



Case report

Bronchopulmonary Kaposi's sarcoma

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A B S T R A C T

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Kaposi's sarcoma (KS) is a highly vascular tumour, which was first described by the Hungarian dermatologist Moritz Kaposi Kohn before the discovery of the human immunodeficiency virus (HIV). Historically, KS has been linked to immunosuppression or to elderly male patients, especially in relation to diffuse cutaneous KS. We describe a case of Bronchopulmonary Kaposi's sarcoma in a patient with AIDS who was successfully treated with HAART and Liposomal Doxorubicin chemotherapy.

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Introduction

Kaposi's sarcoma (KS) is a highly vascular tumour, which was first described by the Hungarian dermatologist Moritz Kaposi Kohn before the discovery of the human immunodeficiency virus (HIV). It is caused by a herpes virus – human herpes virus 8 (HHV-8) – and is considered as one of the most common acquired immunodeficiency syndrome (AIDS) defining illnesses. Bronchopulmonary KS is not an uncommon presentation in patients with AIDS, especially in relation to diffuse cutaneous KS [1].

Case presentation

A 48-year-old Polish man was initially diagnosed with AIDS and skin Kaposi's sarcoma, during a visit to his home country. He presented to our department one year after the diagnosis, following the onset of new symptoms including shortness of breath on exertion, dry cough, and a mildly raised temperature. He looked unwell on admission to the hospital; his observations indicated tachycardia and no particular respiratory signs. The rest of the examination was unremarkable, apart from multiple skin lesions observed on his face, upper trunk, and legs. His medical background included mild asthma.

Laboratory tests showed the following: Haemoglobin 14.3 g/dl, leukocytes $5.1 \times 10^9/l$, CRP 1 mg/l, HIV-1RNA 1050 copy/ml, lymphocytes Ab's $1.33 \times 10^9/l$, CD4 T helper cells 2%, CD4 T helper cells $0.02 \times 10^9/l$, CD8 T suppressor cells 54%, CD8 T suppressor cells $0.71 \times 10^9/l$. The Ziehl Neelsen stain for acid-fast bacilli was negative.

The chest radiograph on admission displayed clear lung fields. A CT scan was subsequently performed, which revealed patchy

ground glass attenuation in all lobes, consistent with atypical infection. Based on these results, the patient was started on a high dose of intravenous antibiotics, to which he initially responded well, but failed to recover completely. Due to the persistence of his symptoms, a fiberoptic bronchoscopy was performed, along with biopsies which were sent for histopathology. During the bronchoscopy, a violaceous lesion was seen at the level of the right upper lobe take-off (Fig 1a). The histopathology indicated poorly formed vascular structures, which were highlighted in the immunohistochemistry for CD31, CD34, and Factor 8 (Fig. 2). This confirmed the suspicion of Kaposi's sarcoma.

The patient was started on HAART and Liposomal Doxorubicin chemotherapy, which led to a marked improvement of his skin lesions and the resolution of the endobronchial Kaposi's (Fig. 1b). It was also noted that his CD4 count and the viral load were stabilised. He was then covered for opportunistic fungal infection with antibiotic prophylaxis.

One year after the chemotherapy, the patient visited the chest clinic with symptoms of shortness of breath occasionally associated with reduced exercise distance, no sputum, and no haemoptysis. Spirometry showed an obstructive picture with reversibility post bronchodilator, which was in keeping with his diagnosis of asthma. A CT scan revealed the findings of bilateral consolidation/atelectasis, which could represent resolving infection. Fiberoptic bronchoscopy revealed no signs of KS; however, one of his sputum samples grew *Mycobacterium Kansaii*, for which he was started on a treatment of antibiotics for eighteen months.

Discussion

Kaposi's sarcoma is a tumour involving blood vessels and lymphatics. It is one of the highly vascular tumours and hence has a high risk of bleeding. KS is associated with the herpes virus (KSHV)

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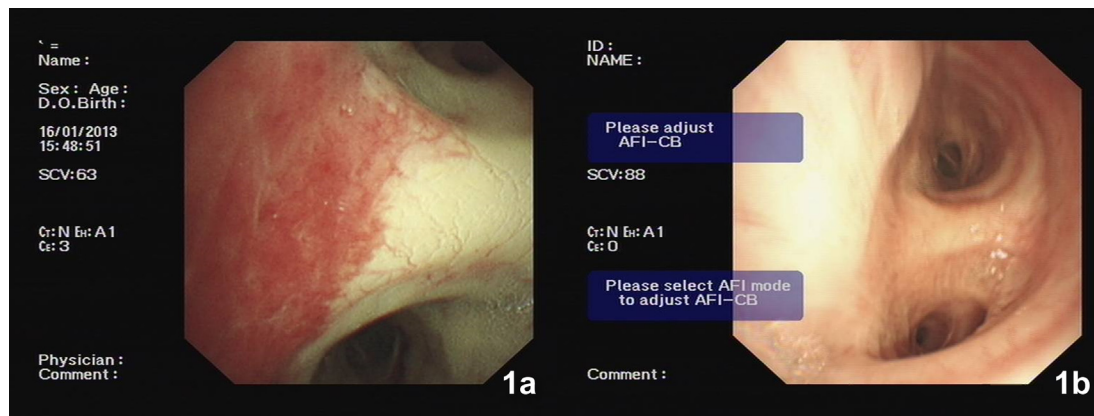


Fig. 1. Appearances of Bronchopulmonary KS pre. (1a) and post (1b) treatment.

