Somato Publications

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Case Report

Getting to the HAART of Kaposi Sarcoma

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Abstract

Kaposi sarcoma is a rare disease classified as a malignancy arising from the vascular endothelium. It is due to infection with human herpesvirus 8, and is increasing in prevalence due to an increase in HIV/AIDS patients. We present the case of a 31-year-old african american male that presented to our hospital for leg pain with a diagnosis of epidemic (HIV-associated) kaposi sarcoma. The patient had severe, exophytic, violaceous papules and nodules coalescing into plaques measuring up to 6 cm by 8 cm with ulceration and overlying fibrinous exudate in atypical sites including his thigh, which was the most severely affected region. This case demonstrates a severe presentation of epidemic kaposi sarcoma and highlights the importance of clinical recognition of life threatening systemic illness.

Keywords: Kaposi Sarcoma, Human Immunodeficiency Virus, Acquired Immunodeficiency Syndrome, Highly Active Antiretroviral Therapy

Introduction

Kaposi sarcoma (KS) is a type of spindle cell malignancy that arises from vascular endothelial cells secondary to infection with HHV-8 [1]. It is a rare disease and has an estimated age-standardized incidence rate of 0.3 to 0.9 per 100,000 males in the United States [2]. It usually presents as a vascular tumor and can involve multiple organs. Cutaneous lesions appear as violaceous papules, plaques or nodules and have a predilection for the face, upper body, oral cavity and genitalia [3]. There are four subtypes of KS including: classic, endemic, epidemic (HIV-associated) and iatrogenic [1]. The classic and endemic subtypes are indolent and more prevalent in mediterranean countries and sub-saharan africa, respectively. The epidemic and iatrogenic subtypes are more aggressive and due to an underlying immunodeficiency from infection with HIV or immunosuppressive therapy, especially in

organ transplant recipients [1]. The aggressive subtypes have a mean survival of 18 months if left untreated [3]. With the apparition of HIV, epidemic KS has become more common and is increasing in prevalence in the southern United States [1]. Kaposi Sarcoma is listed as an AIDS-defining illness by the center for disease control and prevention (CDC), although recent literature has found this topic to be more controversial.(Center for Disease Control and Prevention, 2008; Dalla Pria, Pinato, Bracchi, & Bower, 2019; Daly, Fogo, McDonald, & Morris-Jones, 2014) A recent retrospective study found that 7 of 23 cases of HIV-associated KS occurred in patients with CD4 T cell counts >300 cells/mm^3 [4]. Similarly, a recent review found that the risk of developing KS in HIV positive patients with normal CD4 T cell counts was substantially higher than the general population [5]. This distinction is important for HIV positive patients with KS, such as ours, as the classification of HIV without AIDS has differing clinical consequences from a diagnosis of AIDS. We present a case of severe exophytic epidemic KS involving atypical sites in a patient newly diagnosed with HIV.

Case Presentation

A 31-year-old African American homeless male presented to the emergency department with complaints of left lower extremity pain. He was found to have septicemia and was subsequently admitted to the hospital. Despite homeless status the remainder of this social history revealed low-risk behavior including denial of IV drug abuse, and no male sexual encounters. He also had a past medical history only positive for an unknown, untreated seizure disorder, no history of STIs, with no contributing family medical history. According to the patient he was recently diagnosed with HIV, roughly 3 months prior to this hospital admission.

