Case Report

Pyogenic Granuloma-Like Kaposi's Sarcoma

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Abstract

Kaposi sarcoma (KS) has different clinicopathological presentations, an uncommon form of which is pyogenic granuloma (PG)-like Lesions. This may make its diagnosis challenging, due to its clinical and histological features of both PG and KS. These skin lesions are superficial and protrude outward with subsequent ulceration and secondary infection giving the appearance of a PG. It has been reported in HIV-negative and HIV-positive KS patients. We report a 37-year-old HIV-positive patient on highly active antiretroviral therapy with multiple flesh-coloured skin growths, after initial clinical evaluation, a diagnosis of PG was initially made, however, skin biopsy of the growths revealed KS. We report this case because of rarity of this clinicopathological variant of KS, to which most clinicians misdiagnosed as PG (a benign lesion) with antecedent consequences of delayed treatment. Thus, there is a need for clinicians to do early tissue biopsy of skin growths in HIV/AIDS subjects in order not to miss this rare variant of KS.

Keywords: Highly active antiretroviral therapy, Kaposi's sarcoma, pyogenic granuloma

Introduction

Kaposi's sarcoma (KS) was first described in 1872 by Moritz Kaposi, a Vienna-based Hungarian dermatologist, as a rare multifocal angioproliferative tumor involving blood and lymphatic vessels in elderly men of Jewish origin.[1] It is the most frequent cancer seen in patients with HIV infection.[2] The tumor has four well-defined clinical presentations that have been categorized as classic, endemic (African), transplant-associated or iatrogenic, and acquired immunodeficiency syndrome-associated or epidemic KS.[3]

There are reports of growing number of clinical and/or pathological variants of KS. These variants may make diagnosis difficult with resultant delay and therapeutic implications. Some of the variants may also have prognostic relevance like anaplastic and lymphangioma-like KS.

Pyogenic granuloma (PG)-like KS can exhibit clinical and histologic characteristics of both PG and KS.[4] Clinicians should maintain a high index of suspicion and do an early tissue biopsy for histological diagnosis, so as not to miss this rare variant of KS, especially in HIV-positive patients.

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Case Report

A 37-year-old homemaker diagnosed with HIV Infection 9 months before the presentation was referred to dermatology clinic with an 8 months' history of skin growths located on the thighs, shin of the right leg, foot, and earlobe. Growths on the thighs described as soft sessile and easily traumatized with bleeding and occasional detachment with subsequent recurrence of similar growth on the same site. Other sites reported included the earlobes and the right shin. Another growth on the right shin was described as horn like and hard. She has no associated history of cough, hemoptysis, and shortness of breath, no chest pain; but has easy fatigability. Her baseline CD4 Cells count at HIV diagnosis was 54 cells/ul and was diagnose WHO clinical stage 2. She was immediately commenced on highly active antiretroviral therapy (HAART) (Tenofovir, lamivudine, and efavirenz). No history suggestive of gastrointestinal tract bleeding, no jaundice, and no dysphagia. Has had significant weight loss since onset of illness. Response to HAART has been suboptimal with minimal change in her weight.

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Physical examination revealed a chronically ill-looking woman, who was wasted and pale. Skin examination showed two friable, tender, soft, sessile growths on the lateral and medial aspects of the left and right thighs respectively each measuring about 2 cm × 3 cm. One of which easily detached from underlying hyperpigmented plague with central actively bleeding vessels on detachment [Figure 1]. Similar lesions were also found on the dorsum of the right foot and right earlobe. There were numerous dusky red patches and plaques on the trunk and mucosal surfaces over the hard palate.

Examination of cardiorespiratory, gastrointestinal, and nervous systems essentially normal.

An excision biopsy of the sessile growth together with the underlying plaque for histology showed that; they were composed of proliferating vascular channels containing red cells and aborted binumerous spindle cells arranged in nodules and extend to subcutaneous layer [Figures 2 and 3], histological conclusion of KS was made for both the nodular and wedge biopsy. Immunohistochemistry with latent nuclear antigen-1 (LNA-1), a monoclonal antibody to HHV-8 was not done due limited resources and unavailability in our center.

