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Tonsillar Kaposi Sarcoma in a HIV-Negative Patient: A Case Report and Review of the Literature

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Abstract

Tonsillar Kaposi's sarcoma (KS) is an extremely rare malignancy associated with HIV-positive individuals and usually affects the skin. The most common causative agent is the human herpes virus type 8. Through the literature, there are few case reports about KS with no association with HIV. This case report describes a unique presentation of a 40-year-old healthy male, HIV-negative, managed successfully with tonsillectomy alone and was histopathologically diagnosed as unilateral tonsillar KS.

Keywords

Case Report, Kaposi Sarcoma, Tonsillar Mass, Tonsillectomy

1. Introduction

Kaposi sarcoma (KS) is a multifocal angioproliferative malignancy that primarily affects the skin, mucous membranes, and internal organs [1]. First described by the Hungarian dermatologist Moritz Kaposi in 1872, Kaposi sarcoma has since garnered significant attention due to its association with immunosuppression, particularly in the context of HIV/AIDS [2]. In the head and neck region, Kaposi sarcoma is most often seen on the mucosa of the oral cavity; however, cases involving the mucosa of the pharynx, larynx, and nasal cavity have also been identified. Oral Kaposi sarcoma (OKS) is most often seen on the hard and the soft palates, the gingiva, and the dorsum of the tongue [3]. Isolated involvement of the

tonsils is rarely seen in Kaposi sarcoma [4]. Isolated KS involving the tonsil in an HIV-negative and immunocompetent individual is exceptionally rare, with only a few cases reported in the literature.

2. Case Presentation

We report a case of a 40-year-old medically free man who presented to the ENT clinic with a four-month history of progressive sore throat with no associated dysphagia or airway symptoms. On examination, the patient appeared well and spoke normally, and upon oral exam, a right fungated, painless, ulcerative mass on the right tonsil was observed with no bleeding; the left tonsil was of normal appearance. Fiberoptic laryngoscopy was unremarkable.

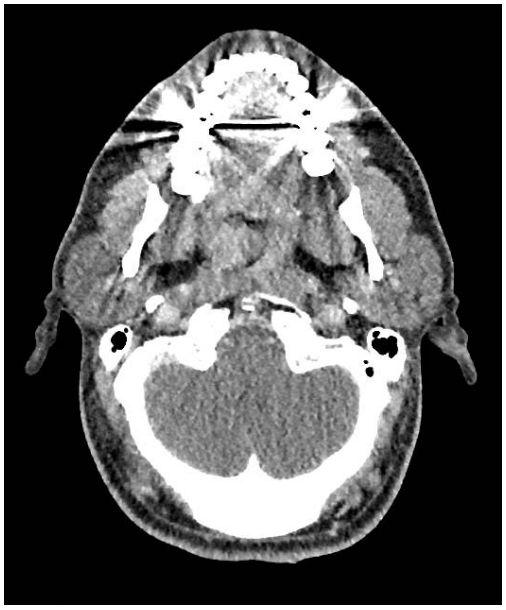
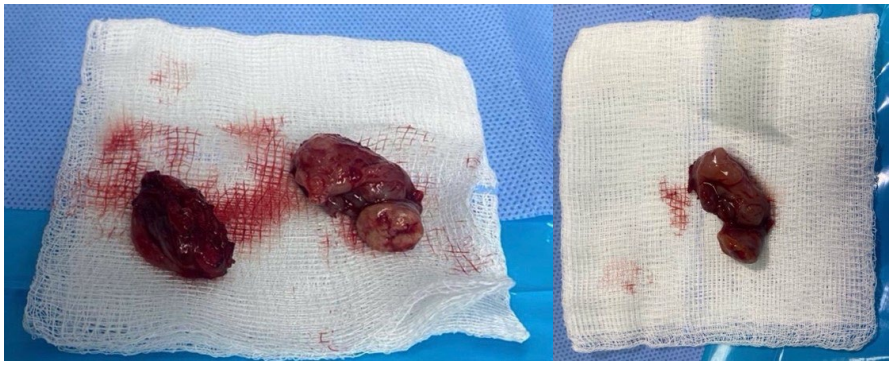


Figure 1. Contrast-enhanced CT imaging of the head and neck revealed a right tonsillar mass about 2 cm * 2 cm.



(A) (B)

Figure 2. (A) Excisional biopsy with bilateral tonsillectomy; (B) Right tonsil.

