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Inflammation and infection

Classical form of Kaposi sarcoma localized in penis

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ABSTRACT

We present you a case of 43 year old man with classical form of Kaposi sarcoma (KS) localized to the Penis who was HIV negative. Detailed pathological and immunohistochemistry characteristic of the tumor was done. Pathology reported it as KS with nodular and polypoid form. Classical form of KS with localization in male genitalia is rare identity and serves as a diagnostic challenge.

Introduction

Kaposi Sarcoma (KS) was first introduced in 1872 by Moris Kaposi, Hungarian Dermatologist. This tumor was found to be associated with Human Herpes virus 8 (HHV-8) in 1994. There are many forms described in the literature: Classical form, mainly found in adults in Mediterranean region, where frequency of HHV8 infection is very high; Endemic form found in 1990, with aggressive pattern in patients with HIV, and those who underwent organ transplantation (immune-compromised), where both transplanted organ can be a source of HHV-8 or the recipient may be pre-infected. Lesions are usually nodular, red, violet, black colour and may be localized anywhere but primarily engaged to the skin, gastrointestinal tract and the respiratory tract. The growth of the tumor can be either very slow or very fast and aggressive. KS arises from lymphatic endothelium and forms vascular spaces, filled with erythrocytes. Tumor cells are spindle shaped, and the lesion is highly vascularized with irregular blood vessels.

Case presentation

A 43 year old man was admitted to the Clinic of Urology at Medical University Pleven. Patient presented with history of tumor formation in distal half of the penis, which started before one month and grew rapidly [Fig. 1]. There were no other subjective or objective symptoms, past medical history and operative intervention linked with our pathology. Patient is heterosexual, married and the partner is clinically healthy. Both the partners had no history of an extramarital sex. Detailed physical examination showed non tender, exophytic growth, red-blue in colour with a narrow base, 2 cm in diameter on the foreskin of the dorsal surface of the penis. There was no palpable enlargement of inguinal lymph nodes and no other pathological finding of external genitalia or anywhere else in the body. Consultation with internal medicine doctor and anesthesiologist did not rule out any other disease. Pre-operative laboratory investigations were found to be normal. Patient underwent circumcision with complete excision of the tumor formation [Fig. 2]. There was 24 months follow-up, during this period patient was found to be healthy without any signs and symptoms.

Histology

The pathological and immunohistochemistry of the lesion was studied in detail. It was characterized as polyp-like, nodular lesion with focal ulcerations. It consists of spindle shaped cells, forming slit-like vessels grouped in bundles. Erythrocytes were found inside blood vessels with infiltration of perivascular lymphocytes and plasma cells seen as a component of the tumor [Fig. 3]. The tumor cells were positive for CD-34 & D-40 [Fig. 4]. Pathology confirmed it as KS in a HIV negative patient.

Discussion

KS is a multifocal process with vascular origin which may affect skin and internal organs. Around 20% of all cases have lesions in genitals but only 3% of these patients have primary lesion localized to the penis[1]. First case of KS was described in 1902. In our case, it is localized to the prepuce of the penis as primary isolated lesion, which is exclusively rare classical form of KS.

This form of disease is characteristic of Mediterranean region which

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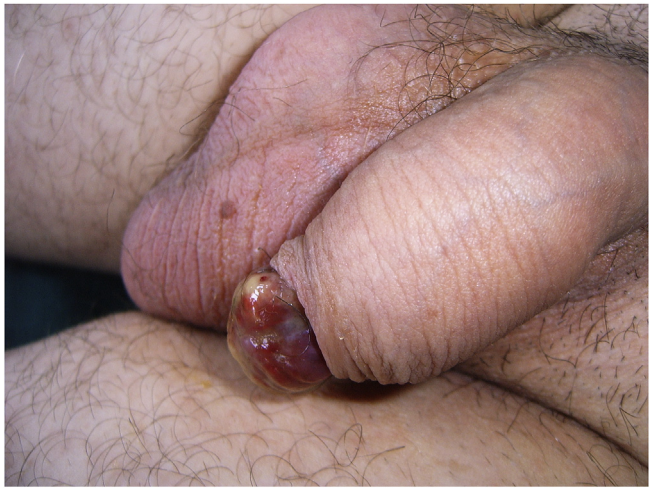

Fig. 1. Tumor formation of the penis.

