

## Laryngeal Kaposi Sarcoma in Non HIV Patient

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### Abstract:

Kaposi's sarcoma (KS) is acutaneous malignancy with multisystemic involvement and the larynx is one of the rare and unusual sites for involvement, we had reported a 75-year-old gentleman, who presented with progressive hoarseness with stidor. He underwent an emergency tracheostomy and fiberoptic laryngoscopy revealed a whitish globular laryngeal mass obscuring the glottic region, The patient also has many cutaneous lesions with involvement of oral cavity, nasal mucosa, nasopharynx biopsy was taken and histopathological study showed evidence of spindle shaped cell connective tissue consistent with Kaposi sarcoma, Laboratory test for (HIV) was negative, patient not diabetic no renal problem, no history of steroid or other drugs, no history of disease that affects his immune status, then we sent him to oncology unit and received systemic chemotherapy (paclitaxil) 225mg/m<sup>2</sup> every three weeks ,he improved and laryngeal lesion healed completely and tracheostomy closed. This case was reported because KS rarely involve the larynx with the narrowing of the airway and only two cases reported, and second thing is that our case was (HIV) negative.

**Key words:** Kaposi sarcoma; unusual location; larynx; HIV negative

### Introduction:

Kaposi's sarcoma (KS) is avascular tumor caused by Human herpes virus 8(HHV8), also known as Kaposi's sarcoma-associated herpesvirus. Kaposi's sarcoma (KS) is a systemic disease which can present with cutaneous lesions with or without internal involvement. Four subtypes have been described:

1- Classic KS, affecting middle aged men of Mediterranean and Jewish descent.

2-African endemic KS.

3-KS in iatrogenically immunosuppressed patients.

4-AIDS-related KS.

The erythematous to violaceous cutaneous lesions seen in KS have several morphologies: macular, patch,

localized or disseminated. KS can involve the oral cavity, lymph nodes, and viscera. Classic KS tends to be indolent, the presentation of AIDS patient with KS as laryngeal emergency is rare and only a few cases being reported in the literature.<sup>(1, 2, 3, 4)</sup> KS may involve every tissue in the human body. Incidence of KS Was 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.<sup>(5)</sup> Mochloulis G et. al. in his 10 years retrospective study showed the commonest site of laryngeal involvement was the supraglottis (65%).<sup>(6)</sup>

The aim from presenting this case to show that;

- 1-larynx is unusual and rare site for KS.
- 2-Good and dramatic response to systemic chemotherapy.
- 3-KS of the larynx can occur in non HIV patient.

### **Case presentation:**

Seventy five years old male patient presented with severe stridor in emergency unit in Kirkuk general hospital, on examining the patient, there were papular lesions ,red to purple in color involving lower extremities, both hands, ENT examination revealed red to bluish nodule involving the oral cavity,palate,nasal cavity,nasopharynx and the larynx was diffusely involved by the tumour(glottic and supraglottic) which made the airway to be narrow so emergency tracheostomy was done for him,after that we proceeded to investigate the patient, so skin and laryngeal biopsy revealed kaposi sarcoma,HIV and hepatitis tests were negative, patient has no other diseases that affect his immune status,patient refered to oncology unit.

In the oncology unit, he received chemotherapy with paclitaxel 225 mg/m<sup>2</sup> every three week. The patient was tolerating chemotherapy courses well with dramatic response after four courses of chemotherapy (three months) regarding improvement in his performance state to optimum and disappearance of the swellings and lymphedema in extremities mainly the lower limbs. Two additional paclitaxel doses had been added with complete disappearance of skin lesions and the laryngeal nodule, and tracheostomy tube

removed and site closed and patient regained his speech.

### **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. <sup>(7)</sup> However, KS resulting in upper airway obstruction in AIDS patients is extremely rare. The epiglottis appears to be the most common laryngeal site. <sup>(6, 8)</sup>

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.

Treatment of laryngeal KS is in general, conservative. It includes 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. <sup>(8)</sup> Low dose radiotherapy to the larynx and systemic chemotherapy had been advocated. <sup>(9)</sup>

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 anti retroviral drugs, the non-nucleoside analog reverse transcriptase inhibitors and protease inhibitors, resulted in improved antiretroviral efficacy in patients with AIDS. <sup>(10)</sup>

Lukawska et. al. reported that more than 50% of patients with cutaneous KS responded to HAART (highly active

anti retroviral treatment),<sup>(9)</sup> 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. Upper airway obstruction secondary to Kaposi's sarcoma, however, is extremely rare, and only two cases have been reported<sup>(11, 12)</sup>. Both were in men with previously diagnosed Kaposi's sarcoma who get hemorrhage and died shortly after tracheostomy. 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. Various chemotherapeutic agents were tested as first- and second-line treatments for CKS in small studies, with promising results. Because of the rarity and distribution of cases, only one randomized trial in patients with CKS has been published<sup>(13)</sup>. paclitaxel, oral etoposide<sup>(14-15)</sup>.

In our patient; the decision was to give paclitaxel as he got low ejection fraction which make use of anthracycline is controversial due to risk of cardiotoxicity and the new evidence were suggesting

paclitaxel belong to taxanes, which are microtubule stabilizing agents that have demonstrated effectiveness against KS associated with AIDS. Few anecdotal reports of the successful treatment of non-AIDS-associated KS with these agents are available in the medical literature<sup>(16, 18)</sup>. In a recent report Fardet et al., described 12 HIV-negative patients with KS who were treated successfully with taxanes after prior cytotoxic treatment had failed<sup>(19)</sup>.

These results suggest that taxanes are highly active in patients with non-HIV KS and the authors believe that drugs could be considered either as first-line or as second-line. Recently, Brambilla et al. have evaluated the clinical efficacy and tolerability of paclitaxel (100 mg weekly) in 17 patients with advanced aggressive and refractory CKS, The response to the therapy was evaluated after 12 weeks, Partial/ complete response was achieved in 14 of 17 patients, The treatment was generally well tolerated, Six patients having relapses, repeated paclitaxel with 5 having a good response therapy.

### **Conclusion:**

1. Larynx can be involved, in association with cutaneous (KS), with narrowing of the airway.
2. Laryngeal (KS) can occur in non (HIV) patients.
3. Laryngeal (KS) has good response to systemic chemotherapy.

**Before treatment**



Figure (1) KS in the larynx



Figure (2) KS in the oral cavity

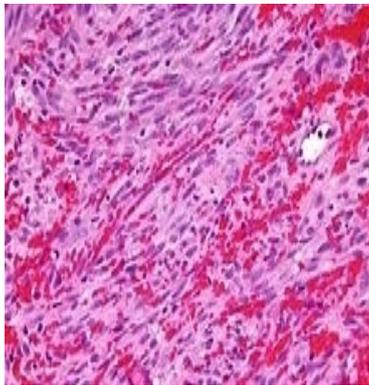


Figure (3) histopathological changes of KS



Figures (4) skin lesions of KS



Figures (5) skin lesions of KS



Figures (6) skin lesions of KS

### After treatment



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