Kaposi's Sarcoma of the Lower Extremity: A Case Study and Literature Review

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Statement of Purpose:

The purpose of this case presentation is to present a case of Kaposi's sarcoma of the lower extremity as well as to review the literature regarding this pathology.

Literature Review:

Kaposi's sarcoma (KS) was first described by Moritz Kaposi in 1872 as an "idiopathic multiple pigmented sarcoma of the skin". KS represents about 1% of all diagnosed cancer cases worldwide (1). The multifocal, hyperplastic tumor is believed to originate from lymphatic endothelial cells (2). KS is classified into four clinical variants: classic, endemic, iatrogenic and acquired immune deficiency syndrome (AIDS)- associated (2). Acquired immunodeficiency syndrome associated Kaposi sarcoma typically presents on the face and trunk as pink to red macules and papules and can be rapidly progressive, leading to disseminated visceral involvement and death (3). Although the incidence of KS as an AIDS defining illness has decreased dramatically since the induction of HAART, it remains the most frequent AIDS associated neoplasm and is a common source of morbidity and mortality (3). AIDS associated KS is rarely reported to present on a distal extremity and there are limited published cases in the literature describing this phenomenon on the feet (3). The purpose of this paper is to review a case of AIDs associated-Kaposi's Sarcoma involving the foot, as well as review the literature related to this topic.

Case Report:

44 year old female presented to the emergency department complaining of a non-healing right foot wound. She stated she had this wound on the bottom of her right foot for approximately 2 weeks. She said that the emergency medical service that transported her to the emergency department expressed fluid from her right lower extremity wound. She stated the fluid was "mustard colored" and bloody. She stated she thought the lesion was initiated by stepping on a sharp object; possibly a nail. The patient had been seeing her podiatrist for management of wound for the past month. The wound has been debrided and an unknown cream was being applied to the right leg. She stated a biopsy of a right leg lesion approximately 6 months ago was Kaposi sarcoma. She reported a complicated left leg fracture that got infected and resulted in a below-knee amputation.

The patient's past medical history consisted of HIV for 27 years, lymphedema, post-traumatic stress disorder, depression, hepatitis C, anxiety, polysubstance abuse, documented noncompliance with medication, obesity, and Kaposi's sarcoma. Her past surgical history included a left below-knee amputation. Her medications included alprazolam, methadone, gabapentin, and sulfamethazole/trimethoprim. She was on Atripla but has been switched to Genvoya (Elvitegravir, cobicistat, emtricitabine, and tenofovir-alafenamide) for mental status issues.

Case Report (cont):

On exam, the patient was noted to have a below-knee amputation on the left side. The patient had nonpalpable dorsalis pedis and posterior tibial pulses secondary to edema. The anterior tibial pulse was audible on Doppler exam. Posterior tibial and peroneal pulses were not audible on Doppler. There was significant lymphedema noted to the right lower extremity. There was a lesion at the plantar aspect of the 1st metatarsal head on the right foot. This lesion was exophytic and measured approximately 2 cm x 2 cm and is 1 cm in height from the surrounding skin (Fig 1). The lesion appeared hyper-granular. Purulent drainage was able to be expressed. There was mild peri-lesional erythema with no streaking or ascending lymphangitis. There are multiple flesh-colored papules to the right. She had mild tenderness with palpation to the right lower leg and foot (Fig 2).

Right foot x-rays were negative for any lytic bony lesions or sclerotic changes. Hyperdense diffuse soft tissue swelling was the only positive finding. A right foot MRI was performed to rule out abscess. The MRI was negative for any discrete soft tissue abscess or osteomyelitis however, there was noted to be extensive skin thickening with multiple enhancing nodules and edema. ABI/PVRs were negative for peripheral arterial disease.

She had a viral load of 66 two months prior and her CD4 was 93. Hepatitis C viral load was 4 million. On admission, her CD4 was 188 and her hepatitis C viral load was 8.96 million. She was afebrile with a WBC of 4.1.

The patient was admitted to the hospital. Excisional skin biopsies were performed 3 days later. Two biopsies were taken; plantar right foot and medial ankle. The pathologist reported findings consistent with Kaposi's Sarcoma. (Figs 3-6). A referral was made to oncology for management.

