Oropharyngeal Kaposi’s Sarcoma from an Immunocompetent Host: A Case Report

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Abstract

Kaposi’s sarcoma is a frequently seen AIDS-related malignant neoplasm in the head and neck region, especially in the oral cavity, but is rarely described in the HIV-negative and immunocompetent individual. We present the iatrogenic Kaposi’s sarcoma affecting soft palate which developed in a 53-year-old male and discuss their histological features and differential diagnosis.

Keywords: Kaposi’s sarcoma; Oropharyngeal; Chemokines

Introduction

Kaposi’s sarcoma (KS) was first described by dermatologist Moritz Kaposi more than a century ago [1]. It is a low-grade vascular tumor and its pathogenesis is closely related to human herpesvirus VIII (HHV-8) [2]. KS has high prevalence in immunocompromised patients, mainly in those with acquired immunodeficiency syndrome (AIDS) or transplant-associated immunosuppression.

KS is more commonly seen in men than women and have four clinical variants such as classic (Mediterranean), endemic (African), posttransplant, and epidemic or AIDS-associated [2,3]. Regardless of the different clinical course and prognosis, all four variants of KS share similar histopathological features and HHV-8 DNA can be found in most cases [3].

Primary KS in oral cavity of an immunocompetent patient (i.e. classic KS) is extremely rare and thus may be misdiagnosed as hemangioma, pyogenic granuloma or other benign vascular lesions clinically. We report classic KS originated from right soft palate of an immunocompetent Taiwanese and demonstrate it histopathological findings and the treatment outcome.

Case Report

A 53-year-old male was referred to our otolaryngology outpatient department due to a soft palatal mass lesion found from local clinic incidentally. He denied recent fever and had no underlying history of diabetes mellitus, hypertension, denied routinely taking any medication or oral supplement. The patient mentioned about oropharyngeal wound in the childhood. The family members of him do not have history of HHV-8 infection.

Physical examination revealed a reddish-to-purplish, lobulated and firm lesion over right supratonsillar fossa, sized approximately 2×2 cm (Figure 1). No cervical lymphadenopathy was palpated. According to his history of present illness and physical findings, the initial impression of this lesion was a benign lesion, favorably a hemangioma.

Figure 1: A reddish-to-purplish, lobulated and firm lesion located on the right side of soft palate. Arrowhead: uvula, Arrow: right tonsil

Transoral en bloc resection of this tumor was performed under general anesthesia was attempted and significant bleeding of the tumor was encountered during the operation. The postoperative recovery was smooth without oral wound bleeding. Histologic examination showed an ill-defined tumor composed of spindle cells separated by slit-like vascular spaces. The spindle cells showed only mild nuclear atypia (Figure 2). No cervical lymphadenopathy was palpated. According to his history of present illness and physical findings, the initial impression of this lesion was a benign lesion, favorably a hemangioma.

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In our case, the sporadic occurrence is categorized in patients with AIDS are called AIDS-associated KS. It is considered rare in non-HIV-infected individuals. Then subsequent studies suggested that the infection of Kaposi's sarcoma–associated herpesvirus (KSHV, also known as HHV-8), results from an opportunistic infection in immunocompromised individuals. This lesion presents similar to KS on clinical examination, because both are multifocal, dome-shaped to polyoid, and red-to-bluish lesions. The differentiation between BA and KS is the circumscription, the presence of epithelioid endothelial cells lining the vessels, the lack of a spindle cell component, and the identification of the causative bacilli on silver stains (Warthin-Starry) in BA. Pyogenic granuloma is a rapidly growing pink-to-red and soft nodules on the skin, or gingival or oral mucosa; it bleeds easily and is often ulcerated. It is considered a reactive lesion that sometimes may be self-limited. However, in the histological analysis, pyogenic granuloma does not have a spindle cell component and eosinophilic globules. Oral hemangiomas may show more dilated vascular spaces, resembling a cavernous hemangioma. Microscopic examination easily distinguished of hemangioma from KS, because the latter present atypical endothelial and spindle cells, eosinophilic globules, and a conspicuous component of reactive plasma cells.

The clinical differential diagnosis of KS includes bacillary angiomatosis (BA), pyogenic granuloma, and hemangiomas. BA is caused by infection with gram-negative bacilli of the Bartonella family, and HHV-8 infection also occurs later in childhood and during adolescence, and through some form of nonsexual contact. For this patient, HHV-8 infection could be through oropharyngeal wound in the childhood. Classic KS can be treated with surgical resection or one of the local treatments, such as radiation therapy. Chemotherapy may be used for widespread skin lesions or for KS that is in the lymph nodes, the lungs, or the digestive tract. Our case was treated with surgery and radical therapy, and there is no evident local recurrence and no further lesions observed after one-year follow-up.

This is one of the few cases of classic KS in ethnic Chinese. Clinicians should be aware that classic KS might be one of the rare occurrences of vascular lesions from the oral cavity. The definitive surgical resection and adjuvant radiation therapy provided adequate tumor control for this patient.

**References**


