Yeni Üroloji Dergisi - *The New Journal of Urology* **2023;18(3):264-267. doi: 10.33719/yud.2023-18-3-1341287**

Olgu Sunumu / Case Report

An Unusual Presentation of Penile Kaposi's Sarcoma in an HIV-Negative Patient with a Circumcised Penis

HIV Negatif ve Sünnetli Bir Hastada Penil Kaposi Sarkomunun Olağandışı Prezentasyonu

Ayberk Iplikci[1], Ahmet Keles[1], Umit Furkan Somun[1], Fatma Yilmazer[2], Gozde Kir[2], Asif Yildirim[1]

[1] Istanbul Medeniyet University, School of Medicine, Department of Urology Istanbul, Turkey

[2] Istanbul Medeniyet University, School of Medicine, Department of Pathology Istanbul, Turkey

Geliş tarihi (Submitted): *2023-08-12*

Kabul tarihi (Accepted): *2023-08-20*

Yazışma / Correspondence

Ayberk Iplikci, MD

Address: Istanbul Medeniyet University, Department of Urology, Egitim Mahallesi, Dr. Erkin Cd., 34722 Kadıkoy, Istanbul / Turkey

E-mail: ayberkiplikci@gmail.com

ORCID

A.İ.	0000-0002-5822-7799
A.K.	0000-0001-5436-1803
Ü.F.S.	0009-0005-1527-8042
F.Y.	0000-0002-9860-5918
G.K.	0000-0003-1933-9824
A.Y.	0000-0002-3386-971X

Özet

Kaposi sarkomu (KS), esas olarak ekstremitelerde ortaya çıkan multifokal hemorajik bir sarkomdur. Penisle sınırlı KS nadirdir ve genellikle edinilmiş immün yetmezlik sendromu (AIDS) ile ilişkilidir. KS'nin penisteki klinik prezentasyonu ve seyri değişkenlik göstermektedir. Burada glans peniste primer maküler lezyon saptanan KS'nin klasik formu olan 27 yaşında bir erkek hastayı sunuyoruz. Daha ayrıntılı değerlendirmelerde immün baskılama veya hastalığın sistemik tutulumuna dair hiçbir kanıt bulamadık. Cerrahi eksizyon uygulanan hastada nüks olmadı ve takibe alındı.

Anahtar Kelimeler: HHV-8, Kaposi sarkomu, Penil nodül

Abstract

Kaposi sarcoma (KS) is a multifocal hemorrhagic sarcoma that occurs mainly in the extremities. KS limited to the penis is rare and usually associated with acquired immunodeficiency syndrome (AIDS). The clinical presentations and courses of KS in the penis demonstrate variability, with limited reports of non-HIV-related primary KS. Herein, we present the case of a 27-year-old male patient with a classic form of KS who had a primary glans penile macular lesion. In more detailed evaluations, we found no evidence of immunosuppression or systemic involvement of the disease. There was no recurrence in the patient who underwent surgical excision, and he was followed up.

Keywords: HHV-8, Kaposi sarcoma, Penile nodule

How to Cite; Iplikci A, Keles A, Somun UF, Yilmazer F, Kir G, Yildirim A. An Unusual Presentation of Penile Kaposi's Sarcoma in an HIV-Negative Patient with a Circumcised Penis. New J Urol. 2023;18(3):264-267. doi: 10.33719/yud.2023-18-3-1341287

INTRODUCTION

Kaposi sarcoma (KS) is a rare angioproliferative disease of the vascular endothelium. KS is a malignant tumor originating from lymphatic endothelial cells. Its close relationship with Human Herpesvirus 8 (HHV-8) infection was demonstrated in 1994 (1). Classical (sporadic), endemic (usually in seronegative individuals for Human Immunodeficiency Virus (HIV) in Africa), epidemic (associated with AIDS), iatrogenic (iatrogenic immunodeficiency as in organ transplant recipients), and non-epidemic (homosexual, HIV seronegative, non-immunocompromised men) are the five types of KS (2). The lesions are asymptomatic, with brown-red, purple, or blue patches, plaques, and nodules located on the lower extremities, especially the ankle and soles (2). Penile KS usually occurs in HIV-positive patients (3). Herein, we present a rare HIV-negative primary penile KS.

CASE REPORT

A 27-year-old male patient, identified as heterosexual, visited the clinic with a painless, purplish nodular lesion in the vicinity of the urethral meatus on his penis, which he had noticed approximately three months earlier. The patient was sexually active and did not have any suspicious sexual intercourse. He had no known illnesses and was circumcised.

On physical examination, a 5 × 4 mm lesion adjacent to the glans penis area was palpated (Figure 1a). No lymph nodes were observed in the inguinal region. The complete blood count, blood biochemistry, and urinalysis results were normal. Enzyme-linked immunosorbent test (ELISA) serology results were negative for Treponema pallidum and HIV.

The abdominal ultrasonography and chest radiography findings were normal. A complete surgical excisional biopsy of the lesion was performed using boundary control (Figure 1b). Histopathological examination of the biopsy specimen revealed spindle cell proliferation and sieve-like vascular enlargement in the dermis.