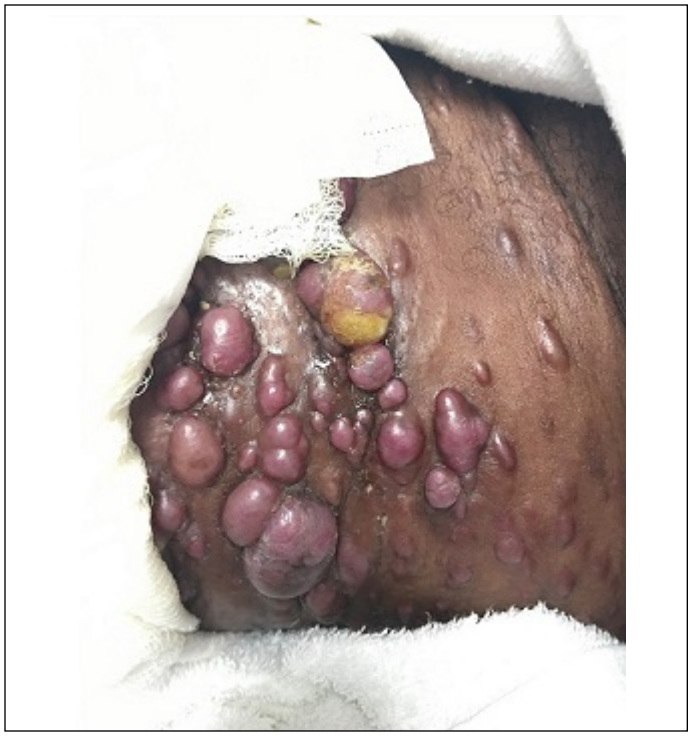

Figure 1: Multiple violaceous papules and nodules measuring up to 0.5 by 0.5 cm that are beginning to coalesce in the inguinal region of the patient.

The pain started five days prior to his admission, was gradual in onset, rated as 8/10, and located in the left calf and hip and had a foul smelling odor. A physical exam revealed a well-developed young male that was alert, oriented, mentally alert and cooperative. The skin examination of the inguinal region showed multiple violaceous papules and nodules measuring roughly 5mm x 5mm (Figure 1). The left lower extremity had extensive lesions that were widespread extending from the groin to the left knee with some involvement of his left lower leg. Numerous nodules and papules were coalescing into exophytic plaques measuring 6cm X 8cm with associated ulceration, necrosis and fibrinous exudate were present along the left leg (Figure 2). The left anterior leg demonstrated similar lesions (Figure 3). The entire left lower extremity demonstrated painful lymphedema.

Further examination revealed sutures present adjacent to several lesions, and the patient reported he had received a biopsy of the area two weeks ago at a local cancer center which had returned with the diagnosis of KS. He reported that he was being closely followed, with plans for chemotherapy and debulking procedures within the next few weeks. He also stated that he was not yet on highly active antiretroviral therapy (HAART), pending his next visit. He was stabilized and discharged over a holiday weekend during which the cancer center was closed. Extensive attempts were then made to obtain records from this facility, but they reported the patient had not been seen at that location for over a decade. Records were then requested from every nearby hospital or cancer center within a 3 hour radius by car, but none confirmed he was a current patient. Attempts were made to contact the patient after his discharge, but they were unsuccessful and the patient was lost to follow up.

Therapy

Therapy for epidemic KS is aimed at the underlying cause.

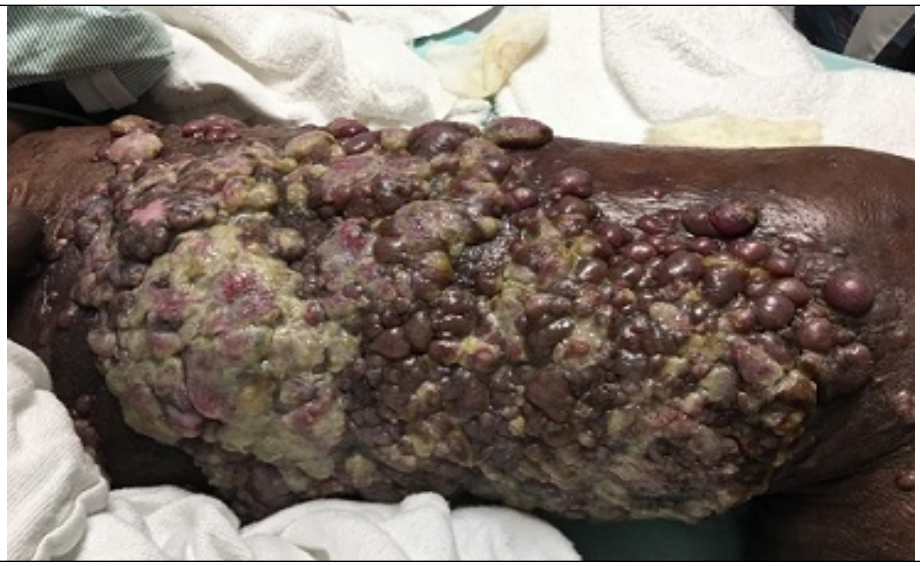

Figure 2: Multiple violaceous papules and nodules coalescing into an exophytic plaque with ulceration and overlying fibrinous exudate on the medial left thigh of the patient

