

Case report

Lymphadenopathic kaposi sarcoma in an immunocompetent young patient: a case report



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Key words: Lymphadenopathic, Kaposi sarcoma, immunocompetent

Received: 13/08/2015 - Accepted: 01/12/2015 - Published: 12/09/2017

Abstract

Kaposi's sarcoma (KS) is a vascular lesion that usually originates from several sites in the mid-dermis extending into the dermis. Infection from human herpes virus type 8 (HHV-8) is the mostly associated cause. Several articles reported cases of KS, first in Africa, then worldwide because of its close association with HIV / AIDS. KS may also be due to iatrogenic immunosuppression of chronic steroid use, high level of expression of many cytokines and angiogenic growth factors. It can involve skin, mucous membranes, lymph nodes and viscera. We report a case of a 24-year-old immunocompetent, HIV negative male who presented with indolent lymphadenopathy, after adenectomy and histological and immunohistochemical examination revealed a KS. The patient did not have skin lesions. Refusing any other therapy, our patient still lives healthy. This very rare case shows that KS does not always equal immunodeficiency.

Pan African Medical Journal. 2017;28:20. doi:10.11604/pamj.2017.28.20.7726

This article is available online at: <http://www.panafrican-med-journal.com/content/article/28/20/full/>

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Introduction

Kaposi sarcoma (KS) is a low-grade neoplasm that was first described by Moricz Kaposi in 1872 [1]. It is commonly diagnosed in patients who have acquired immunodeficiency syndrome (AIDS), human herpes simplex virus type 8 infection, or underwent immunosuppression or organ transplantation. Lymphadenopathic KS in an immunocompetent adult has been rarely reported [2, 3], most cases are HIV related and they are mainly cutaneous. We report an exceptional case of KS with cervical node involvement in an immunocompetent young male.

Patient and observation

A 24-year-old Moroccan heterosexual male, working as a street vendor, consulted 3 months ago for an isolated right digastric lymphadenopathy, without dysphonia, dysphagia, dyspnea, or epistaxis. He had no medical history of chronic cough or tuberculosis infection. The patient was single without any sexual activity and reported no drug addiction. A cervical physical examination revealed digastric lymphadenopathy on the right measuring 20x15mm. The left lymph nodes were normal. The rest of the physical examination was unremarkable reporting no peripheral skin lesion. Nasofibroscopy and panendoscopy were normal, the intradermal tuberculin test's reaction measured 3mm, the blood cell count was also normal. A computed tomography was performed and showed no other locations. A cervicotomy was made, allowing surgical excision of the lymphadenopathy. Pathological examination showed a dense spindle cell proliferation. The tumor cells showed typical nuclei and were arranged around vascular slits of different sizes. Extravasation of erythrocytes was observed. Immunohistochemical staining showed intense expression of HHV-8 in tumor cells. The final diagnosis was lymph nodal Kaposi's sarcoma (Figure 1 and Figure 2). Two ELISA tests for HIV were negative. After a multidisciplinary discussion, it was decided to perform a selective right neck dissection on the levels II, III, IV and V, which would be followed by radiotherapy with strict regular monitoring. Although the patient refused the treatment, he is still alive in good condition with no signs of local recurrence or other symptoms after 18 months.

