THE MEDICAL BULLETIN OF
SISLI ETFAL HOSPITAL

DOI: 10.14744/SEMB.2023.46034
Med Bull Sisli Etfal Hosp 2024;58(2):241–243

Case Report

Primary Classic Penile Kaposi's Sarcoma in a Middle Age Circumcised HIV-Negative Patient: Presentation of an Unusual Case

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Abstract

Kaposi's sarcoma should be considered in the differential diagnosis in young patients with penile lesions and no risk factors.

A 37-year-old heterosexual man with no other medical history applied presented with a non-itchy and painless penile lesion, for three months. The HIV 1-2 serology was negative via ELISA test. Histopathological analysis of the lesion revealed a tumor composed of atypical spindle cells, below a partially ulcerated surface. There was also an abundance of plasma cells admixed within the neoplastic cells. The patient was diagnosed as HIV-negative, HHV-8 positive Kaposi sarcoma.

Although penile Kaposi sarcoma is extremely rare, classical Kaposi sarcoma should be considered in the differential diagnosis of penile lesions.

Keywords: HHV-8, HIV, Kaposi sarcoma

Please cite this article as "Ergul RB, Ortac M, Tonyali S, Pehlivan G, Ozturk Sari S, Hurdogan O, et al. Primary Classic Penile Kaposi's Sarcoma in a Middle Age Circumcised HIV-Negative Patient: Presentation of an Unusual Case. Med Bull Sisli Etfal Hosp 2024;58(2):241–243".

Kaposi sarcoma (KS) is a multifocal antiproliferative disease that originates in the lining of the lymph or blood vessels. It usually arises on mucocutaneous surfaces, but also in the respiratory and gastrointestinal systems. The lesions are macroscopically red, purple, and black nodules. Human herpesvirus-8 (HHV-8) plays an important role in the pathogenesis of KS. The association of KS and Human Immunodeficiency Virus (HIV) is highlighted in the literature.[1] Penile lesions of KS are extremely rare in HIV-negative and circumcised men, which highlights the importance of the present case report. Few cases of KS with isolated penile lesions among patients without HIV infection have been published in the literature.[2-3]

This study aims to discuss the etiology and clinical presentation of an HIV-negative circumcised man with KS with a penile lesion.

Case Report

A 37-year-old heterosexual man with no other medical history presented with a non-itching and painless penile lesion for three months. Physical examination revealed a non-itching and painless, dark red-blue exophytic nodule,

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Submitted Date: February 03, 2023 **Accepted Date:** April 25, 2023 **Available Online Date:** June 28, 2024

sized 7-8 mm, at the ventral side of the penis, abutting the coronal sulcus (Fig. 1a). There were no mucocutaneous lesions or enlarged inguinal lymph nodes. The patient denied smoking, use of illicit drugs, and penile trauma. The HIV 1-2 serology was negative via ELISA test. Consultation with the dermatology clinic and excision of the nodule for pathological examination were recommended. The lesion was excised under local anesthesia by protecting surgical margin. The skin was sutured primarily. Histopathological analysis of the lesion revealed a tumor composed of atypical spindle cells, below a partially ulcerated surface. There was also an abundance of plasma cells admixed within the neoplastic cells (Fig. 2a). Immunohistochemical analysis for the anti-HHV-8 antibody showed strong nuclear positivity (Fig. 2b). The histopathology was consistent with a classical HHV-8 positive KS. Contrasted thoracic and abdominopelvic computed tomography revealed no pathologic lymph nodes. The patient was diagnosed with HIV-negative Kaposi's sarcoma. No local recurrence was observed three months after surgery (Fig. 1b).

Discussion

KS is classified into four groups according to etiology, including classic, endemic, epidemic, and iatrogenic subtypes. The classical form of KS was originally described by Moritz Kaposi in 1872. The incidence is 0.01 per 100,000 / year for the United Kingdom and 0.2 per 100,000 / year for the United States of America.[4]

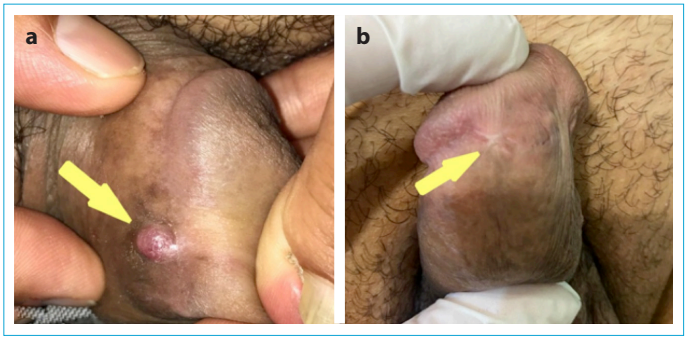

Figure 1. (a) Preoperative image of the penile lesion **(b)** Postoperative image of the penis.

