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CASE REPORT

Kaposi's sarcoma: An unusual penile lesion in a HIV negative patient

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Summary ***Kaposi's sarcoma (KS) of the penis is a very rare lesion and it is usually observed in HIV-infected patients. We introduce a case of KS of the penis in a 75 years old HIV negative patient with a peripheral T-cell lymphoma. He came to our attention with a painful ulcerated red lesion on the glans that stretched from the urethral meatus to the coronal skin. This lesion was found to be a KS balanopreputial in the classical variant. Penile KS must be included in the differential diagnosis of genital diseases especially when the clinical features of the lesion are aspecific and diagnosis can be made histologically by performing a biopsy.***

KEY WORDS: Kaposi sarcoma; Penile lesion; HIV negative; HHV-8; T-cell lymphoma.

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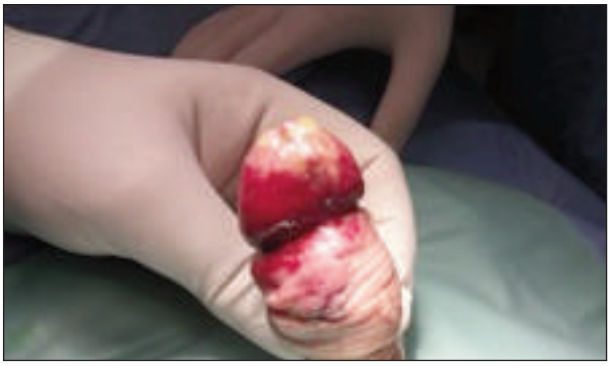
INTRODUCTION

Kaposi's sarcoma (KS) is a malignant neoplasm that typically arises in the skin of the extremities but any area of the face and trunk could be involved. Furthermore it often occurs in the lymph nodes and in the visceral organs (1-3). Penile occurrence is very uncommon so the diagnose and the treatment could be a hard challenge to the urologist. We introduce a very rare case of penile KS in a patient with peripheral T-cell lymphoma without HIV infection.

CASE REPORT

A 75 year-old man came to our attention with a paraphimosis arisen 2 days earlier. He reported a 5 month painless ulcerated red lesion on the glans extending from the urethral meatus to the coronal skin. (Figure 1).
He did not complain urethral discharge nor there were palpable inguinal lymph nodes. He was diabetic, hypertensive and in 2014 it was diagnosed a T-cell lymphoma. So he was treated with COMP 6 cycles, then bendamustin 4 cycles and finally romidepsin 6 cycles with the remission of the disease.
A subtotal circumcision and a glans biopsy were performed. The histological examination could only provide evidence of immunohistochemical staining for HHV-8 both in the stromal cells and in the endothelial ones. There were no localizations of T-cell lymphoma.
Urinary exam and blood laboratory tests were normal. HIV antibody test by ELISA was negative.
The abdominal CT scan showed no inguinal lymph nodes or other suspected lesions.
Due to an extended lesion and to the absence of a clear diagnose it was decided to perform a glansectomy.
The histopathology examination described a classical balanopreputial KS.
Six months later the patient was negative for local recurrence but a secondary gastrointestinal lesion was detected during a whole body CT scan and a gastric biopsy was performed. The patient is currently under therapy with interferon.

Figure 1.
Ulcerated red lesion on the glans extending from the urethral meatus to the coronal skin.



DISCUSSION

KS, described for the first time by *Moritz Kaposi* in 1872, is a rare neoplasm that origins from the endovascular cells in a multifocal way (1).
The behavior of the disease varies from a singular indolent lesion localized in the skin, to a fleeting extensive respiratory and gastrointestinal visceral involvement (2).
Four clinical variants have been described: classic, African endemic, iatrogenic or transplanted associated and HIV-related (3).
Before the 1980s classic KS was rare and it increased because of the AIDS epidemic. Nowadays the introduction of highly active anti-retroviral therapy (HAART) resulted in a reduction of the KS incidence (4, 5).
Primary classical KS of the penis is rare in HIV negative patients (6) and in the literature has been reported only in

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19 cases (7). It usually affects patients between the fifth and eighth decade of life living on the Mediterranean coastal areas where the HHV-8 infection is widespread (6). The pathogenesis of the disease is unclear but HHV-8 infection seems to be an essential cofactor for the development of the KS irrespective of the variant, clinical presentation or prognosis (1, 8).
Beside a KS our patient presented an immunological T cell disorder and an HHV-8 infection.
Probably the T-cell immunity plays a major role in the HHV-8 control and in the development of KS both in viral immunosuppressed patients both in iatrogenic ones.
However no studies have shown a clear correlation between the presence of an immunological T disorder and a KS (9).
The typical clinical feature of a penile KS is a purple papule or macule located on the glans, foreskin, frenulum, urethral meatus or coronal sulcus (6, 10). It rarely affects the shaft or the scrotum. Nodules, plaques, multiple papules, wart-like or pedunculated lesions are less common (5-6).
The differential clinical diagnosis includes pyogenic granuloma, moluscum contagiosum, condiloma acuminate, glomus tumor, bowenoid papulosis, nevi, melanoma and lymphoma (7). The conclusive diagnosis is made by biopsy. The histological pattern is not so different from KS seen in other areas of the body showing groups of spindle cells, extravascular erythrocytes, and macrophages filled with hemosiderin (1). The main target of therapies is to decrease the symptoms, reduce the size, the number of the lesions and to delay the disease progression (11). Many therapeutic approaches have been described: Surgical excision, cryosurgery, radiation therapy, thermo-photoablative, laser therapy, local and systemic chemoterapy, α- and β-interferon (6).
For small and single lesions surgical excision is recommended, while for multiple skin lesions or large-size is recommended radiation therapy. Systemic chemotherapy has been employed in systematic forms (10).
In the case of radical surgical excision, local recurrence is rare (6). However it is important not to neglect the patient's follow-up because a penile unique lesion could be the manifestation of a disseminated Kaposi's sarcoma. As reported by a Greek Author, 82% of patients with classic KS refer a gastric asymptomatic lesion. They recommend therefore the execution of a gastroscopy in every patient with a new diagnose of KS (11-12).

Conclusions

In conclusion the spectrum of penile lesions is broad and the clinical assessment is often insufficient to reach a diagnosis and an adequate choice of treatment. Penile KS must be included in the differential diagnosis of the genital diseases in particular when there are not specific-feature penile lesions so a biopsy is advisable for reaching a histological diagnosis.

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