Warning Signs: Kaposi’s Sarcoma in Late HIV Diagnosis

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Abstract

- Kaposi sarcoma (KS) is a vascular neoplasm considered an AIDS-defining illness, with a rate of occurrence of 12% in previously diagnosed patients and is associated with HIV-8.
- A 29-year-old Black male with a past medical history of non-Hodgkin’s lymphoma, Type 2 Diabetes Mellitus, and no preventative care presented to an infectious disease clinic following a positive HIV test. He had papular, hyperpigmented, nongruntic lesions present on his lower extremities for the last six months. Multiple ED visits and unsuccessful treatment of perceived cellulitis and eczema.
- Despite HAART therapy, his lesions continue to ascend to the groin with inguinal lymphadenopathy and extend from the knees to hip flexors. He was diagnosed with KS via biopsy. Refractory to chemotherapy, lesions progressed to his colon and small intestines. After admission for recurrent pleural effusions and atelectasis from pulmonary KS, the patient expired.
- This patient has an unusual disease timeline of KS with rapid dissemination of disease to internal organs leading to pulmonary failure, despite chemotherapy and prompt initiation of HAART treatment. There is no literature known for a patient with a similar timeline despite well-studied nature of his condition, treatment, and epidemiology.

Case Report

- He noted papular, hyperpigmented, nongruntic lesions present on his lower extremities for the last six months (Figures 1, 2).
  - Leukopenia was previously treated for eczema and cellulitis with no improvement and subsequent increase in edema in his lower legs.
  - DVT was ruled out during a previous medical visit.
  - A daily combination of bictegravir 50 mg/emtricitabine 200 mg/tenofovir 25 mg (Biktarvy) and tenofovir alafenamide/emtricitabine (Symtuza) were initiated.
- Despite HAART therapy, skin changes continued to worsen. Kaposi’s sarcoma was confirmed via skin biopsy.
- Chemotherapy was initiated.
- A 1-month later patient developed rectal discomfort, diarrhea and lymphedema with progression to his groin and penis (Figure 3).
- An anal pap smear and biopsy of a nodular lesion at the squamocolumnar junction confirmed visceral Kaposi’s sarcoma.
- Patient developed a pleural effusion and intra-abdominal swelling. Endoscopy showed KS lesions in the distal transverse colon and polyps in the rectum (Figure 4).
- There was increasing concern for Immune Reconstitution Inflammatory Syndrome (KS-IRIS) due to the rapidly worsening lymphadenopathy and bilateral lower extremity pain after initiation of HAART.
- Patient developed increased fluid retention, abdominal distention and dark colored emesis and increased pleural effusion. Colonoscopy was performed with further extension of lesions (Figure 5). The patient was admitted to the ICU for respiratory distress and expired due to acute hypoxic respiratory failure and anoxic brain injury. No autopsy was requested.

Introduction and Methods

Introduction

- Kaposi sarcoma (KS) is a well-known vascular neoplasm that is associated with an infection with human herpes virus 8 (HHV-8), also known as Kaposi sarcoma associated virus (KSHV).
- Discovered in 1872 by Moritz Kaposi, KS has since been divided into four types: classic, endemic, iatrogenic, and AIDS-associated.
- Classic KS is the "idiopathic multiple pigmented sarcoma of the skin," typically found to affect more men than women and is more often in people from the Mediterranean area, as well as in Central and Eastern Europe [1, 8].
- KS is considered an AIDS-defining illness, and AIDS-related KS is one of the most common neoplasms in patients with HIV worldwide. More often seen in men who have sex with men (MSM) than in other HIV risk groups, which prompted investigation into other etiologies [5]. HHV-8 was later identified in biopsy studies and found to be the etiologic cause of all subtypes of KS.
- Detections of HHV-8/KSHV found in peripheral blood mononuclear cells preceded development of KS, which supported the conclusion that HHV-8 was the defining marker behind KS [5].
- Given that HHV-8 is a virus that can avoid cellular defenses and subvert immune system processes through its genetic mechanisms, it is an important consideration for patients in high-risk demographics [3].

Methods

Literature over the past 40 years were reviewed along with case reports, review articles and online resources pertaining to Kaposi’s sarcoma. The goal of the literature review was to investigate the presentation of Kaposi’s sarcoma and its presentation associated with HIV and Castleman disease, and the timeline of disease progression of Immune Reconstitution Inflammatory Syndrome (KS-IRIS).

Discussion

- This is a case of Kaposi’s sarcoma (KS) in a patient with a positive HIV status, presenting initially with cutaneous lesions and progressing to visceral involvement [12, 13]. Due to early diagnosis, treatment of HIV with HAART therapy, as well as with more targeted viral susceptibility in chemotherapy, cases of AIDS-related KS are extremely rare, with an incidence of 1397.11 per 100,000 person years, approximately 1.3% [2, 7, 11].
- There was concern in this case for exacerbation of KS upon initiation of HAART treatment, known as Immune Reconstitution Inflammatory Syndrome (KS-IRIS) [9, 10].
- Castleman Disease was also a concern for this patient due to his HIV status confirmation 6 months after the KS had developed.
- This case identifies the need for the promotion of early and regular HIV testing as well as provider education in identification of HIV associated skin conditions. The USPSTF recommends annual HIV testing for adults ages 15-65, and especially for those at an increased risk of infection [4]. Pre-exposure prophylaxis (PrEP) is recommended for those at high risk of contracting HIV [6].
- This patient satisfied two of highest risk demographics as an African American male and MSM.
- The health care providers who treated the patient for his lesions prior to the diagnosis of KS provided failed therapies.
- The patient did not receive HIV testing until he presented to the ID clinic.
- This patient did not receive prophylactic therapy for HIV.

References

- Levy JA, Ziegler JL. Acquired immunodeficiency syndrome is an opportunistic infection and Kaposi's sarcoma results from secondary herpesvirus infec
- Saleski and Ashwin Sharma, MA, MSIII for advising and supporting the preparation of this project. Thank you to Melissa at Spartanburg Regional for her communication in providing images for the use of this project. Thank you to Saleski and Ashwin who were extremely helpful in providing support and financial assistance for the preparation of this project to be presented today. In addition, we are grateful to the patient LA and his family for their consent and kindness to use the case for educational purposes.

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