

Localisation of mediterranean Kaposi's sarcoma in Morgagni's ventricle

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Abstract. *Localisation of mediterranean Kaposi's sarcoma in Morgagni's ventricle.* **Objective:** Head and neck involvement in Kaposi's Sarcoma (KS) is not unusual. However, laryngeal involvement is a relatively infrequent manifestation and ENT specialists should consider it in differential diagnosis in laryngeal lesions of AIDS patients and/or subjects from the Mediterranean area.

Methods: Case report and review of the literature in English.

Clinical Case: Male patient presenting with a three-month history of cough and acute dyspnoea. Laryngoscopy identified a laryngeal mass occluding the glottic plane. Tracheotomy was then performed and the laryngeal lesion was removed.

Histopathology showed neoplastic spindle cells that were positive to immunostain with CD-31 and CD-34, and immunoreactivity for HHV-8 was present. A diagnosis of KS was then suspected and confirmed after dermatological inspection.

Conclusion: Purple vascular mass lesions should lead in ENT to a high index of suspicion to exclude systemic diseases. Laryngeal KS must be included in the differential diagnosis of pigmented laryngeal lesions to plan correct management.

Introduction

The first description of Kaposi's sarcoma (KS) was by Kaposi in 1872,¹ who reported an unusual tumour consisting of small brownish-red cutaneous nodules, mainly involving the skin of lower extremities in a multifocal fashion.

The development of KS is related to several factors: genetics due to race and geographical distribution and infection because KS is found principally in patients with acquired immunodeficiency syndrome (AIDS).

Infection by HHV-8, a new human herpes virus described by Chang in 1994,² was considered necessary but not sufficient for KS development after HHV-8 DNA was found in tissues of patients affected by KS. The development of KS also requires compromised immune status.

Head and neck involvement in Kaposi's sarcoma (KS) is not unusual.³ However, laryngeal involvement is a somewhat infrequent manifestation. The literature describes about 50 cases of laryngeal KS.

There are four types of epidemiological KS: AIDS-related or epidemic, iatrogenic, African or endemic, and the classic or Mediterranean type. Three clinical forms of KS have been noted: localised nodular, locally aggressive and generalised. HIV-negative patients with KS of the larynx have been reported.^{4,5}

Males seem to be affected most (91%). The majority of patients with laryngeal KS have advanced HIV infection and were antiretrovirally naive.

Presenting symptoms may include hoarseness, throat discomfort, cough, dysphagia, stridor and

dyspnoea. Laryngoscopy may show laryngeal oedema or more often a purple vascular mass lesion.

We report on the first case in the literature in English of KS in Morgagni's ventricle.

Case report

A 65-year-old white male complaining of cough and hoarseness over the past three months attended the emergency room of our department for episodic dyspnoea and stridor. The history was negative for cigarette smoking and alcohol consumption. The patient presented with dysphonia and inspiratory stridor. The indirect laryngoscopy showed a reddish sessile mass arising from the superior aspect of the right true vocal cord. The diameter of the lesion at the largest point was 1.5 cm,

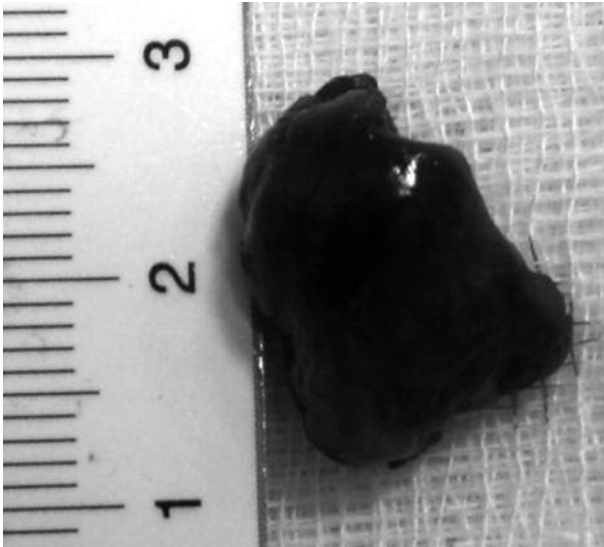


Figure 1
Laryngeal Kaposi's Sarcoma mass excised

compromising the respiratory passage. Vocal cord motility was normal, as was the rest of the mucosa. Head and neck clinical examination did not reveal lymph node disease.

The patient underwent an urgent tracheotomy because of the stridor, low oxygen saturation (average of 93%) and severe dyspnoea.

Pre-operative routine blood tests results were normal, as were thoracic radiographs.