On the neck exam, there were no palpable lymph nodes. Laboratory investiga-

tions were unremarkable, including an HIV test and complete blood count. Contrast-enhanced CT imaging of the head and neck revealed a right tonsillar mass about 2 cm by 2 cm with heterogeneous enhancement (**Figure 1**). Contrast-enhanced CT imaging of the chest, abdomen, and pelvis was unremarkable. Excisional biopsy with bilateral tonsillectomy was performed without complication (**Figure 1**, **Figure 2**), and histopathological examination confirmed the diagnosis of Kaposi sarcoma, as shown with serial sections of the right tonsil revealing an ulcerated surface squamous cell layer with underlying hypercellular spindle cell proliferation arranged in alternating fascicles and admixed with extravasated RBCs. The tumor cells merge with the blood vessels. The tumor cells showed a mild to moderate degree of nuclear atypia with frequent mitotic activity of about 10/10 HPF. Immunohistochemical examinations identified CD31, CD34, and HHP-8. No evidence of tumor cell necrosis. The background showed reactive follicular lymphoid hyperplasia. The surgical margins are tumor-free. Histopathological examination of the left tonsil showed reactive follicular lymphoid hyperplasia and mild chronic inflammatory infiltrate. The surgical margins are tumor-free. Postoperatively, the patient showed no recurrence or systemic disease for two years of follow-up. Informed consent was obtained from the patient for this report.

3. Discussion and Literature Review

The presented case of Kaposi sarcoma (KS) involving a fungated ulcerative mass on the right tonsil in an HIV-negative individual highlights the importance of considering Kaposi sarcoma in the differential diagnosis of oral lesions, even in immunocompetent patients. While Kaposi sarcoma is commonly associated with immunosuppression, particularly in the context of HIV/AIDS, it can also occur in individuals with intact immune function, albeit less frequently.

In this case, the absence of dysphagia or airway symptoms initially led to the consideration of benign etiologies for the tonsillar mass. Contrast-enhanced CT imaging reveals heterogeneous enhancement of the lesion, which may be variable.

Finding Kaposi sarcoma in HIV-positive but does not exclude the diagnosis. Kaposi sarcoma lesions can demonstrate variable enhancement patterns in imaging studies, and the presence of enhancement does not rule out the possibility of Kaposi sarcoma [5] [6].

A definitive diagnosis is made by biopsy. The differential diagnosis of oral Kaposi sarcoma includes capillary angiofibroma, pyogenic granuloma, lymphoma, oral candidiasis, atrophic candidiasis, and other benign lesions [7].

In this case, histopathological examination of the biopsy specimen confirmed the diagnosis of Kaposi sarcoma, characterized by spindle-shaped cells, vascular proliferation, and extravasated erythrocytes. The absence of HIV infection in this patient underscores the multifactorial etiology of Kaposi sarcoma and suggests that other factors, such as genetic predisposition or chronic inflammation, may contribute to its pathogenesis in immunocompetent individuals.

The management of oral Kaposi sarcoma in HIV-negative patients is similar to

that in HIV-positive individuals and depends on various factors, including the extent of disease involvement and the presence of comorbidities. Ozbudak *et al.* report a case of a 68-year-old male. Diagnosed with tonsillar-oesophageal KS managed with radio/chemotherapy [8]. Keleş E *et al.* also present a case of a 72-year-old HIV-negative female with tonsillar Kaposi sarcoma who was managed with tonsillectomy followed by chemotherapeutic agents with no recurrence in the first year follow-up [9]. Latini *et al.* demonstrate a 37-year-old male HIV-negative also diagnosed with tonsillar KS treated with tonsillectomy alone with no recurrence for 2 years of follow-up [10]. Sikora AG *et al.* described two cases of 34- and 49-year-old males. HIV negative diagnosed with KS treated with tonsillectomy with no recurrence in 3 years of following up [4]. Surgical excision with bilateral tonsillectomy was performed in this case as a definitive treatment approach. While localized lesions may benefit from surgical resection or radiation therapy, systemic chemotherapy may be indicated for more extensive or refractory disease [8].

4. Conclusion

This case highlights an unusual presentation of Kaposi sarcoma as a fungated ulcerative mass on the tonsil in an HIV-negative individual. Clinicians should maintain a high index of suspicion for Kaposi sarcoma in patients presenting with atypical oral lesions, regardless of their HIV status. Prompt biopsy and histopathological examination are crucial for accurate diagnosis and appropriate management of this aggressive malignancy. Further research is warranted to elucidate the underlying mechanisms of oral Kaposi sarcoma pathogenesis and to explore novel therapeutic strategies for this challenging disease entity.

Patient Consent

The patient was consented.

Ethics Committee Approval

Our research gets approved by an ethical committee. The name of ethics committee approval is: Asser Institutional Review Board, the national registration number with NCBE-KACST, KSA: (H-06-B-091), IRB Log No: REC 3-7-2024.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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