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Photo Vignette

A 29-year-old man with exophytic Kaposi sarcoma and edema of the bilateral legs in the setting of immune reconstitution inflammatory syndrome.

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Abstract

A 29-year-old man with a history of HIV, previously noncompliant with antiretroviral therapy, restarted highly active antiretroviral therapy (HAART) 4 weeks prior to the sudden development of multiple tender exophytic friable tumors and subcutaneous nodules of the thighs. Herein we present a patient with Kaposi sarcoma in the setting of immune reconstitution inflammatory syndrome.

Case Synopsis

A 29-year-old man with a history of HIV, previously noncompliant with antiretroviral therapy, presented with multiple tender exophytic friable tumors and subcutaneous nodules of the thighs (Figure 1 and Figure 2). In addition, he complained of progressive edema of the bilateral lower extremities of approximately 5 months' duration. Of note, the patient had restarted highly active antiretroviral therapy (HAART) 4 weeks prior to the onset of his symptoms. His most recent CD4 count was 156. An excisional biopsy was performed on one of the exophytic lesions.



Figure 1. Multiple vascular papules and nodules on the thigh



Figure 2. Exophytic nodule

A biopsy of one of the exophytic tumors (Figure 3) demonstrated interlacing bundles of spindle cells with poorly formed slit-like vessels. Red blood cell extravasation was present and eosinophilic hyaline globules were seen within the cytoplasm of the spindle cells. Stains for CD34, CD31, and HHV-8 were positive, compatible with a diagnosis of Kaposi Sarcoma. Warthin-Starry, GMS, PAS, and Giemsa stains were negative.

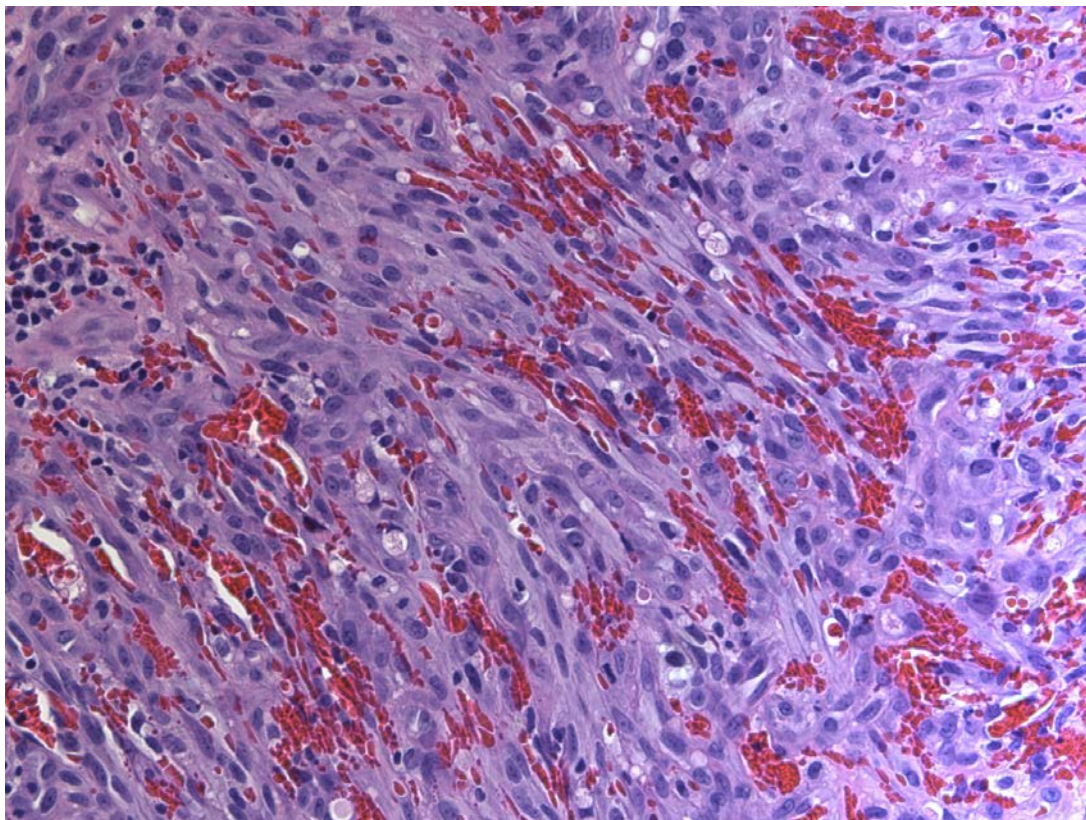


Figure 3. Interlacing spindle cell bundles with slit-like vessels

Despite continued treatment with HAART, the patient's lesions and edema progressed. He eventually required systemic chemotherapy consisting of liposomal doxorubicin 20mg/sq meters every 3 weeks. After 2 doses, he experienced approximately 80% reduction in tumor burden as well as significant improvement in his edema. Of interest, the patient was also found to have a positive rapid plasma reagin (RPR). Subsequent lumbar puncture revealed a diagnosis of neurosyphilis and the patient was treated with a 2-week course of IV penicillin without further complication.

Discussion

Moritz Kaposi, a Hungarian dermatologist, first described Kaposi Sarcoma (KS) in 1872 after he observed an unusual multifocal sarcoma in 5 patients. KS is a spindle cell proliferation derived from virally modified circulating endothelial cells. Human herpesvirus type 8 (HHV-8) has been implicated in all four major variants of the disease: classic KS, African endemic KS, KS in iatrogenically immunosuppressed patients, and AIDS-related epidemic KS [1]. The incidence of Kaposi Sarcoma has declined since the advent of HAART, however, KS has been reported as a rare manifestation of immune reconstitution inflammatory syndrome (IRIS). In a cohort study by Bower et al., patients with IRIS-associated KS were more likely to have tumor-associated edema and higher CD4 counts compared to HIV patients whose KS did not progress while undergoing antiretroviral therapy [2].

Clinically, typical lesions of Kaposi Sarcoma appear as red to violaceous patches, plaques, or nodules. More rare presentations include exophytic, infiltrative, telangiectatic, keloidal, and ecchymotic variants [1]. Classic KS tends to involve the lower extremities symmetrically, whereas AIDS associated KS (AIDS-KS) more often presents with multiple tumors of the head, neck, and trunk. Rapid progression and dissemination are also more frequently observed in AIDS-KS [3,4]. The most common sites of extracutaneous involvement are the lymph nodes, gastrointestinal tract, lungs, and liver [5]. Surprisingly, gut involvement is often asymptomatic in classic KS, but massive, potentially fatal hemorrhage can develop in AIDS-associated and iatrogenic KS.

Diagnosis can be confirmed through skin biopsy; alternatively endoscopic, pleural, or transbronchial biopsy may be used to establish a definitive diagnosis. Therapeutic options vary considerably based on both the clinical course and immune status of the patient. Localized disease may be treated by surgical excision, intralesional therapy, laser, or radiotherapy. Patients with disseminated disease often require systemic chemotherapy; liposomal anthracyclines represent a standard therapy for advanced AIDS-associated KS [1]. Regardless of therapeutic modality, recurrences can occur years after treatment [4]. Similarly to treatment options, prognosis varies considerably with KS. Those with classic disease rarely die of their disease, but AIDS-related generalized KS carries a poor prognosis with a 3-year survival rate close to 0% without therapy [1].

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