Primary Kaposi Sarcoma of Penis in HIV Negative Patient

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Abstract
Primary Kaposi sarcoma of penis is very rare. We will introduce a 47 years old male patient referred to our clinic from dermatology service, in this report. The patient suffered from itchy penile papules around coronal region. The lab tests had revealed a negative serology of HIV but tissue PCR was positive for Human Herpesvirus-8 (HHV8). Histological findings were compatible with Kaposi sarcoma. Primary Kaposi sarcoma of penis is rare but could occur in HIV negative patients.

Keywords: Primary; Kaposi sarcoma; Penis


Introduction
Kaposi Sarcoma (KS) is the most common vascular neoplasm. Any skin area could be involved, including the genitilia. Skin lesion types have described as: macula or patch, plaque and nodule [1]. Sarcomas of penis are very uncommon, representing less than 5% of malignant tumors in this area. KS is the most common sarcoma of the penis and the second one is leiomyosarcoma [2]. Primary presentation of KS on penis is not common, but more often observed in AIDs patients, whose lesions are "aggressive form", and only approximately 2-3% cases have shown penile KS lesions as first manifestation of disease [3].

Case Report
The patient was 47 years old male, suffered from itchy penile papules which the first one had started to grow 5 years ago. The first manifestation of his disease had appeared by a violet subcoronal papule (about 5 mm in diameter). During the last three months the lesion had extended rapidly and appeared multiple lesions similar to the first lesion, around the coronal region. He didn't have any history of fever, weight loss, or mucosal involvement. He had unprotected extra marital sex with a female partner but 2 years after initiation of lesions. His wife was free from Sexually Transmitted Diseases (STDs). The patient didn't have a history of chemotherapy, radiotherapy, HIV infection, dermatologic disorders and disease and medications with immune suppression. In his past medical history, there was no report of any kind of disease or surgery, but only he underwent surgical remove of right urethral stone, 7 years ago. On examination, there were papular indurate glandular and subcoronal lesions which some of them were crusted. No inguinal or iliac nodes were palpable and other sites of skin and mucosal surface had not involved by tumor (Figure 1, 2).

Laboratory tests include: Cell Blood Count (CBC), hemoglobin, urine analysis and urine culture were normal. Serology of HIV, HBV, HCV tests were negative in two times but the HHV8 was detected in tissue by PCR method in biopsy sample. Wedge biopsy of the lesions had shown a tumor which was composed of spindle cells, around the blood spaces and ectatic capillaries. Fibrotic connective tissue with hemosiderin deposition and mild infiltration of lymphoplasmocytic inflammatory cells had enveloped spindle cells and blood spaces. Those histological findings confirmed Kaposi form vascular proliferation (Figure 3, 4).

The abdominopelvic ultrasonography had shown renal stone in the middle calix of left kidney with mild fullness and other findings were normal. Computed Topographic scan (CT scan) of thorax, abdomen and pelvic were normal too. Endoscopic evaluation of urinary tract and gastrointestinal tract were normal, without tumoral involvement. After oncologic consult, the patient underwent radiotherapy.

Discussion
Kaposi sarcoma has classified to classic and epidemic forms. Classic forms have often occurred in elderly patients, black equatorial Africans, patients with lymphoma or immune deficiencies. In contrast, epidemic types are related with AIDs [1]. Primary
Kaposi sarcoma of penis often has been seen in HIV infected patients and in patients with competent immune system, it has occurred in elderly patients. Reports of primary KS of penis in HIV patients are very rare in literatures [3].

According to some studies, primary presentation of KS on penis is reddish-purple to bluish nodules. Other types of lesions such as papules, plaques and wart like lesions are less common [4-9]. Our patient had presented primary KS of penis by subcoronal papules. KS is a proliferative disease, has characterized by angiogenesis, endothelial spindle cell growth (KS cells), inflammatory cell infiltration, and edema. These lesions have reflected immune dysregulation characterized by CD8+ T-cell activation, production of Th1 cytokines, and angiogenic factors. This process has induced generalized activation of endothelial cells. No specific therapy is curative. Treatment needs to be individualized, based on the patient’s clinical and immunologic status. Highly Active Anti-Retroviral Treatment (HAART) results in clinical improvement of KS lesions and prolongation of time to treatment failure. Numerous anecdotal reports document KS regression on HAART alone. Anti-KS activity of HAART appears to reflect immune system reconstitution and, to a lesser extent, suppression of HIV replication. HIV protease inhibitors are also potent anti-angiogenic molecules that could affect KS pathogenesis. Localized, cutaneous KS lesions could be treated using radiation, laser, cryotherapy, or intralesional injections of antineoplastic medicines. Cytotoxic chemotherapy has indicated for patients who have not responded to HAART and patients with life-threatening or visceral disease [1]. In some studies, palliative excision of lesions of KS on penis
without chemotherapy has related with recurrence after 6 months [10]. After oncologic consult and evaluation of the patient’s condition, we planned radiotherapy for our patient.

Acknowledgment
None

Conflict of Interest
Authors have no conflict of interest.

Authors’ Contribution
Hossein Karami designed this case report, analysed the data and wrote the paper. Alireza Bagher Tabrizi and Mohammad Yaghoobi contributed to the data entry, literature review and writing-up process.

References