Case Report

Unusual oropharyngeal Kaposi's sarcoma about a case not associated to the human immunodeficiency virus

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Abstract

Kaposi's sarcoma (KS) is almost always associated with acquired immunodeficiency syndrome but is rarely described in the HIV-negative and nonimmunosuppressed individual, especially in the head-and-neck region. We present a case of 32-year-old HIV-negative, immunocompetent man with an oropharyngeal KS. The complete surgical resection of the tumor alone was used as a therapeutic modality. Clinical presentation, histological features, and treatment are discussed and compared with a review of the literature.

Keywords: HIV negative, Kaposi's sarcoma, oropharyngeal, treatment

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INTRODUCTION

Kaposi's sarcoma (KS) is a low-grade vascular tumor. Moritz Kaposi, a Hungarian dermatologist from the University of Vienna, was the first to describe this condition as "idiopathic multiple pigmented sarcoma of the skin."[1] This tumor affects more the skin and the mucous membrane than other tissues of the human body.[2,3] Many parts of the body can be invaded: stomach, lungs, intestine.[2,3] The mucous membrane of the oral cavity is the part of the head-and-neck region where the KS is usually found but other parts can be involved such as the nasal cavity, tonsillar mucous, pharynx, and larynx.[4,5]

The prevalence of KS is high in immunocompromised patients, mainly patients with acquired immunodeficiency syndrome (AIDS) or immunosuppression related to the organ-transplant process, but it is rarely seen in the HIV-negative and immunocompetent individuals. According to statistics, KS is more frequently found in men than women. Oropharyngeal primary KS of the nonimmunocompromised patients is very rare and most of the time is clinically misdiagnosed as a pyogenic granuloma, hemangioma, or other benign vascular tumors.[2,3] The pathogenesis of Kaposi's sarcoma in immunocompetent patients is not yet well-known.[5]

Here, we present an endemic KS originated from left soft palate extending to the posterior wall of the uvula of an immunocompetent Congolese treated by surgery and demonstrate its histopathological findings and the treatment outcome.

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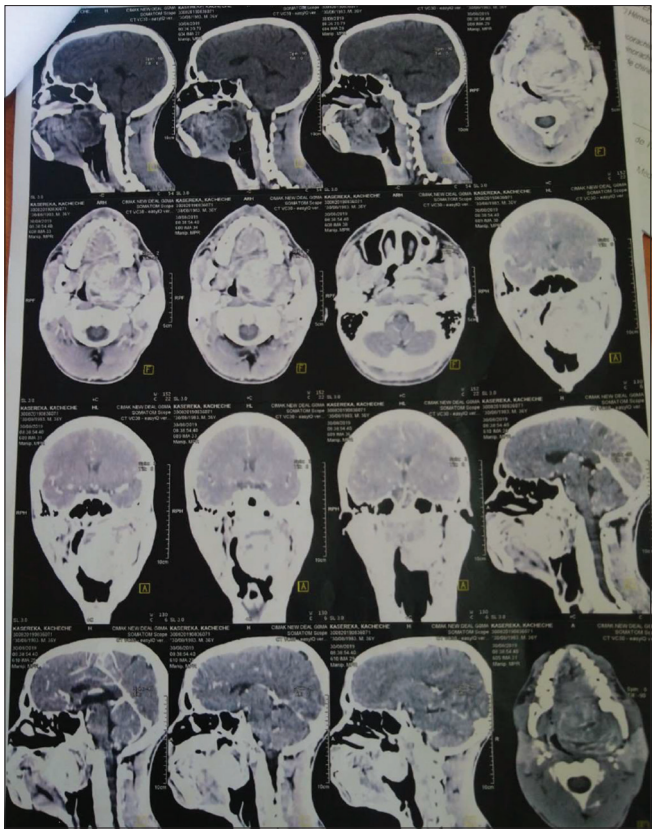

Figure 1: Computed tomography scan showing the mass over left supratonsillar fossa

CLINICAL PRESENTATION

A 32-year plumber was admitted in otorhinolaryngology outpatient department at North-Kivu Provincial Hospital (regional referral and teaching hospital) with 2 years' history of the painful throat, hoarse voice, difficulty in swallowing and in breathing. There is no history of cough. One year after the onset of painful throat, he developed a voice change which became progressively worse. Three months later, he started getting difficulty in swallowing first to solids and after to liquid foods. Six months before admission, he had a sensation of a mass in his throat reason why he often wants to clear his throat. He also noted a bulging in his throat. At the same time, he started experiencing difficulty in breathing progressively worsening, especially at night. There is no history of intermittent night sweats and fever. He noted an unquantifiable weight loss. He had a normal appetite and no abdominal pains were noted. On examination, he was alert with a fair nutritional status. He was afebrile, not pale, and no jaundice. No finger clubbing was noted. All vitals are in the normal range. He had a voice change "hot potato voice" type and was often clearing his throat. The uvula was deviated at left and we noted a bulging of the soft palate which made it not possible to visualize the posterior