Following adequate preparation, patient while still on HAART was commenced on KS specific chemotherapy with significant clinical improvement evidence by shrinking, dryness, and falling off of the growths.

Discussion

KS is a low-grade vascular endothelium malignancy that has been categorized into four clinico-epidemiological forms: (a) Classic KS, mostly seen in elderly men of Middle East, Eastern European, and Mediterranean ancestry, (b) The endemic or African KS, (c) The iatrogenic or immunosuppression-related KS, and (d) HIV/AIDS-related KS.[3] The HIV/AIDS-associated KS commonly presents as aggressive and disseminated disease with oral, skin, lymph node, and visceral involvement.

PG-like-KS is a rare clinicopathological variant of Kaposi sarcoma (KS) with clinical and histological features of both PG and Kaposi sarcoma.[4] This condition is characterized by superficially located nodular KS lesions that become protuberant and elicit the development of a peripheral epidermal collarets.[5] Traumatized lesions may undergo ulceration and became inflamed such that may be misdiagnosed as PG. Cases of PG-like KS has been reported in both HIV-positive and HIV-negative individuals.[4] In one report, the PG-like KS was initially thought to be a PG of the nasal mucosa.[6] While another author reported the PG-like KS on the hands of 3 patients, which is a common location for PG.[5] For our patient she is HIV-positive and the PG-like KS lesions were located on the thighs, earlobe, and dorsal aspect of the toes.

PG-like KS case was also reported by Michael Megaly and collique in the United States of America (USA) as a foul-smelling mass between the second and third toes of the right foot of a 30 year old HIV-positive man who has been off HAART, which had been slowly increasing in size and nonresponsive to antibiotics for presumed infection.[7]

Figure 1: A flesh coloured nodular pyogenic granuloma-like swelling on the medial aspect of the right thigh, though painted with gentian violet by the patient

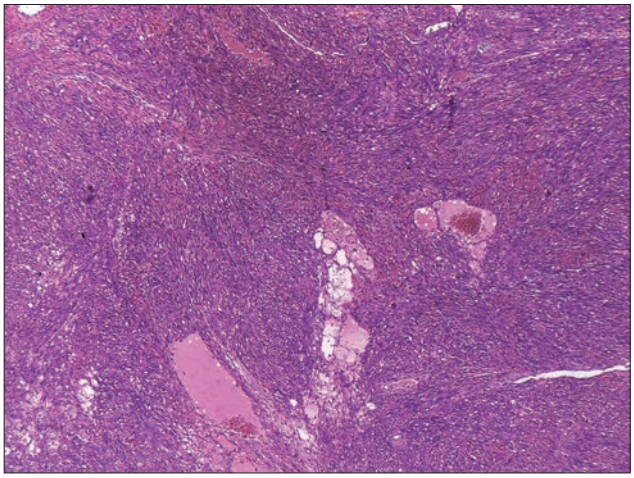

Figure 2: Proliferating vascular channels containing red cells and aborted binumerous spindle cells arranged in nodules (×40 magnification)

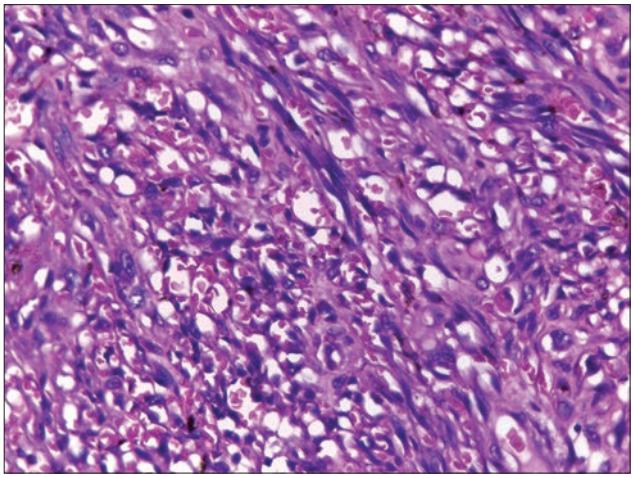

Figure 3: Proliferating vascular channels containing red cells and aborted binumerous spindle cells arranged in nodules and extend to subcutaneous layer. (Higher magnification, ×400)