The patient underwent a direct microlaryngoscopy, revealing that the true origin of the lesion was above the arcuate line in the fundus of the right Morgagni's ventricle. The lesion was completely excised (Figure 1) by means of laser-assisted surgery. The tracheostomy was then closed in the same intervention.

Histopathology revealed the proliferation of neoplastic spindle cells arranged in intersecting fascicles, which form slit-like vascular spaces of various dimensions containing red cells (Figure 2). The specimens were positive to immunostain with CD-31 and

CD-34. Nuclear immunoreactivity for HHV-8 was present, confirming the suspected diagnosis of KS (Figure 3).

The patient was then readmitted for further analysis and to evaluate the possibility of HIV infection. No signs of HIV infection were found. Blood tests were repeated and ELISA examination for HIV, CMV and EBV viruses were also performed. All values were in the

normal range. No virus infection was found.

A total body dermatological examination was then conducted, showing typical KS nodular lesions on both feet (Figure 4). The definitive diagnosis of Mediterranean or classic KS was made.

The patient was then followed-up. After one year, no recurrent laryngeal lesions were detected,

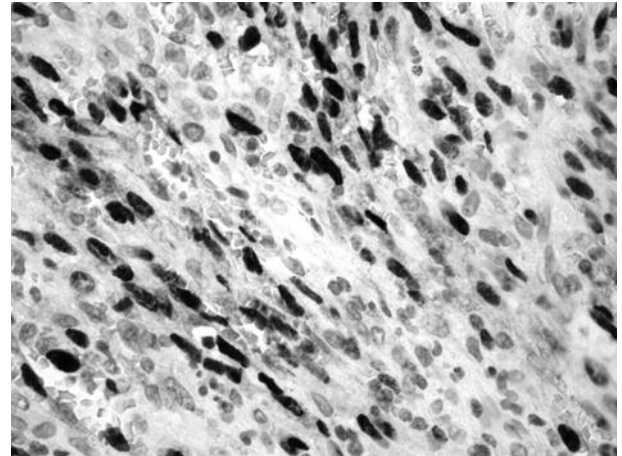


Figure 2
Neoplastic cells showing nuclear immunoreactivity for HHV-8. (Strept-ABC; original magnification 400×).

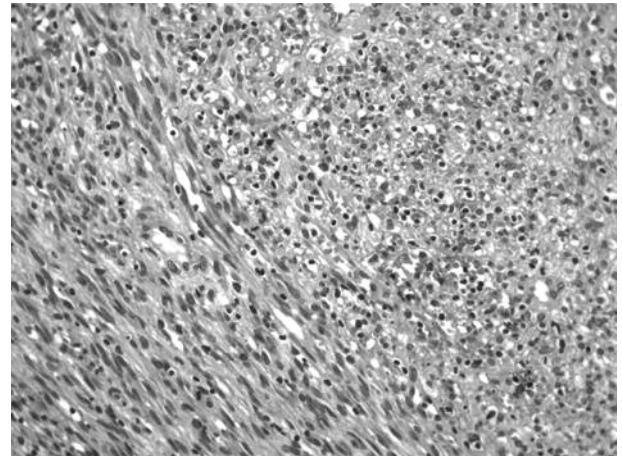


Figure 3
This section shows the proliferation of the neoplastic spindle cells arranged in intersecting fascicles which form slit-like vascular spaces of various dimensions containing red cells. (Hematoxylin-Eosin staining; original magnification 200×).



Figure 4

Nodular skin lesions on the right foot, typical of Classic Kaposi's Sarcoma.

and the skin lesion did not show any change.

Discussion

Mediterranean type or classic KS occurs in the older population of the Mediterranean area and it affects men 15 times more often than women. The nodular lesions are mainly situated in the lower extremities; they are indolent and infrequently cause morbidity. Conversely, the epidemic form of KS occurs in immunocompromised subjects (AIDS-related). The lesions in this form are aggressive and multicentric.

About 40% of AIDS patients will develop KS during the course of the disease, and two-thirds of

these patients will have head and neck involvement.⁶

The lesions are reddish-purple, multicentric macules and nodules. Histologically, the typical features of KS are spindle-shaped cells with numerous vascular channels and extravasated erythrocytes. Deposits of haemosiderin are present in a collagen stroma. An inflammatory infiltrate may be present. Immunostain is performed with HHV-8 antibodies to confirm the viral infection and hence the diagnosis.