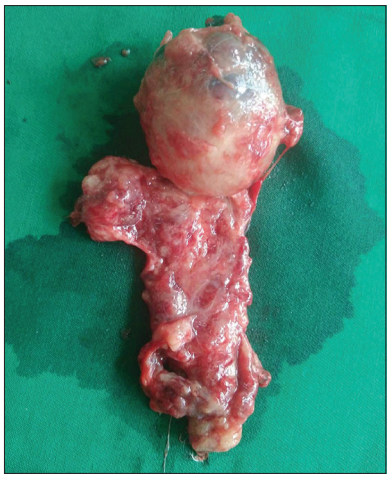

Figure 2: Kaposi's sarcoma removed after surgical excision

wall of the pharynx. No cervical lymphadenopathy was palpated. The cardiovascular and abdominal examinations were unremarkable. Physical examination revealed a reddish-to-purplish, non-tender, and firm lesion over left supratonsillar fossa, sized approximately 6 cm × 4 cm. According to his history of present illness and physical findings, the initial impression of this lesion was a benign lesion, favorably a hemangioma. Computed tomography scan was done and confirmed the presence of the mass [Figure 1]. Tracheostomy was done before we perform the complete transoral resection of the tumor under general anesthesia [Figure 2]. Moderate bleeding was encountered during the surgical procedure. The pathologist report described a "vasoformative tumor composed of hyperchromatic spindle-shaped cells (SC), vascular slits, and extravasated red blood cells. The spindle cells showed only mild nuclear atypia. Immunohistochemical stain showed positive for human herpesvirus 8 (HHV-8) that confirms the diagnosis of KS with no margin involvement." Serological tests for HIV and Epstein–Barr virus were both negative. The patient remained free of disease after 1 year of follow-up.

DISCUSSION

KS, in many studies, has been described as a vascular disorder characterized by proliferation of spindle-SC, the formation of neovessels, inflammation, and edema. Because of the absence of all features of malignancy, it is considered as an intermediate neoplasm. According to many published papers, it has been discovered that KS is linked to immunosuppressive factors such as AIDS, chemotherapy, diabetes, posttransplantation.[2-4] It is well known that KS is one of the major opportunistic diseases which frequently occur in HIV-positive patients where its reported incidence in the head-and-neck region was

found to be as high as 40%–67% with predomination of cutaneous lesions. However, the incidence of KS in patients with HIV-negative was only 14% in the head-and-neck region. Chang *et al.* established the relation of KS with the infectious agent. They demonstrated herpesvirus-like DNA sequences in AIDS-associated KS tissue.[6] Other published papers mentioned that KS-associated herpesvirus (also known as HHV-8) could explain the occurrence of the neoplasm by disorganizing the control of cellular proliferation, secreting chemokines which may induce the vessel formation suppress the immune type 1 helper-T-cell responses.[5,6] As said previously, although KS in the oral cavity is commonly found in HIV-positive patients, it is considered to be rare in non-HIV-infected individuals[2,3,7]

Classically, KS appears as red, purple, or violaceous macules, papules, or plaques; later, they can become nodules or exophytic and sometimes ulcerative lesions. Lesions are usually located to multiple parts of the skin but also lymph nodes and viscera can be involved. Oral cavity lesions ulcerate more often than skin lesions. Because of the ulceration, lesions within the mouth present with pain, burning, and bleeding. Neoplastic-looking forms lesions have also been described in the literature.[2,8] The pathogenesis of KS is now recognized as multifactorial. It is associated with immunosuppression state and genetic and environmental factors have been discovered to influence its pathogenesis. The association with HHV-8 and HHV-8 DNA sequences has been established according to results from many studies where 95% of KS lesions are found in patients with both AIDS and non-AIDS KS. However, transmission modalities of HHV-8 are still unknown, but the incidence of KS among HIV-positive homosexual males was found to be high, this pleads in favor of possible sexual transmission (through feces). Most of all HIV patients with KS present oral cavity and rectal lesions, which suggests local direct spreading. There is a limited number of papers focused on the incidence of KS in AIDS-negative patients in the head-and-neck region.[6,8]

KS has main clinical presentations: classic (sporadic), African (endemic), iatrogenic (immunosuppression associated), and AIDS associated (epidemic). The epidemic type is most focused on scientific publications. Nevertheless, all clinical forms present similar histopathological features in spite of different clinical course and prognosis. All clinical forms present similar histopathological features even if their clinical course and prognosis are different.[4,5] A lot of mononuclear inflammatory cells including mast cells are often discovered with scattered erythrocytes and hemosiderin deposits. The perivascular proliferation of spindle cells may be noted but atypical cells are minimal. Other more advanced lesions present as nodules and plenty of small capillaries or dilated vascular channels with proliferating sheets of sarcomatous or atypia of spindle cells, most of the time with many large erythrocytes and abundant hemosiderin deposition.[5]

About 60% of KS in AIDS-negative patients are situated on the skin (lower and upper limbs or trunk), and the head-and-neck area is rarely involved. The most affected part of the head-and-neck region remains the oral cavity; the oropharyngeal and the conjunctiva mucosa have been found to be involved.[3,8]

The etiology of KS up now remains unclear. However, several epidemiologic and environmental factors and the immunosuppression seem to play a role in the development and clinical course of the condition.[3,7]

The main treatment modalities of KS include:

1. Systemic treatments (chemotherapy and biological, with the use of interferon) for its immunomodulating and angiogenetic properties
2. Local treatments (surgical excision; radiotherapy) indicated for selected and small lesions.

There is still no consensus on an optimal regimen of treatment for KS in the literature, but systemic therapy is often used in disseminated, progressive, or symptomatic KS. Surgical excision and radiotherapy are reserved for local lesions.[5,9]

Our case was treated with surgery only in the absence of radiotherapy, and there is no evident local recurrence and no further lesions observed after a 1-year follow-up. In the light of research, it has been advocated that new advances in understanding the pathogenesis of KS, especially the role of vessel formation and growth factors, may help in the future the development of new therapeutic modalities and allow the establishment of the standardized protocol.[5,7,9] Clinicians should be aware that classic KS might be one of the rare occurrences of vascular lesions from the oral cavity. The complete surgical resection alone in the absence of adjuvant radiation therapy provided adequate tumor control for this patient, so we suggest that this therapeutic should be considered as an option of treatment in low-income countries where radiation therapy is not easily available such as the Democratic Republic of Congo.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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