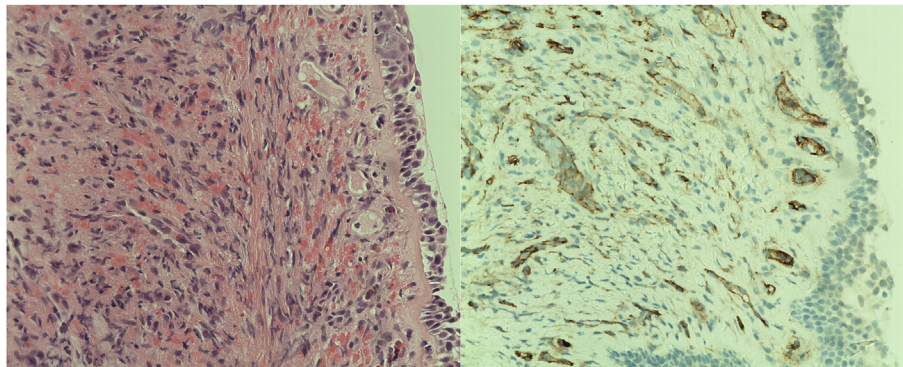


Fig. 2. Histopathology appearances of Bronchopulmonary KS.

and this has been considered as the causative factor for KS, with a mechanism involving the reactivation of the latent virus in immunocompromised patients [2]. Historically, KS has been linked to immunosuppression or to elderly male patients; however, there are reported cases which suggest its occurrence in immunocompetent young patients as well.

There are four main subtypes of KS with different clinical presentations: epidemic or AIDS related; endemic or African; classic or sporadic; and immunocompromised. The KS form which is related to AIDS has mostly been linked to homosexual men. There are fewer reported cases among women and heterosexuals [3].

Although a few cases have been reported as isolated visceral manifestations of KS, it is very uncommon for visceral involvement to occur without skin manifestations [4,5]. Visceral disease may affect the gastrointestinal tract, lungs, liver, and spleen. Pulmonary KS can present a wide range of symptoms and signs: progressive dyspnoea, dry cough, and low grade fever are the most frequent presentations [6]. Imaging, particularly CT, plays an important role in the diagnosis of KS lesions within the lungs. However, in order to confirm pulmonary involvement, it is crucial to perform a bronchoscopy [4,7].

There are many factors to be considered before deciding on how to treat KS. The National Cancer Institute recommends four different modalities of treatment that can be used in conjunction with the HIV treatment: radiation therapy, surgery, chemotherapy, and biological therapy. Since the introduction of highly active antiretroviral therapy (HAART) there has been a significant reduction in the rates of AIDS defining illnesses in general, including KS [8,9].

A study conducted by Gbabe et al. looked into the outcomes of treating patients with HAART alone, as against the usual usage of HAART and chemotherapy together [10]. They concluded that using HAART and chemotherapy together is superior to using HAART alone, especially with regard to progressive diseases. However, the results have not identified any difference between various chemotherapy regimens. Liposomal doxorubicin is widely used as the first line of chemotherapy, since it has been proved to be more effective when compared to other agents [11].

In conclusion, KS is one of the frequent complications of AIDS. In most cases, Bronchopulmonary KS occurs in patients who have had cutaneous involvement. In the author's experience of several cases, endobronchial biopsy has not caused significant haemorrhaging and has, in fact, helped in the confirmation of the diagnosis. Combining HAART and chemotherapy is the current standard treatment for Kaposi's sarcoma.

References

- [1] Garay S, Belenko M, Fazzini E, Schinella R. Pulmonary manifestations of kaposi's sarcoma. *CHEST J* 1987;91(1):39–43.
- [2] Gijshi O, Bottero V, Veettil MV, Dutta S, Singh VV, Chikoti L, et al. Kaposi's sarcoma-associated herpesvirus induces *nrf2* during de novo infection of endothelial cells to create a microenvironment conducive to infection. *PLoS Pathog* 2014;10(10):e1004460.
- [3] Shiels R. A history of kaposi's sarcoma. *J R Soc Med* 1986;79(9):532.
- [4] Gasparetto TD, Marchiori E, Lourenço S, Zanetti G, Vianna AD, Santos AA, et al. Pulmonary involvement in kaposi sarcoma: correlation between imaging and pathology. *Orphanet J Rare Dis* 2009;4(18).

- [5] Ayalew G, George A, Mongia S, Yudis C, Ponnambalam AJ, Sidhu GS. Kaposi sarcoma with extensive pulmonary involvement in an immunocompetent host. *CHEST J* 2010;138(4). Meeting Abstracts, 125A-125A.
- [6] Kang E, Staples C, McGuinness G, Primack S, Müller N. Detection and differential diagnosis of pulmonary infections and tumors in patients with aids: value of chest radiography versus ct. *AJR. Am J Roentgenol* 1996;166(1):15–9.
- [7] Kumar T, Epstein M, Markovskaya Y, Narasimhan M, Rosen M, Talwar A. Bronchoscopy and endobronchial disease in patients with human immunodeficiency virus infection. 2011.
- [8] Rubinstein PG, Aboulafia DM, Zloza A. Malignancies in hiv/aids: from epidemiology to therapeutic challenges. *AIDS* 2014;28(4):453–65.
- [9] Dufour V, Cadranet J, Wislez M, Lavole A, Bergot E, Parrot A, et al. Changes in the pattern of respiratory diseases necessitating hospitalization of hiv-infected patients since the advent of highly active antiretroviral therapy. *Lung* 2004;182(6):331–41.
- [10] Gbabe OF, Okwundu CI, Dedicat M, Freeman EE. Treatment of severe or progressive kaposi's sarcoma in hiv-infected adults. *The Cochrane Library*; 2014.
- [11] Nasti G, Errante D, Santarossa S, Vaccher E, Tirelli U. A risk and benefit assessment of treatment for aids-related kaposi's sarcoma. *Drug Saf* 1999;20(5):403–25.