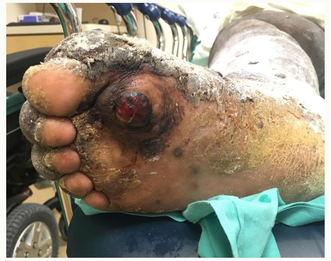

Figure 1: Right foot lesion

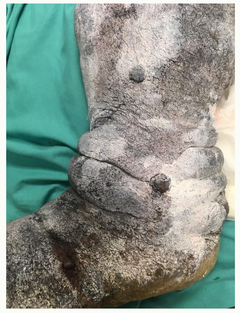

Figure 2: Right lower extremity lesions

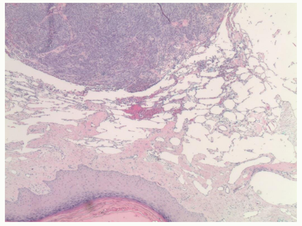

Figure 3: Skin (bottom) and tumor (top)

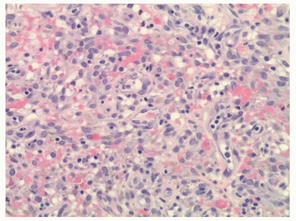

Figure 4: Tumor cells forming slit like spaces with extravasated red cells and hemosiderin

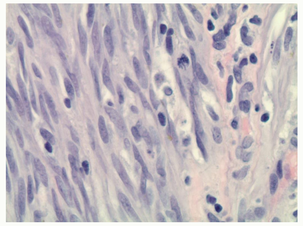

Figure 5: Tumor spindle cells with mitoses and hemosiderin granules

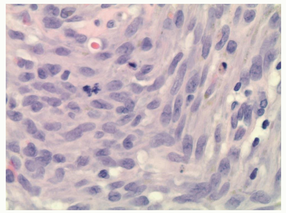

Figure 6: Slit like spaces formed by tumor cells

Discussion:

We report a case of Kaposi's Sarcoma on the plantar right foot in an HIV positive patient. Prior to the AIDS epidemic there were only about 200 cases of KS found for every million people in United States (4). During the AIDS epidemic the rate of KS in this country increased more than 20 times peaking to about 47 cases per million people per year in the early 1990s (4). Because of the new treatments for AIDS, KS has become less common in the United States, and it is now occurring at a rate of about 6 cases per million people per year (4). Human herpevirus 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 (5).

Discussion (cont):

Lesions are typically patches or plaques, which can become nodular, infiltrative, exophytic, lymphadenopathic, echymotic, telangiectactic or keloidal in nature (2). Bilaterally symmetric patches or plaques of the lower extremities, which progress to nodules, are often the initial presentation of early stage disease (2). Initial diagnosis of KS is made clinically, but histological confirmation is essential for an accurate diagnosis (2). Initial disease stage histology typically shows a normal epidermis with the dermis displaying increased spindle cells and vascular structure proliferation within a network of collagen and reticular fibers (2). Polymorphic inflammatory cell infiltrate with prominent plasma cell component can often be seen, along with surrounding proliferation of endothelial-like cells and fibroblasts (2).

Many histological variants of KS have been defined. Grayson and Pantanowitz (2008) have divided these histological variants into four main groups: (a) KS lesions that encompass usual variants related to disease progression (i.e., patch, plaque, and nodular stages); (b) variants alluded to in the older literature, such as anaplastic and telangiectatic KS; (c) more recently described variants, such as hyperkeratotic, keloidal, micronodular,pyogenic granuloma-like, ecchymotic, and intravascular KS; and (d) KS lesions as a consequence of therapy (e.g., regressing KS) (6).

In classic Kaposi sarcoma, treatment must be individualized and includes nonintervention, radiotherapy, surgery, topical imiquimod, cryotherapy, laser, photodynamic therapy and antiviral drugs or, if extensive skin or mucosal lesions are present, liposomal doxorubicin or other chemotherapeutic agents (7). The classic form of Kaposi sarcoma usually has an indolent course and a good prognosis, with few deaths directly attributable to the condition itself (7).

References:

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