Pneumocystis Pneumonia Outside of The HIV Population: a Differential Diagnosis Not To Be Missed.

Grace Martinez Pena MD, Richard Jesse Durrance MD, Talal Arab MD, Samir Sarkar MD, Farshad Bagheri MD.
Department of Internal Medicine. Jamaica Hospital Medical Center. 8900 Van Wyck Expressway, Jamaica NY 11418.

Introduction

Pneumocystis jiroveci pneumonia (PCP) is one of the most well-known opportunistic complications in patients with advanced HIV (AIDS). While incidence of PCP in this population has decreased largely due to the implementation of prophylaxis and retroviral therapy, PCP has become an emerging and under-appreciated complication in patients with other forms of immunodeficiency - especially those receiving long-term corticosteroid therapy or chemotherapy. This opportunistic infection has emerged as a challenge due to the associated high mortality rate (30.6%). We present the case of a non-HIV patient undergoing treatment for chronic lymphocytic leukemia (CLL) who developed PCP.

Case

A 66-year-old male with pertinent medical history of CLL diagnosed 6 years ago, on fludarabine and cyclophosphamide, presented with shortness of breath and cough. The shortness of breath was described to occur on exertion and the cough was dry. No history of substance use was reported.

On exam, the patient was hypotensive (84/69 mmHg), severely hypoxic, and in moderate respiratory distress. Physical exam was remarkable for decreased breath sounds in the left lower lung field. Lab analysis confirmed hypoxemia and mild leukopenia with mild normocytic anemia. CT pulmonary angiography ruled out pulmonary embolism however diffuse ground glass parenchymal opacities and bilateral blebs and bullae within upper lobes were appreciated.

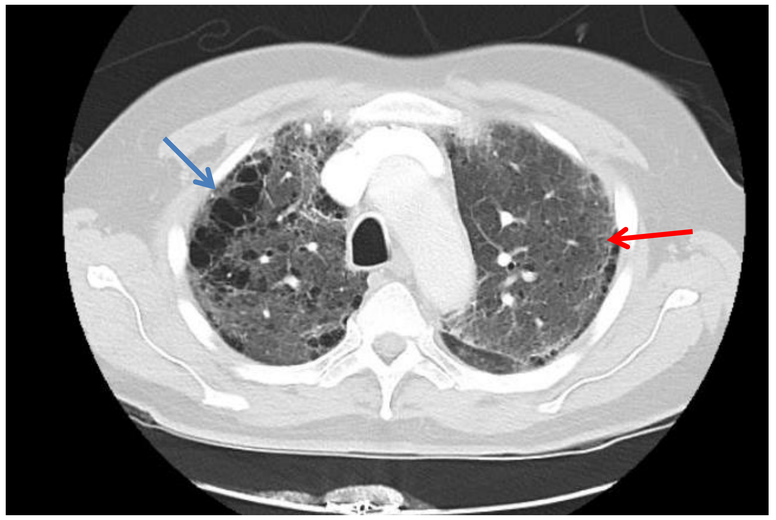

Figure 1. CT scan of lungs showing bilateral ground glass opacities (red arrow) and blebs (blue arrow).

The patient was started on empirical antibiotics and supportive treatment for healthcare associated pneumonia. Given the poor clinical evolution and concurrent chemotherapy, there was a concern of opportunistic infection. Antibiotics were switched to trimethoprim-sulfamethoxazole, and bronchoscopy produced samples with markedly elevated LDH, but with cytology negative for PCP. The patient continued to deteriorate, developing respiratory failure and septic shock. Given the clinical suspicion for PCP, antibiotics were suspended and systemic steroids were initiated with slow clinical improvement. Subsequent lab analysis showed elevated β-D-glucan and sputum PCR analysis was positive for PCP. After subsequent clinical stabilization and laboratory confirmation, trimethoprim-sulfamethoxazole was again initiated, with progressive clinical improvement and pulmonary pathology recovery.

Discussion

PCP is an underappreciated clinical challenge in the immunocompromised population suffering from respiratory symptoms outside of the AIDS population. Studies have shown that healthy individuals can act as a reservoir for Pneumocystis in which the bacteria can still multiply in the alveoli. However, pneumocystis cannot become pathogenic until the immune system becomes compromised, as occurred in this patient and in an ever-increasing patient population. PCP can then become a life threating disease due to its rapid evolution to respiratory failure, as occurs in almost one third of cases. This requires physicians to maintain a high index of suspicion, especially in patients with hematological malignancies, autoimmune disease, organ or bone marrow transplant recipients, those receiving systemic steroids or monoclonal immunotherapy, and those with solid organ tumors. This case exemplifies the importance of persistent clinical consideration of PCP in one such immunocompromised patient, leading to an aggressive workup despite early false-negative results, and subsequent treatment leading to a favorable clinical outcome. An appreciation of this entity would favor patient outcomes in an increasingly more predominant patient population.

References

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