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CASE REPORT

Primary classic penile Kaposi's sarcoma in an elderly HIV-negative patient: report of a case and review of the literature

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Summary

Kaposi's sarcoma (KS) is a neoplastic vascular tumor. The case of a 91-year-old HIV-negative and HHV-8-positive with two reddish-purple lesions (one pedunculated) on the inner layer of the prepuce is described. Although primary penile KS is a relatively uncommon lesion in HIV-negative men, dermatologists and urologists should consider this possibility when treating non-specific penile lesions.

Key words: classic Kaposi's sarcoma, HIV-negative, human herpesvirus 8 infection, penis, primary involvement

Introduction

Kaposi's sarcoma confined to the penis is extremely rare and is most often observed in AIDS patients, who usually develop an aggressive clinical course [1-4] and in 2-3% of the cases penile KS lesions may be the first manifestation of the disease. In 1993 Swierzewski et al. [5] reviewed 51 cases of KS of the penis and genitalia; among them, 37 cases have been regarded as isolated classical KS without any association with AIDS or immunosuppression [3,6,7]. In 1994 Chang et al. identified DNA sequences of a new herpes virus (HHV-8 or KSHV) in KS patients with AIDS- associated KS [8].

The case we describe herein is interesting for its unusual site of presentation, the oldest age of onset, the association with HHV-8 infection and HIV-negative serology.

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Case presentation

A 91-year-old non-circumcised man presented to the urological department because of the discovery of a lesion on the inner layer of the prepuce. Examination revealed two lesions, one pedunculated, located lateral to the frenulum on the internal sheet of the prepuce. The lesions were 15 mm and 5 mm in diameter, reddish-purple with smooth surface and well-defined margins (Figure 1).

There was no past history of homosexuality, drug abuse or blood transfusion. Examination of the skin and oral cavity, gastroscopy, colonoscopy and

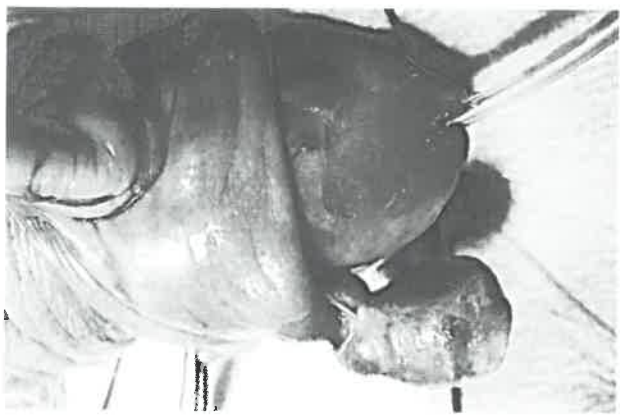


Figure 1. Reddish-purple pedunculated lesion on the inner layer of the prepuce.

computed tomography did not reveal similar lesions elsewhere.

The patient was seronegative on two occasions, 3 months apart, for HIV-1 and HIV-2 in enzyme-linked immunosorbent assays (ELISA).

Excisional biopsy of lesions with a safety zone of 2 mm was performed. Histological examination of both lesions revealed in the mid dermis, a well-circumscribed area composed of slit-like vascular channels, and proliferation of spindle cells in a fascicular or whorl-patterned arrangement (Figure 2). Erythrocytes were present within slit-like and extravasated between the spindle cells. Inflammatory cells (lymphocytes and plasma cells) and dilated vessels were seen at the periphery of the lesion. The epidermis showed focal ulceration (in one lesion). Immunohistochemically the tumor cells expressed a variable positive reaction for antibody to factor VIII-related antigen and CD34. Polymerase chain reaction (PCR) for HHV-8 from a paraffin-embedded skin specimen was positive only in the neoplastic cells. The pathological diagnosis was Kaposi's sarcoma (Figure 1). Ten months postoperatively the patient is disease-free.

Discussion

Kaposi's sarcoma confined to the penis is a rare finding. In the last 20 years most reported cases of penile KS are strongly associated with HIV infection [9]. The first case of KS involving the penis was reported in 1986 by Seftel et al. [1]. Lowe et al. [4] noted that 20% of male AIDS patients with KS eventually developed lesions in their genitalia. KS with penile skin lesion is extremely uncommon in human immunodeficiency virus (HIV)-seronegative patients. Only 12 well-documented cases of primary penile KS in HIV-negative subjects (age range: 43-78; mean age: 57 years) have been reported in the English literature [2,6,10-14]. In our case the patient's age was very advanced compared to the average age of onset of classic KS (6th-7th decade).

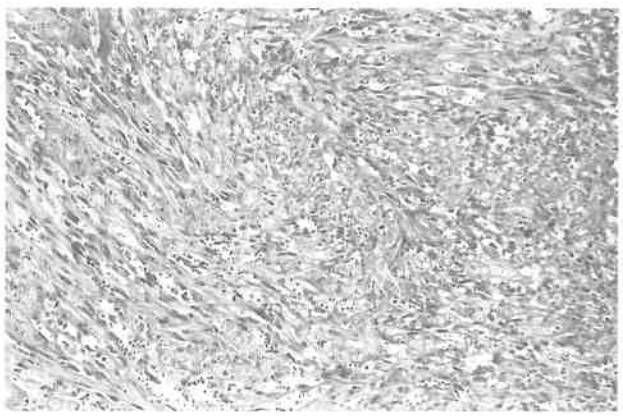


Figure 2. Histology of the biopsy specimen showed dense aggregates of spindle cells, capillaries and extravasated erythrocytes within a circumscribed area in the upper to middle dermis (H&E ×125).

Primary penile KS usually presents as a single reddish-purple to bluish nodule [2,6,10,12]. Multiple papules [2,11], nodules [14,15] or plaques [3] and wart-like or pedunculated lesions, as in our case, are less common presentations [13,16]. Ulcerative nodules, as in our case, have been described in two patients [17]. The majority of cases of KS of the genitalia involve the glans penis [1,13,16]. KS of the prepuce, as in our case, is rare with only three HIV-negative men reported previously [15,16,18]. Lesions may also involve the coronal sulcus [2] and the shaft [3,13].

The clinical course of the disease is variable due to non unified follow-up. In our case the clinical presentation and course where characterized by two non-specific lesions on the penis (foreskin) and no evidence of recurrence after 10 months of follow-up. Recurrence of KS at this site is rare if the primary tumor is adequately removed [3,19]. Onset of new lesions, distant from the primary tumor, may be observed after a period of 2 months-2 years [2,10,11]. Spontaneous regression has been reported by Casado et al. [15] in one case in which the lesions were very small (3 to 7 mm). Penile gangrene with extensive KS in patients with AIDS has been reported by Klein and Lowe [9]. Recent studies show a strong relationship between KS and infection with HHV-8 found in the neoplastic cells and in the blood by serology [20]. It is believed that HHV-8 is necessary but not sufficient to cause the disease, with other predisposing conditions such as gender, immunosuppression and genetic susceptibility playing a major contributory role [7,21].

Most studies suggest that there are at least two possible pathways of transmission: sexual in adults and nonsexual in children probably by saliva [22-24]. In our case the localization of KS on the foreskin and the lack of other manifestations elsewhere suggest sexual transmission.

In conclusion, primary penile KS is a rare disorder in HIV-negative men. Although rare, isolated KS of the penis must be considered in the differential diagnosis of red-purple lesions of the penis. Any association with AIDS should be sought before any treatment.

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