The Diagnosis of Pulmonary Kaposi Sarcoma in a Patient with AIDS

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Introduction

Kaposi sarcoma (KS) is the most common malignancy in patients with HIV/AIDS. Pulmonary KS occurs in 6-32% of patients with active mucocutaneous disease. Diagnosis of pulmonary KS is challenging, as clinical features are nonspecific and may be indistinguishable from pneumonia. Furthermore, pulmonary KS is a significant cause of morbidity in HIV/AIDS patients. We present this case to review the clinical, radiological, and bronchoscopic findings of pulmonary KS.

Learning Objectives

1. Recognize the constellation of findings which suggest the diagnosis of pulmonary Kaposi Sarcoma.
2. Understand the implication of pulmonary Kaposi Sarcoma on overall mortality.

Case Report

History of Present Illness:
A 24 year old male recently diagnosed with HIV started on HAART one week prior to admission presented to the Emergency Department with a three week history of cough, pleuritic chest pain, shortness of breath, abdominal pain, and malaise. Patient also endorsed subjective fever. No recent sick contacts.

Past Medical History:
Patient was diagnosed with HIV three weeks prior to admission. Most recent CD4 7, with HIV viral load 73,169. Started on STRIBILD (elvitegravir, cobicistat, emtricitabine, tenofovir disoproxil fumarate) therapy.

Physical Exam:
T 38.8°C, oxygen saturation 92-98% on room air, BP 110/70, HR 95.

Pertinent findings: violaceous, macular lesions of face and oral cavity, oral candidiasis, bibasilar cracks, and tenderness at the right upper quadrant with a negative Murphy’s sign.

Pertinent Labs:
VBG: 7.41/42/32/27
WBC 4.3
Hgb 12.9
LFTs WNL
Lactic acid 1.6
LDH 249
Blood cultures negative
CD4 count 7
HIV viral load 73,169

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Imaging

Figure 1. Admission Chest X-Ray: Diffuse bilateral patchy opacities, focal consolidation of the left lower lobe, bilateral pleural effusions.

Figure 2. Hospital Day 3 Chest X-Ray: Increase in bilateral perihilar and bibasilar opacities.

Figure 3. Hospital day 5 Chest CT: Disseminated bilateral ill defined patchy, nodular opacities. Worsening bilateral pleural effusions.

Figure 4. Subsequent Admission Chest CT: Disseminated nodules, peribronchial opacities, mediastinal and hilar lymphadenopathy, bilateral pleural effusions.

Figure 5. Bronchoscopy: Diffusely erythematous tracheobronchial mucosa.

Pathology

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Hospital Course

The patient was empirically started on ceftriaxone for community acquired pneumonia. Over the course of admission he had increasing oxygen requirements. Coverage was broadened to include azithromycin, trimethoprim/sulamethxazole (PJP dosing), and prednisone (PJP dosing). Chest CT revealed bilateral airspace consolidation, patchy nodular opacities, bilateral pleural effusions, pulmonary edema, and bilateral axillary lymphadenopathy. Pleural fluid was exudative with negative culture and cytology. Bronchscopy revealed diffusely erythematous tracheobronchial mucosa without discrete lesions. BAL studies for infection and malignancy were negative. Axillary lymph node biopsy revealed HHV8+ spindle-shaped neoplastic cells. EGD/colonoscopy confirmed Kaposi involvement of the gastrointestinal tract. Doxorubicin therapy was initiated with improvement of symptoms. Unfortunately the patient was re-admitted twice for respiratory failure, ARDS and eventually passed away. The cause of death was likely visceral involvement of KS.

Discussion

Patients with pulmonary KS commonly present with subacute dyspnea, cough, and fever. Chest radiograph commonly demonstrates hilar adenopathy, peripheral nodules, and pleural effusions. Classic chest CT findings include interstitial thickening, a bronchovascular distribution of perihilar infiltrates, poorly defined bilateral pulmonary nodules, fissural nodularity, and bilateral pleural effusions. Bronchoscopy adds diagnostic value when Kaposi’s lesions are visualized, however patients may have parenchymal disease without tracheobronchial involvement. BAL studies most often reveal alveolar hemorrhage, and may be helpful in ruling out infection. Trans-bronchial biopsy has low diagnostic yield, given the patchy involvement of lesions. The mainstay of treatment for pulmonary KS is chemotherapy and HAART, however therapy is palliative rather than curative. Patients have brief periods of disease-free remission and an overall poor prognosis. Notably, two-thirds of patients with pulmonary KS have superimposed opportunistic infection, which is the predominant cause of mortality rather than the malignancy itself. Despite the advent of highly active antiretroviral therapy (HAART), pulmonary KS carries a poor prognosis with a median survival time of 1.6 years. Timely recognition of pulmonary KS and initiation of appropriate therapy is paramount in improving mortality from this disease.

References: