Scrotal Kaposi's Sarcoma in HIV-negative patient: A case report and review of the literature

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ABSTRACT

Kaposi's Sarcoma (KS) is a malignancy that generally effects the skin, and can be systemic with internal organ involvement. It originates from the vascular endothelium. KS's relationship with human immunodeficiency virus (HIV) infection is well known. Isolated scrotal KS in the urogenital system is quite rare and scrotal KS in an HIV-negative patient is limited to a few cases. In this case report, the biopsy result from the violaceous nodular lesions on the scrotum of the HIV-negative 81-year-old patient was found compatible with KS and a pathology was not detected in the systemic screening. With a diagnosis of isolated scrotal KS, the patient underwent surgical excision aimed at the lesions on the scrotum. KS is rare in HIV-negative patients and it is associated with human herpes virus-8 infection.

Keywords: Human herpes virus type-8; Kaposi's Sarcoma; Scrotal Kaposi.

Introduction

Kaposi's sarcoma (KS) is an angioproliferative disease of the vascular endothelium.^[1] Four subtypes of KS have been categorized. These are classified as classic KS, African type (endemic) KS; transplantation-associated KS and human immunodeficiency virus/acquired immune deficiency syndrome (HIV/AIDS)-related KS.^[2] While its most commonly observed location in the urogenital system is penis, KS with a scrotal localization is rare. In this article we aimed to present scrotal KS in the HIV-negative patient and its surgical treatment with reference to the literature.

Case presentation

The 81-year-old male patient was admitted to the dermatology clinic with a complaint of bleeding, red-purple painful swellings in the scrotal region presenting for 8 months. On dermatological examination a total of 10-12 intra-scrotal lesions with papular or papulonodular

features with a diameter of 3-10 mm were seen. The livid ones were purplish, and some of them were hemorrhagic. Any pathology other than the scrotal lesions and a lymphadenopathy (LAP) were not detected in the physical examination of the patient. A history of type 2 diabetes mellitus, coronary angiography and coronary stent implantation was elicited. The pathology result of the excisional biopsy obtained with the prediagnoses of angiokeratoma and pyogenic granuloma was CD34 (+) in the immunohistochemical analyses. Besides, the human herpes virus-8 latent nuclear antigen-1 (HHV-8 LNA-1) and nuclear staining was detected which was found compatible with KS. The routine hemogram, biochemistry, hepatitis B and C serology, anti-HIV, thorax and abdomen computer tomography (CT) and upper and lower gastrointestinal tract endoscopic tests results were within normal limits. Because of the presence of intrascrotal growth of the lesions, secondary infection and ulceration (Figure 1), total excision was performed under spinal anesthesia (Figure 2). Administration of interferon alpha-2a was planned for other lesions after surgery (Figure 3).

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Figure 1. Preoperative appearance. Violaceous and ulcerative nodules on scrotum

Figure 2. Intraoperative appearance. Total excision of the lesions and primary closure

Discussion

Kaposi's sarcoma was first presented to the literature by Moritz Kaposi in 1872 under the name idiopathic multiple pigmented sarcoma (known classical type).^[3] The etiopathogenesis of the disease has not been clearly explained. Several factors are re-

Figure 3. Postoperative appearance. Other scrotal lesions

sponsible in the etiopathogenesis of KS and its relationship with HIV infection is well known. Another virus related to KS is the HHV-8 whose concomitancy has been proven by epidemiological studies. HHV-8 contains homologues of cellular genes which stimulate cell proliferation, inflammation and angiogenesis and suppress apoptosis as well.^[4-6] Other factors predisposing to the formation of KS include gender, immunosuppression, cytokine activation and genetic predisposition.^[6] In our patient, the immunohistological examinations that would explain the etiology have revealed HHV-8-positivity.

Kaposi Sarcoma is characterized by few or widespread multifocal, brown-violescent or dark red patches and papules, plaques and/or deep nodular skin lesions. Its classical form is often seen in older male patients of Mediterranean or Ashkenazi descent and it is localized in the mucocutaneous tissues, more commonly affecting the lower extremities and feet with its nodular lesions and presents as a clinical entity rarely showing visceral involvement.^[7]

In the urogenital system, extragenital KS most commonly affects penis.^[1] Scrotal KS is rarely seen. The first isolated scrotal KS was described by Vyas et al.^[8] in the year 1976. Other than our case, HIV-negative KS with only scrotal localization and no other urogenital localization has been reported as 3 case reports in the literature.^[1,9,10] Eight cases of HIV positive or negative scrotal KS (incl. ours) have been reported in the literature.^[1,8-13]

The diagnosis of KS is based on histopathological examination. In a study investigating internal organ involvement, asymptomatic stomach involvement has been calculated as 82% in classic KS. For this reason, authors suggest routine gastrointestinal system screening in patients diagnosed as classic type KS.^[14] Endoscopy is the preferred method of screening and our patient's panendoscopic, radiological, and systemic dermatological examination did not reveal any pathologies.

The main objective of KS treatment is to relieve the symptoms of the disease, reduce the size and number of skin and internal organ

lesions and delay the progression of the disease. There is a wide range of treatment options available after diagnosis and the evaluation of the size, localization and number of lesions were made, ranging from total excision to chemotherapy, cryotherapy, laser ablation, electrocautery, radiotherapy, interlesional or systemic injection of cytotoxic agents and alpha and beta interferon as an adjuvant therapy, photodynamic therapy and photodynamic treatment with imiquimod or nitrogen mustard.^[15] Gümüşay et al.^[10] have administered 30 Gy palliative external radiotherapy to the scrotum of an 80-year-old HIV-negative patient with early stage scrotal KS and reported that the patient's lesions regressed by 90% following treatment. In other case-based treatments mentioned in the literature, small lesions had been usually surgically removed. We planned to treat the recurrent lesions that may develop after the total surgical excision of the lesions in the scrotum with interferon alpha-2a.

In conclusion, scrotal KS is a rare vascular neoplasm that can originate from skin lesions progressing to widespread internal organ involvement. KS is rarely seen in HIV-negative patients and treating the lesions with surgical excision and administering intralesional treatment during follow-up can yield good results in patients presenting a scrotal mass not associated with systemic involvement.

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

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