prophylaxis.[22]

Hypoxemia, the most characteristic laboratory abnormality, can range from mild (room air arterial oxygen partial pressure [PaO_2] ≥70 mmHg or alveolar-arterial gradient [A-a gradient] <35 mmHg) to moderate (A-a gradient ≥35 to <45 mmHg) to severe (A-a gradient ≥45 mmHg). Oxygen desaturation with exercise is often abnormal but is nonspecific.[23] Elevation of lactate dehydrogenase levels to >500 mg/dL is common but also nonspecific.[24] The chest radiograph typically demonstrates diffuse, bilateral, symmetrical “ground-glass” interstitial infiltrates emanating from the hila in a butterfly pattern[20]; however, in people with HIV with early disease, a chest radiograph may be