

Case report

A rare case of laryngeal Kaposi's sarcoma

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Abstract

Kaposi's sarcoma (KS) is the most common malignancy observed in patient with acquired immune deficiency syndrome (AIDS). It rarely causes upper airway obstruction. We report a 39-year-old gentleman, a former intravenous drug user with AIDS and Hepatitis C positive who developed progressive hoarseness with stidor. He underwent an emergency tracheostomy and direct laryngoscopy revealed a whitish globular laryngeal mass obscuring the glottic region. A biopsy of the mass was taken and the histopathological report showed evidence of spindle cell connective tissue, consistent with Kaposi's sarcoma. It is important for clinicians or surgeons to maintain a high index of suspicion for the diagnosis of laryngeal KS in immunodeficiency patient even without cutaneous manifestation.

Keywords: Airway obstruction, Laryngeal, Kaposi's sarcoma, HIV.

Introduction

Kaposi's sarcoma (KS) is a rare subcutaneous lesion linked mainly with patients suffering from AIDS. In HIV infection, Kaposi Sarcoma is an AIDS-defining disease. It is usually an indolent vascular tumour with some variance depending on the epidemiologic subtype, of which there are four: Classical type, African-endemic KS, Iatrogenic KS and Epidemic AIDS-related KS.^{1,2,3}

The presentation of AIDS patient with KS as laryngeal emergency is rare and only a few cases being reported in the literature.^{1,4} It was first described in 1872 by the Hungarian physician Moritz Kaposi. The characteristic feature of this lesion is multifocal violaceous nodules with the predilection for the skin of the lower extremities. KS may involve every tissue in the human body. Incidence of KS has been reported as high as 20% in

homosexual men who have HIV, 3% in heterosexual intravenous drug users, 3% in women and children, 3% in transfusion recipients and 1% in hemophiliacs.⁵ Mochloulis G et al in his 10 years retrospective study showed the commonest site of laryngeal involvement was the supraglottis (65%).⁶

Case report

A 39-year-old gentleman with AIDS and Hepatitis C presented with 4 month history of progressive hoarseness and stridor. He was a former intravenous drug user with a history of sexual promiscuity. Fiberoptic laryngoscopy revealed a large, whitish globular mass occupying about two-thirds of the laryngeal inlet, obscuring the glottic region (Fig 1). There were no cutaneous manifestations of the disease. The emergency tracheostomy was performed under local anesthesia as patient had

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impending upper airway obstruction. Direct laryngoscopy revealed the above mass, as well as oesophageal candidiasis. A biopsy of the mass was taken and the histopathological report showed evidence of spindle cell connective tissue, consistent with Kaposi's sarcoma (Fig 2). Subsequent staining and cultures for tuberculosis and fungal organisms were negative. He has not encountered any significant complications and was evaluated for radiation therapy. He was discharged home with the tracheostomy and plans for outpatient follow-up visits.

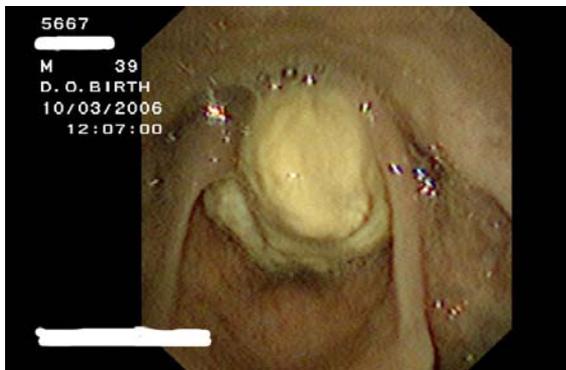


Figure 1: Laryngeal view showing mass obstructing the airway

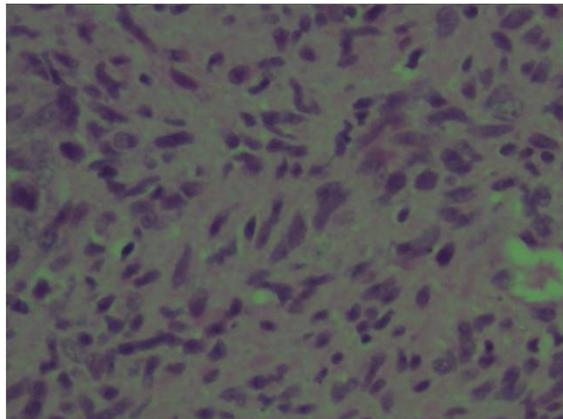


Figure 2: Slide showing presence of spindle cell

Discussion

Kaposi's sarcoma (KS) of the head and neck has been well described in the literature. KS of the upper aerodigestive tract is not unusual; the palate and gingiva are the most frequently involved sites.⁷ However, KS resulting in upper airway

obstruction in AIDS patients is extremely rare. The epiglottis appears to be the most common laryngeal site.^{6,8} As with any laryngeal disorder, the presenting symptoms can range from hoarseness and dysphagia to stridor or complete airway obstruction. Other symptoms commonly seen with laryngeal involvement are pain, bleeding and speech abnormalities.

The diagnosis can usually be easily established by fiberoptic laryngeal examination. After establishing the diagnosis, therapy is usually aimed at symptomatic relief.

Urgent intervention is indicated for lesions producing acute or impending airway obstruction. In this case, endotracheal tube intubation was impossible to be done because the mass was located at the supraglottic region and obstructing a direct view of the glottis. Tracheostomy should be immediately offered and should always be considered prior to any treatment protocol, since local therapy can often exacerbate airway compromise as mucositis and soft tissue edema develop.

Mochloulis G *et al* had experienced of significant hemorrhage and developed acute airway obstruction after performing biopsy of the KS lesion of the larynx.⁶ Therefore, he didn't recommend biopsy of suspected laryngeal KS. In our case, we were able to do direct laryngoscopy and took biopsy from the lesion using cold instrument without any complication. Schiff *et al* in his report of 2 cases had performed a biopsy using a carbon dioxide laser without major complication.⁸ Histologically, the distinctive features of KS are spindle-shaped cells with a random orientation, many extravasated erythrocytes, and thin vascular slits occurring in a reticular network of collagen fibers.

Treatment of laryngeal KS was in general, conservative. It consists of systemic and

local therapies. Systemic therapy is usually reserved for patients with rapidly progressing and/or widespread disease while local therapy is usually used to palliate pain and to improve function.⁸ Low dose radiotherapy to the larynx and systemic chemotherapy had been advocated.⁹ Apart from that, treatment of the HIV itself is important as KS often responds to the improvement in immunological status. In the late 1990s, the introduction of 2 new classes of antiretroviral drugs, the non-nucleoside analog reverse transcriptase inhibitors and protease inhibitors, resulted in improved antiretroviral efficacy in patients with AIDS.¹⁰ Combinations of three or more antiretroviral agents from at least two different classes became known as

HAART (Highly Active Antiretroviral treatment). Lukawska et al reported that more than 50% of patients with cutaneous KS responded to HAART.⁹ The cutaneous response is not as immediate as the virological and immunological responses, taking 3-6 months to improve. He had evaluated that mucosal presentation of KS has an equivalent response rate to cutaneous KS. Palliative radiotherapy was planned to this patient as he presented with life-threatening clinical disease progression. During the pre-HAART era, radiotherapy had an important role in the management of low-volume cutaneous KS. However, this method is now less common as a first-line treatment, and is becoming a second-line or third-line treatment after chemotherapy.

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