Title

Disseminated Kaposi Sarcoma A Cautionary Tale

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Abstract

Introduction: Cytokine dysregulation and immunosuppression are important risk factors in the oncogenesis and proliferation of Kaposi Sarcoma (KS).[1] This is especially relevant in patients with HIV/AIDS who often receive glucocorticoids for treatment of opportunistic infections. Within this patient population, corticosteroids have been associated with KS disease progression and increased morbidity and mortality[2], thereby presenting a diagnostic challenge to clinicians. In this report, we examine the complex presentation and challenges in diagnosis of disseminated KS associated with steroid use and paradoxical KS-associated immune reconstitution inflammatory syndrome.

Case Report: A 33-year-old transgender female with a history of HIV/AIDS presented with chronic, progressive dyspnea and cough. She was admitted to the hospital three weeks prior to this admission due to the unremitting nature of these symptoms. At that time, she had a CD4 count of 80 cells/mm^3, viral load of 260,000 copies, and chest imaging with multifocal, perihilar, bronchocentric opacities (Figure 1). An extensive infectious work-up including tests for tuberculosis and a direct fluorescent antibody (DFA) sputum stain for pneumocystis pneumonia (PCP) was unremarkable. Despite a negative DFA, the patient was empirically treated for PCP with both trimethoprim-sulfamethoxazole and steroids given her persistent hypoxemia. She was started on highly active antiretroviral therapy (HAART) and discharged home. Although she initially experienced mild relief, she ultimately developed worsening dyspnea prompting her return to the hospital. On presentation, she was febrile, tachycardic, and hypoxic. There were numerous well-demarcated, violaceous, raised lesions along her bilateral lower extremities as well as a solitary lesion on the hard palate (Figure 2). Tender bilateral lymphadenopathy (LAD) in the anterior cervical chain and painless bilateral inguinal LAD was noted. Pulmonary auscultation revealed diffuse rhonchi and reduced breath sounds bilaterally. Laboratory blood work was notable for white blood cell count of 14.2 K/uL, c-reactive protein 17 mg/L, and CD4 165 cells/mm^3. Chest computed tomography (CT) demonstrated significant worsening of dense mass-like peribronchovascular consolidations with obliteration of segmental and subsegmental bronchi and bronchioles (Figure 1). Due to the refractory and worsening respiratory symptoms, the diagnosis of PCP was revisited. Given the worsening radiographic findings, lack of improvement with antibiotics, and interval development of multiple violaceous mucocutaneous lesions in the setting of recent steroid administration and initiation of HAART, there was high suspicion for disseminated KS complicated by immune reconstitution inflammatory syndrome (IRIS). A bronchoscopy was performed with visualization of multiple intraluminal macular, erythematous lesions consistent with KS. Core needle biopsy of an inguinal lymph node. confirmed the diagnosis of KS. A diagnosis of disseminated KS with pulmonary involvement exacerbated by steroids and likely complicated by IRIS was established. Given her persistent fever, tachycardia, and dyspnea throughout the hospital course, steroids were discontinued given concerns for further exacerbation of KS and possible KS inflammatory cytokine syndrome (KICS). The patient was started on liposomal doxorubicin and HAART was continued. Her respiratory symptoms slowly improved, and interval chest imaging demonstrated marked improvement of her pulmonary lesions (Figure 1). The patient now reports being able to jog.

Discussion: Here we present a case of disseminated KS accelerated by steroids and complicated by IRIS. Pulmonary involvement of KS portends a poor prognosis and empiric use of steroids with underlying unrecognized KS can lead to rapid disease progression. KS can be further exacerbated by systemic inflammatory responses including IRIS and KICS. This case highlights the importance of close monitoring for the interval development of KS lesions in the setting of both HAART initiation and steroid use in patient's diagnosed with HIV/AIDS. Further studies are warranted to determine the clinical utility of KS screening prior to the initiation of HAART. Improved understanding of the role that complex cytokine dysregulation and immunosuppression play in the precipitation or progression of KS may inform care in these complex patients.