Discussion

Described in 1872 as an idiopathic multiple pigmented sarcoma, Kaposi's sarcoma is a rare angioproliferative disease [2]. Four clinical subtypes are recognized: classic (sporadic, mainly in elderly patients of European, Jewish, and Mediterranean descent), African (endemic, mainly in Sub-Saharan Africa), iatrogenic (immunosuppression-associated, principally renal transplant-associated), and epidemic (AIDS-related) [4]. Lymphadenopathic KS also called African or endemic KS is common in different parts of Africa and particularly prevalent among young Bantu children of South Africa, who present with localized or generalized lymphadenopathy. The disease in these patients is extremely aggressive [5]. The etiology is unknown but infection from human herpes virus type 8 has been suggested. Majority of the KS cases are HIV related and they are mainly cutaneous. Very few cases have visceral involvement. Those cases involve mainly the gastrointestinal tract and are diagnosed by endoscopic and histological studies [3]. Pulmonary and lymphadenopathy forms are rare and every time less frequently encountered [3]. Lymphadenopathic KS in an immunocompetent adult is a rare occurrence in the post-HIV/AIDS era. KS associated herpes-virus reactivation is necessary for the development of KS. First reports of KS came from Africa, and only later from all around the world due to the close association with HIV/AIDS. Prior to this however, KS was very frequent in Eastern Europe, Italy and the United States where it existed in an indolent form in the elderly men of Jewish ancestry [3]. KS may also be due to iatrogenic immune suppression from chronic use of steroids, elevated degree of expression of numerous cytokines and angiogenic growth factors including TNF alpha, IL-6, bFGF, HIV tat protein and oncostatin M [3]. KS is usually linked with HIV. It is more common in males than in females, having a sex ratio of 15:1 with predilection for elderly males [6]. KS in Morocco is predominantly a male disease which exhibits some special characteristics, including disseminated skin disease at diagnosis, a more common visceral or lymph node involvement and a less frequent association with second malignancies. In the large series reported by Errhani et al. in the same department where our patient was diagnosed, only 4 cases of lymph node involvement were recorded, all of them showing skin and visceral lesions as well [7].

Our case is unique because it occurred in a young immunocompetent patient with only lymphadenopathy and no skin

involvement. Histologically, the spindle cell variety is the slowest growing while the anaplastic is described as the most aggressive. The mixed cell has an intermediate growth rate [8]. Typically they consist of a proliferation of spindle and endothelial cells, extravasations of red blood cells, haemosiderin-laden macrophages, and, in early cases, an inflammatory cell infiltrate [9]. Kaposi's sarcoma management depends on the type, the extent/dissemination of lesions, and the organs involved. Local treatment modalities are surgical and laser excision, cryotherapy, radiotherapy, intralesional vinblastine or vincristine injections, and topical application of vinca alkaloids, bleomycin or retinoids. Systemic therapy includes highly active antiretroviral therapy (HAART), IFN-alpha for minimal and/or indolent cutaneous disease, and chemotherapy (Paclitaxel, ABV (doxorubicin, bleomycin, vincristine), vincristine, bleomycin, liposomal anthracyclines) for rapidly progressive visceral disease [4].

Conclusion

This case demonstrates that Kaposi's sarcoma can be observed in an immunocompetent patient with only lymph node involvement, and in the absence of cutaneous involvement, and that even in the era of AIDS, KS is not a synonym of immunodeficiency.

Competing interests

The authors declare no competing interests.

Authors' contributions

All authors have read and agreed to the final version of this manuscript and have equally contributed to its content and to the management of the case.

Figures

Figure 1: The lymph node is entirely erased and replaced by a sarcomatous proliferation formed by spindle cells surrounding irregular vascular spaces

Figure 2: The nuclear immunostaining for HHV8 shows spindle cells surrounding the vascular spaces

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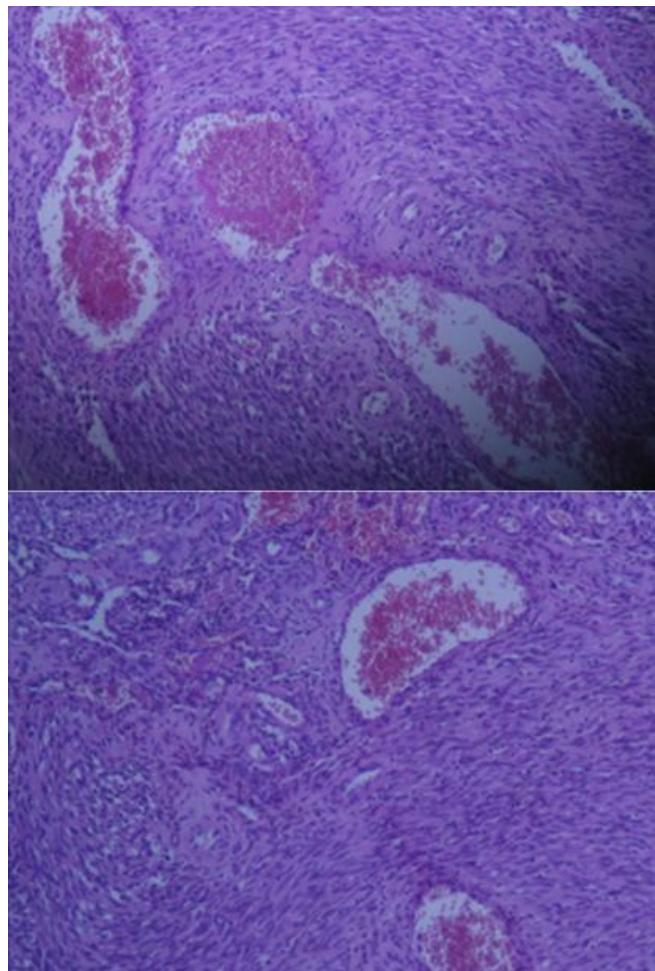