Histopathological examination revealed a dermal tumor consisting of extravasated red blood cells and intersecting spindle cell fascicles arranged around slit-like vascular cavities, mixed with scattered inflammatory cells. ETS-related gene (ERG) and HHV 8 expression was observed, and PanCK expression was negative (Figure 2). The surgical margins were negative. The distance of the tumor to the surgical margin was 3 mm at its closest point. The patient was not prescribed a systemic treatment. Following a 3-month follow-up, no recurrence of the disease was observed (Figure 1c).

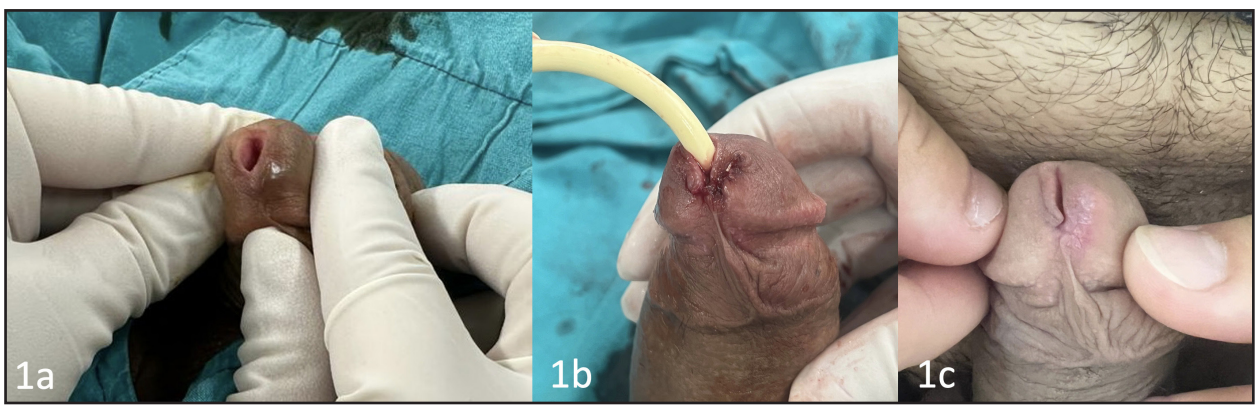

Figure 1a. A purplish macular lesion on the ventral side of the glans penis. **1b.** View immediately after excision of penile lesion. **1c.** Third-month postoperative view.

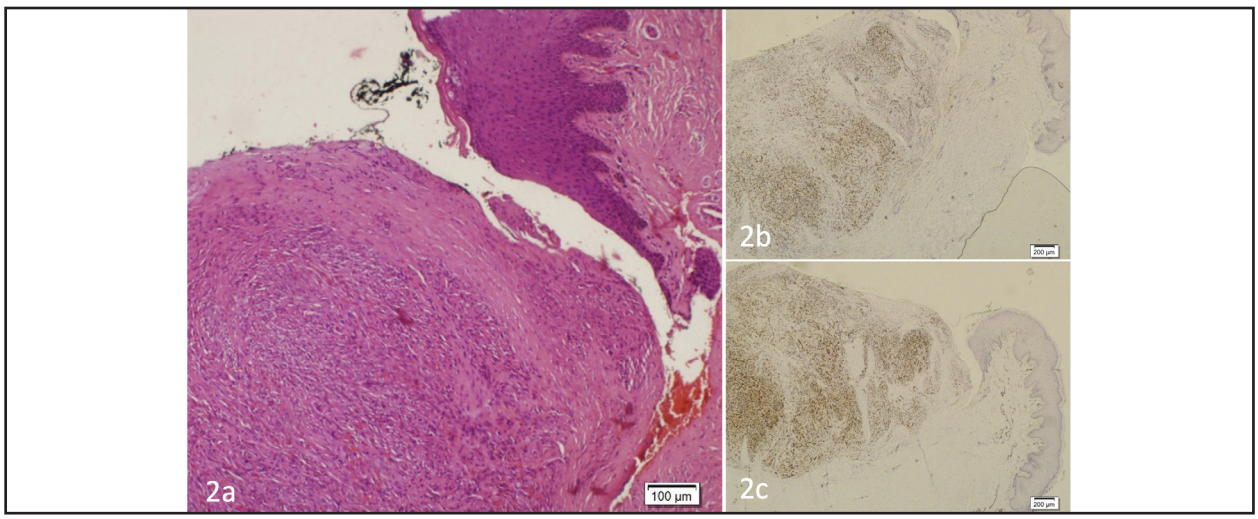

Figure 2a. Spindle cell proliferation with intracytoplasmic lumen containing red cells. **2b.** Neoplastic cells are strongly positive for human herpes virus type 8 latent nuclear antigen 1. **2c.** Positive staining for ERG

DISCUSSION

Kaposi sarcoma was first described by Moritz Kaposi in 1872 and is called 'multiple benign pigmented idiopathic hemorrhagic sarcoma' (4). KS is a multifocal angioproliferative disease originating from endothelial cells (5). The primary symptom is plaque or nodular structures, which appear especially on the skin of the extremities and, to a lesser extent, on other organs. A pathological diagnosis can usually be made using conventional hematoxylin and eosin (H&E) staining. Vascular proliferation in the dermis shows some characteristic features, such as an increase in the number of vessels without endothelial cell coating, the presence of extravasated blood, and the expression of endothelial markers by spindle cells (5). This multicentric angioproliferative disease, which mainly involves the skin, rarely causes mucosal or internal involvement (6). Although genital lesions are seen in 20% of KS cases, only 3% have a primary localized lesion in the glans penis, as in our case (3,6).

