**Pneumocystis carinii** Pneumonia in a Human Immunodeficiency Virus–Uninfected Patient with Sickle Cell Crisis

*Pneumocystis carinii* pneumonia (PCP) usually occurs in the setting of preexisting immunosuppression. We present a case of PCP that occurred in an HIV-negative woman with severe sickle cell disease.

We present a case of *Pneumocystis carinii* pneumonia (PCP) that occurred in a woman with sickle cell pain crisis but no clinical evidence for a predisposing immunodeficiency state. PCP is the most common opportunistic infection in acquired immunodeficiency syndrome (AIDS) patients; before 1981 it was a rare condition [1–3]. *Pneumocystis carinii* was first recognized in Europe during World War II as a cause of interstitial plasma cell pneumonia, a disorder common in institutionalized, premature, and debilitated infants [4]. Before the AIDS epidemic, most patients with PCP were either malnourished or were receiving immunosuppressive therapy for various conditions, with neutoplasms, organ transplantations, or a disorders treated with corticosteroid or cytotoxic drugs being the most common [5, 6]. Reports of PCP in well-nourished immunocompetent adults remain extremely rare [7–11]. We conducted a MEDLINE search of literature published between 1966 and 1999, using the search terms “*Pneumocystis carinii* pneumonia,” “opportunistic infections,” and “sickle cell disease,” and were unable to discover an antecedent case.

A 30-year-old black woman with transfusion-dependent sickle cell anemia and known pulmonary hypertension was admitted with a probable sickle cell–related “acute chest syndrome.” She was given broad-spectrum antibiotics, patient-controlled analgesia, saline hydration, and a red cell transfusion. Her complete blood count was remarkable for moderate hemolysis and leukocytosis (52% neutrophils, 32% lymphocytes, 12% monocytes, 3% eosinophils, and 1% basophils). Chemical analysis of serum found nothing remarkable, with the exception of a lactate dehydrogenase level of 354 IU/L. A central line analysis of serum found nothing remarkable, with the exception of 12% monocytes, 3% eosinophils, and 1% basophils). Chemical analysis of serum was negative for bacteria, viruses, acid-fast bacilli, and fungi. Antimicrobial therapy was changed to trimethoprim-sulfamethoxazole and corticosteroids. The patient showed steady clinical improvement, was extubated, and was subsequently discharged from the hospital.

At the time of discharge, ELISA for HIV-1/HIV-2 antibodies was negative, and HIV PCR virus load was below the level of detection. Her CD4 count was 211 cells/mm³, and a subsequent CD4 count 4 months later was 1096 cells/mm³. Repeated HIV PCR for virus load remained below the level of detection. After discharge she continued to require frequent hospitalizations for pain crises and had not developed any malignancies. The patient expired of a sickle cell chest crisis 18 months after discharge. Repeat evaluation for *Pneumocystis carinii* was negative. An autopsy was not performed.

Today, the majority of PCP cases are related to AIDS, but there are a number of non-HIV disorders that have been associated with PCP as well. Cancers and their related therapies are well-described risk factors for PCP [12–15]. Among patients with malignancy, corticosteroid use appears to be a principal risk factor.

In our patient, we were unable to identify a risk factor for her development of PCP. HIV-1 antibody and PCR virus load tests were repeatedly negative. There were 2 prior and 2 subsequent hospitalizations for intramuscular injection site abscesses requiring incision and drainage. The patient had not received any immunosuppressive or cytotoxic drugs. On admission, the patient was not biochemically malnourished (albumin level, 4.0 g/dL), although she was 9% below her ideal body weight (height, 173 cm; weight, 58.3 kg). Subsequently, unfavorable nutritional parameters developed during her 38-day hospitalization (albumin level, 2.8 g/dL; prealbumin level <70 g/dL). There were no cases of PCP in patients in adjacent rooms during the preceding 2 months. With the exception of her sickle cell disease and eventual impaired nutritional status, there were no identifiable risks for immunosuppression at the time of admission. Her CD4 count while she was in the intensive care unit was low (211 cells/mm³) and can be explained by the severity of her acute illness; it was possibly sufficient to result in the development of her PCP [16]. Her CD4 count was 1096 cells/mm³ 4 months after discharge.

In summary, this is the first described case of PCP that occurred in a patient with sickle cell anemia who didn’t have identifiable risk factors. The presentation closely resembled the clinical features associated with sickle cell disease-related “acute chest syndrome.” In our patient, progressive hypoxemia was initially attributed to progressive pulmonary hypertension, a known late-stage complication of sickle cell disease. This case highlights the difficulty of discerning on clinical grounds the occurrence of what appears to be a rare, but treatable, cause of diffuse pneumonitis and respiratory failure.
Acknowledgments

We would like to thank Dr. Thomas Ward (Chief of Infectious Disease, Portland VA Medical Center) for his critical appraisal and review and Dr. Thomas Deloughery (associate professor, Division of Hematology-Oncology) for advise and counsel.

Priscilla Le and Alan J. Hunter
Department of Medicine, Oregon Health Sciences University, Portland, Oregon

References