The two largest series of patients affected by laryngeal KS have been reported by Abramson *et al.*⁷ in 1970 with 13 patients and by Mochloulis⁸ in 1996 with 17 cases. Classic KS of the larynx is a rare

Table I

The literature relating to Kaposi's sarcoma, including the present case*

Author	Year	Cases	Site involved	Local treatment	Type
Reynolds ¹⁰	1965	1	Arytenoids-Epiglottis	Tracheotomy	Classic
Coyas ¹¹	1983	1	Larynx	Tracheotomy	Classic
Abramson ⁷	1970	13	Larynx		Classic
Moritsch ¹⁶	1978	1	Larynx-Hypopharynx	Tracheotomy	Classic
Abemayor ³	1983	1	Epiglottis		Epidemic
Gnepp ¹²	1984	2	Anterior commissure (1) Larynx-Hypopharynx (1)	Excision (1) Tracheotomy (1)	Epidemic
Weidauer ¹⁷	1986	1	Larynx	Excision	Epidemic
Levy ¹⁸	1990	1	Glottic		Epidemic
Friedman ¹³	1996	6	Larynx	Intralesional vinblastine (6) Tracheotomy (1)	Epidemic
Mochloulis ⁸	1996	17	Supraglottic (11) Glottic (8) Subglottic (3)	Tracheotomy (1) Radiotherapy (5)	Epidemic
Schiff ¹⁹	1997	2	Epiglottis (2)	Biopsy (1) excision (1)	Epidemic
Beitler ⁹	1996	1	Larynx	Tracheotomy (1)	Epidemic
Gras ²⁰	1999	1	Supraglottic	Intralesional injection	Epidemic
Alkhuja ²¹	2001	1	Larynx	Tracheotomy	Epidemic
Ares ²²	2005	1	Larynx	Tracheotomy	Epidemic
Angouridakis ⁴	2006	1	Vocal cord	Excision	Classic
Ashurov ⁵	2007	1	Larynx		Classic
Dispenza [*]	2008	1	Larynx	Tracheotomy and excision	Classic

entity with less than 20 cases reported in the literature (Table 1).

The clinical presentation of laryngeal KS is variable, depending on the site and the stage of the disease. The symptoms range from dysphonic voice to stridor and dyspnoea due to upper airway obstruction. In the literature, ten cases of laryngeal KS needed tracheotomy,⁸⁻¹³ including the present case (Table 1).

Diagnostic biopsies were performed without complications in several patients, but biopsies of such vascular lesions have been associated with brisk and potentially fatal bleeding.⁸

The diagnosis of KS is generally made after histopathology, particularly in Classic KS, because of the lack of manifest extralaryngeal symptoms. In AIDS patients, the index of suspicion is higher than in other patients affected by the Mediterranean form.

Therapeutic options include: low-dose irradiation, intralesional chemotherapy, ablation and systemic therapy if there is disseminated disease. Obstructing laryngeal lesions require urgent intervention with tracheotomy. Depending upon the location of the lesion, a tracheotomy may contribute to mortality as a result of fatal haemorrhaging.⁹ A tracheotomy is generally adopted in these patients when transoral intubation for general anaesthesia is difficult or impossible, and in cases of severe dyspnoea. Furthermore, surgeons should bear in mind that Kaposi's nodules may be bleeding lesions, and so intraoperative haemorrhaging during transoral surgery may be well managed if the airways are saved. Surgical excision is adopted for isolated lesions and for those obstructing the airway. The tran-

soral modality for the surgical removal of the lesions is the first choice because it allows for the prompt and good visualisation of the glottic plane and rapid postoperative healing.

In our case the lesion was removed after airway patency was restored by tracheotomy. Tracheotomy may be closed once the lesion is removed.

Friedman *et al.* adopted the intralesional injection of Vinblastine sulphate, with an appreciable response in over 75% of cases. Only one patient required a tracheotomy for upper airway obstruction.¹³

The Mediterranean type of KS does not generally contribute to patient mortality and the treatment of asymptomatic laryngeal lesions may be conservative⁸, as in other laryngeal lesions.^{14,15} In the epidemic type, death is secondary to other AIDS-related disease processes.

Conclusion

Laryngeal KS is a rare clinical entity. To the best of our knowledge this is the first case reported in the English literature of KS affecting the laryngeal ventricle. The clinical investigation generally shows a mass without malignant features. The definitive diagnosis is established after histopathology, but the index of suspicion is high, especially in AIDS patients. It must be included in the differential diagnosis of pigmented laryngeal lesions. Urgent treatment may be required in cases of acute stridor due to an occluding mass. Classic KS is not an aggressive disease and patients usually die of other diseases, but follow-up is mandatory to prevent loco-

regional recurrence or new primary lesions.

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