Kaposi sarcoma is most commonly associated with Acquired Immunodeficiency Syndrome (AIDS). In the literature review, involvement was the first sign of HIV and AIDS in very few of the patients presenting with KS involving the penile region (7). In the case published by Tammam et al. in 2022, a 35-year-old male patient was admitted to the hospital with an ulcerated penile lesion and systemic findings. After receiving antiviral and antibacterial treatment for a while, the patient who remained without follow-up and had low treatment compliance died in a metastatic state shortly after diagnosis despite surgery (7).

Primary KS of the penis may also occur even more rarely in HIV seronegative patients, as in our case. The first case of solitary penile KS with HHV8 positivity in an HIV seronegative patient was published by Morelli et al. in 2003 (8). Another publication in which cases with primary penile involvement are evaluated belongs to Cito et al. evaluated 33 cases of KS where the penis was the first site of origin. According to epidemiological evidence, there is a strong association between disease pathogenesis and HHV-8 infection. Most patients with penile KS had positive results in serology HHV-8 research. (9). Our case supports the literature in this respect.

Kaposi sarcoma is more common in men, with a reported male/female ratio of 3:1. Few cases have been reported in individuals under 50 (10). Our patient is unusual because of his young age. Primary penile KS clinical course is variable, but local recurrence

is rare. There is no standard treatment method for primary penile KS. In the literature, some cases underwent local surgical excision, radiotherapy, laser treatment, and chemotherapy (3,9). To date, there has been no standardized follow-up. In general, local recurrences are rare if the primary tumor is completely removed (9).

CONCLUSION

Although penile Kaposi sarcoma is a rare condition in HIV-negative men, it should be considered in the differential diagnosis and treatment of nonspecific lesions in the penis. A rare presentation of KS may present as a single lesion on the penis without any known risk factors. Therefore, histological evaluation is recommended for patients with penile lesions. The treatment should be customized according to the clinical and immunological status of the patient.

Financial Disclosure: The authors declared that this study has received no financial support.

Conflict of Interest: The authors declare that they have no conflict of interest.

Informed Consent: Written informed consent was obtained patient who participated in this case.

REFERENCES

1. Chang Y, Cesarman E, Pessin MS, et al. Identification of herpesvirus-like DNA sequences in AIDS-associated Kaposi's sarcoma. Science. 1994;266(5192):1865-1869. https://doi.org/10.1126/science.7997879

2. Vangipuram R, Tyring SK. Epidemiology of Kaposi sarcoma: review and description of the nonepidemic variant. Int J Dermatol. 2019;58(5):538-542. https://doi.org/10.1111/ijd.14080

3. Micali G, Nasca MR, De Pasquale R, Innocenzi D. Primary classic Kaposi's sarcoma of the penis: report of a case and review. J Eur Acad Dermatol Venereol. 2003;17(3):320-323. https://doi.org/10.1046/j.1468-3083.2003.00747.x

4. Braun M. Classics in Oncology. Idiopathic multiple pigmented sarcoma of the skin by Kaposi. CA Cancer J Clin. 1982;32(6):340-347. https://doi.org/10.3322/canjclin.32.6.340

5. Cesarman E, Damania B, Krown SE, Martin J, Bower M, Whitby D. Kaposi sarcoma. Nat Rev Dis Primers. 2019;5(1):9. Published 2019 Jan 31. https://doi.org/10.1038/s41572-019-0060-9

6. Mahzouni P, Taheri D, Goukizadeh A. Primary Kaposi's Sarcoma of penis. J Res Med Sci. 2005;(1):38-39.

7. Tammam A, Abdulrahman A, Ebrahim M, et al. Penile Kaposi Sarcoma as an initial manifestation of HIV infection: A case report and literature review. IDCases. 2022;29:e01576. Published 2022 Jul 19. https://doi.org/10.1016/j.idcr.2022.e01576

8. Morelli L, Pusiol T, Piscioli F, et al. Herpesvirus 8-associated penile Kaposi's sarcoma in an HIV-negative patient: first report of a solitary lesion. Am J Dermatopathol. 2003;25(1):28-31. https://doi.org/10.1097/00000372-200302000-00006

9. Cito G, Di Costanzo R, Morselli S, et al. Primary penile Kaposi's sarcoma in HIV-seronegative patient: a case report and literature review. Int Braz J Urol. 2020;46(5):825-842. https://doi.org/10.1590/S1677-5538.IBJU.2020.05.03

10. Di Lorenzo G. Update on classic Kaposi sarcoma therapy: new look at an old disease. Crit Rev Oncol Hematol. 2008;68(3):242-249. https://doi.org/10.1016/j.critrevonc.2008.06.007