Fukunaga described a case of PG-like KS that was initially misdiagnosed as PG based on the clinical appearance of the skin lesion, but subsequent biopsy and histology confirmed PG-like KS.[8] Histopathological investigation of the case showed distinctly exophytic growth, nodules, spindle cells, vascular structures, cellular atypia, and mitotic activity.[8] This case resembles clinically misdiagnosed KS. Due to it consists of clinical features of PG and histopathological features of KS.

Possible factor that may have contributed to the development of KS in this patient is severe immunosuppression (CD4 cells count of 57cell/ul) just a month before onset of KS lesions despite immediate commencement of HAART.

There is no officially accepted system for staging KS; however, several classification systems were proposed, namely, AIDS clinical trial group (ACTG) classification[9] and the Mitsuyasu classification.[10] The ACTG system uses three variables: tumor extent (T), immune status (I), and systemic symptoms (S) with each variable classed as good risk (0) or poor risk (1).[9] Another system classified KS into Stages I to IV, namely, Stage I: maculonodular (macules and nodules confined only to the lower extremity); Stage II: infiltrative (plaques with some nodular involvement primarily located in the lower extremities); Stage III: florid (multiple angiomatous plaques and nodules dispersed throughout the lower extremities usually ulcerated); and Stage IV: disseminated disease (similar to Stage III disease but extending beyond the lower extremities).[11]

Using above two classification systems our patient falls into ACTG-T0 (good risk), I1 (poor risk), and S1 (poor risk). When the other classification system is applied she falls into stage IV disease.

Recent observations have led to a new grouping of histologic KS variants: (a) KS variants associated with disease evolution; (b) older literature KS variants; (c) recent KS variants; and (d) KS variants related to therapy outcomes.[12] PG-like KS are considered to be part of the "recent KS variants" group and can be very difficult to distinguish from PG given overlapping histologic features, such as epidermal collaret resulting from nodular prominence, ulceration, inflammation, and lobular proliferation of capillaries.[4]

Immunohistochemical staining with HHV-8 LNA-1 has been shown to exhibit high sensitivity and specificity for diagnosing KS and help to differentiate KS from its mimickers. Cheuk *et al*. studied 50 cases of KS and 53 cases of KS-mimickers, and findings showed that 100% of KS cases were positive for HHV8 LNA-1 while 100% of KS-mimickers were negative.[13] This study showed immunostaining of LNA-1 exhibits high sensitivity and specificity for diagnosis of KS and is useful for distinguishing it from mimickers. The KS-like PG closely resembles KS histologically, and it has been reported that immunohistochemistry in such lesions may be positive for smooth muscle actin and factor VIII-related antigen, which are typically negative in KS.[13] However, due to limited resources and unavailability, immunohistochemical studies in our setting could not be done. For our patient, diagnosis of PG was made based on the clinical appearance of the flesh colored superficially located exophytic nodules, and histology.

Treatment options for localized disease include; Surgical excision, external beam radiation, laser therapy, cryotherapy, photodynamic therapy, topical tretinoin gel, and intralesional vinblastine. Systemic chemotherapy is used for widespread lesions as it is in the index case where intravenous paclitaxel was used. Another treatment modality for AIDS-related KS is immune reconstitution with HAART drugs. The index patient is on combination of tenofovir, lamivudine, and efavirenz which will help with the immune reconstitution.

Conclusion

KS can present as rare and atypical variants, such as lymphangioma-like KS, classic KS with a sarcoid-like granuloma presentation and PG tous-like KS. PG-like KS may be misdiagnosed as PG and as such treatment may be delayed. Clinicians should maintain a high index of suspicion when lesions resemble PG especially in HIV-positive patients and do an early tissue biopsy for histological diagnosis, so as not to miss this rare variant of KS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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