Figure 1: The lymph node is entirely erased and replaced by a sarcomatous proliferation formed by spindle cells surrounding irregular vascular spaces

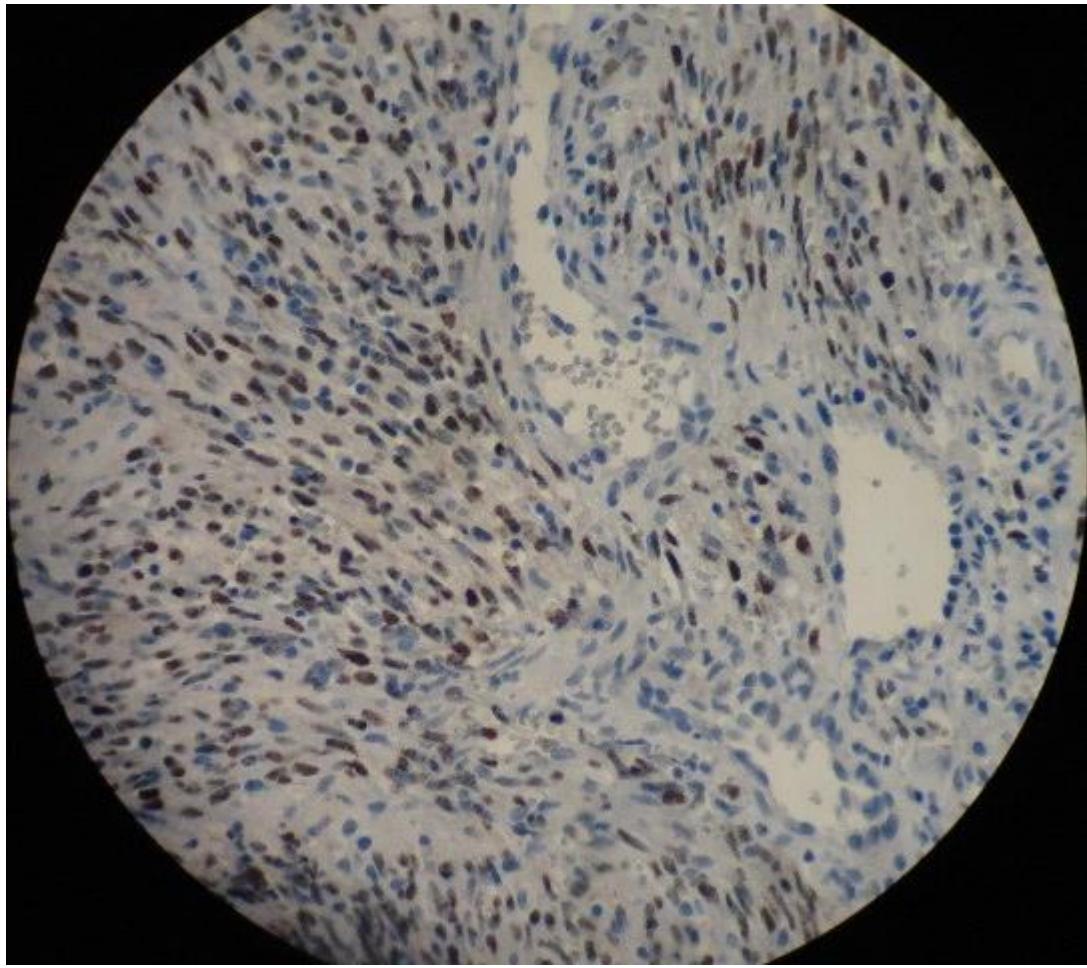


Figure 2: The nuclear immunostaining for HHV8 shows spindle cells surrounding the vascular spaces