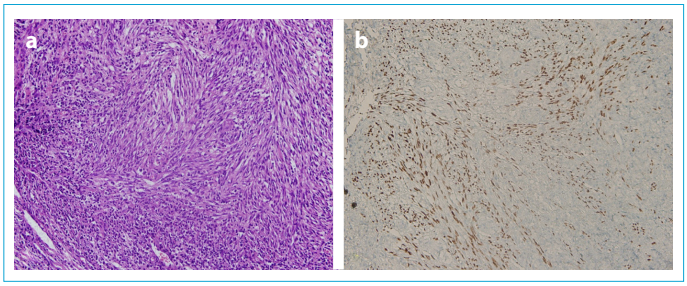

Figure 2. (a) Spindle shaped tumors cells, arranged in a whorled fashion with accompanying abundant plasma cells (Hematoxylin&Eosin, x200) **(b)** Immunohistochemical analysis for anti-HHV-8 antibody shows strong nuclear positivity in spindle tumor cells (anti-HHV-8, x200).

In circumcised men, penile lesions are rare and typically asymptomatic. The diagnosis of penile lesions can be challenging due to the lack of the physicians' familiarity with the lesion and embarrassment of the patient. Hence the accurate diagnosis is generally delayed.[5] KS can present with a solitary penile lesion. According to current literature, the primary occurrence of KS on the penis is rare, and mostly observed in patients with HIV.[6]

The lesions of KS are mostly localized on the lower extremities, as patches, plaque nodular, papular, ulcerated, granulomatous, and verrucous lesions. However, lesions can rarely appear in other areas such as the face, oral mucosa, and genitalia.[7] The penis is one of the rarest presentation sites for classical KS, particularly in circumcised patients. Recently Cito et al.[8] published a case report and review of the literature, and reported that only 33 HIV-negative patients with a penile lesion have been diagnosed as classical KS so far.

Although the etiopathogenesis of KS is not clearly understood, Human Herpes Virus 8 (HHV-8), named Kaposi's sarcoma-associated Herpesvirus, is strongly associated with KS. Almost all patients with KS have HHV-8; while the minority of people with HHV-8 develop KS.[9] Our patient also had positive serology for HHV-8.

Generally, classic KS is more common in elderly male patients. Studies show that the incidence rate in males is 2-3 times higher than in females. In Cito et al.'s review of the literature,[8] they reported that the mean age of the patients with KS was 55.7 years (range 26-78 years). Our patient is 37 years old, had a single partner and was circumcised. This example shows that classical KS should be kept in mind when evaluating young patients with penile lesions and no risk factor.

The appropriate treatment algorithm for KS has not yet been described. Several treatment options, including local excision, laser therapy, topical agents, localized radiation therapy, and chemotherapy, are recommended depending on the stage of the disease.[10] Local excision is usually recommended for small lesions, as in our patient. The patient showed no recurrence or metastasis after three months from the time of surgery.

Kaposi sarcoma of the penis is rare and generally observed in older patients with HIV. However, young patients with a penile lesion and no risk factors can also be diagnosed with classical KS. For this reason, classical Kaposi sarcoma should be considered in the differential diagnosis.

Disclosures

Informed consent: Informed consent was obtained from the patient included in the study.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship Contributions: Concept – R.B.E., M.O.; Design – R.B.E.; Supervision – M.O., A.K.; Fundings – R.B.E., O.H., S.O.S.; Materials – S.T.; Data collection &/ or processing – G.P.; Analysis and/ or interpretation – M.O., S.O.S.; Literature search – S.T., O.H.; Writing – R.B.E., G.P.; Critical review – A.K.

Use of AI for Writing Assistance: None declared.

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