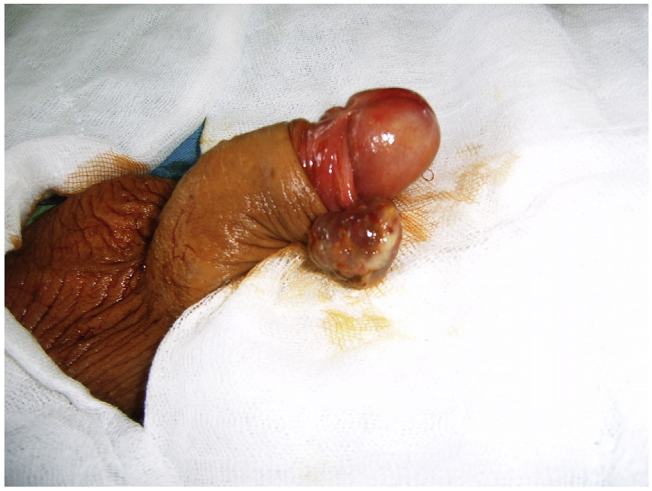

Fig. 2. Excision of the tumor.

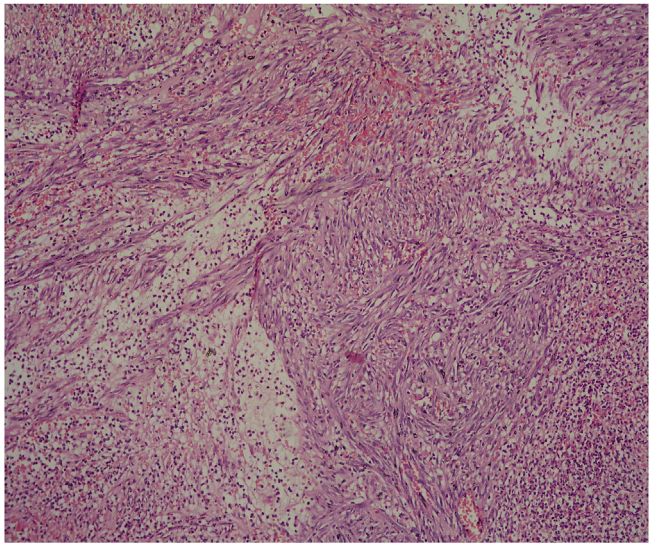

Fig. 3. HE x 25.

frequently affects patients during 6th and 7th decade of their life. Usually lower limbs are affected[2]. It is caused by HHV-8 which is a necessary component but not significant factor for the progress of disease. In our case presence of lesion only in penis suggests sexually transmitted infection. Patient never had extra marital sex with male or female partner. The wife of the patient was not having any skin or mucous lesions and was free from sexually transmitted diseases. In Greece classical form of KS is having frequency of 0.47/100,000 people per year and seen to occur in elderly, lesions disseminating skin, usually accompanied by lymphedema or visceral distribution[3]. The most frequent form of KS is nodular 85.3%[4]. Many patients are asymptomatic – 48.8%, most common complaints are pain - 26.7%, oedema- 21.2%, bleeding – 14.9%, itching – 3.9%[4]. Patients with classical form of KS have higher chances of development of Non-Hodgkin's lymphoma and malignant melanoma[5]. The method of treatments are simple excision, radiotherapy, chemotherapy, cryotherapy, laser and photodynamic therapy. During involvement of internal organs systemic chemotherapy is used.

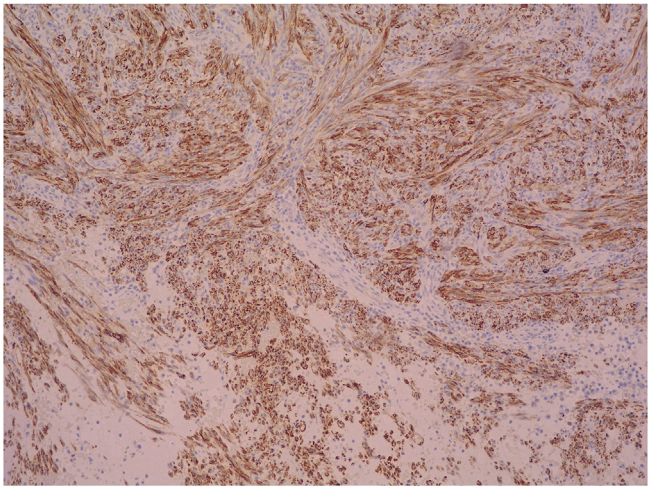

Fig. 4. D2-40 × 25.

Conclusion

Presence of tumor lesion in the genital area should be differentially diagnosed with KS. In doubt, serological test for the presence of HIV must be done.

Conflicts of interest

The authors declare that they have no competing interests.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2019.100856.

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