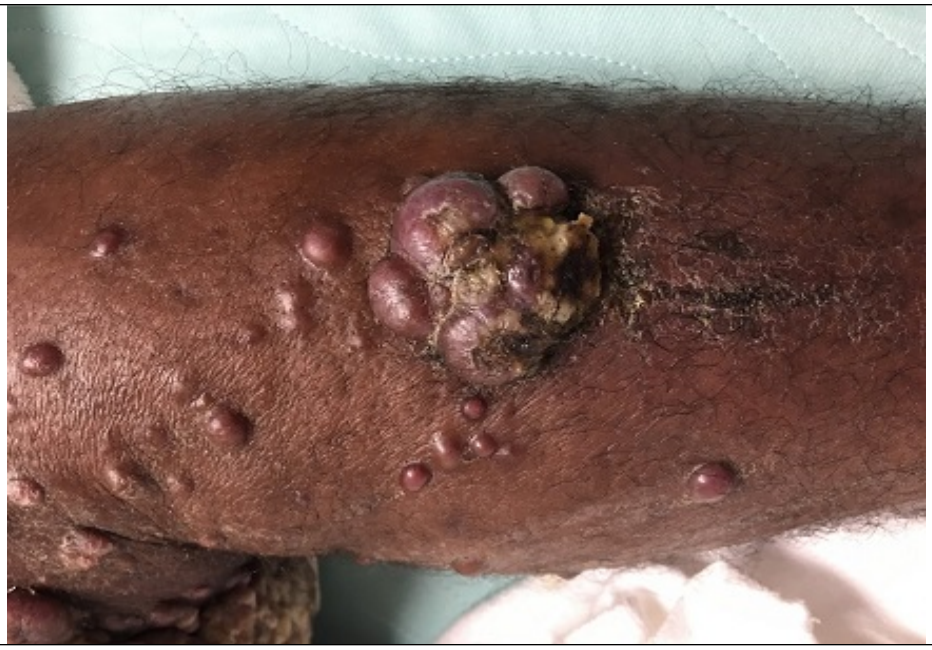

Figure 3: Multiple violaceous papules and nodules present on the left anterior leg of the patient. Some lesions are coalescing into an exophytic plaque with fibrinous exudate and overlying crust.

HAART is the preferred treatment for epidemic KS, as it decreases viral loads and increases CD4+ T cell counts, which leads to prompt improvement in KS [3,6]. In severe cases of epidemic KS, such as our patient, debulking with chemotherapy is useful [6]. The patient stated that he was receiving this debulking therapy and that he would soon be started on HAART. The patient history was confounded by his apparent orientation and mental awareness, but was later found to be confabulating his medical history.

Discussion/Conclusion

Epidemic KS is a severe disease with cutaneous lesions that have a predilection for the face, upper extremities and mucosal sites. It only occasionally present with ulcerative lesions or painful lymphedema [3]. Our case demonstrates the rare presentation of exophytic KS that has associated painful lymphedema with an atypical distribution, as the most significantly and severely affected areas were the lower extremities.

Epidemic KS is an AIDS defining illness that can have significant morbidity and mortality if left untreated [2]. Our patient presented with a history and physical examination clinically suggestive of KS with a recent diagnosis of HIV, though the patient was later discovered to be an unreliable historian in regards to his pathological diagnosis and that healthcare provider that obtained it. Due to the patient's apparent clear understanding of his diagnosis and treatment plan, it was elected to forego obtaining a biopsy and initiating therapy for the patient's KS and HIV/AIDS to avoid interfering with the treatment centers established diagnosis and treatment plan. Our patient was a member of the homeless population, which has a higher incidence of psychiatric illness [7]. A psychiatric consultation may have uncovered an underlying illness that would explain his confabulation. This could have prompted a more serious investigation into the patient's treatment at another facility and would have ensured adequate follow up and treatment prior to his discharge.

While this patient could have benefitted from receiving HAART to treat his underlying immunodeficiency and associated KS, he was lost to follow up and it is unclear if he ever received this treatment for his life threatening diagnoses. Our case highlights the importance of identifying the lesions of KS and their association to an underlying diagnosis of HIV/AIDS, as our patient had clinically apparent KS lesions for a much longer time than the 3 months prior to admission when a positive HIV test result was first obtained.

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