

A Case Report of Intraluminal Tracheal Kaposi Sarcoma in Hiv-Infected Child

A Kanyamuhunga^{1,2,*}, D C Stefan^{1,3}

¹ Department of Pediatrics and Child Health, Faculty of Health Sciences, Stellenbosch University, South Africa

² Department of Pediatrics and child Health, Kigali University teaching Hospital, Rwanda

³ South African Medical Research Council, Parrow, Cape Town, South Africa

ABSTRACT

Kaposi's sarcoma is the most common HIV-associated malignancy; it is extremely associated with Human Herpes Virus 8 (HHV8). It usually starts on the skin and may later disseminate to visceral organs. Pulmonary Kaposi's sarcoma is uncommon especially in children and very rare as an obstructive mass in trachea. We are reporting a case of HIV-associated Kaposi's Sarcoma involving the lung parenchyma and obstructing the trachea.

Keywords: Kaposi's sarcoma - HIV - child - Intra-tracheal mass.

INTRODUCTION

Kaposi's sarcoma (KS) is a low-grade mesenchymal tumor involving the blood and lymphatic vessels that mostly affects the skin and may cause disseminated disease in a variety of organs. It is classified as sporadic KS, African-endemic KS, epidemic AIDS-related KS and non HIV-induced immunosuppression KS [1].

Despite being described as the most common neoplasm in patients with AIDS, its sporadic presentation is not rare and can sometimes elude clinical suspicion. It is seen as a cutaneous lesion typically affecting the skin of the extremities in classic-sporadic KS form. Epidemiologically it is most often observed in elderly patients. Pulmonary Kaposi's sarcoma is an unusual pre-mortem diagnosis in AIDS patients. A postmortem study found pulmonary KS in 47% of patients with cutaneous disease². In clinical studies pulmonary KS has been described in 3-13% of patients with AIDS overall, 6-32% of patients with AIDS and cutaneous KS and about 10% of patients with AIDS presented with respiratory symptoms [2,3]. In this paper we describe clinical features of 14-year-old girl with Kaposi's sarcoma obstructing the trachea causing a severe respiratory distress.

CASE PRESENTATION

A 14-year-old girl was admitted in Pediatrics Oncology Department at Tygerberg Hospital in February 2014, four months after being diagnosed with HIV and she was immediately started on highly active anti-retroviral therapy (HAART): ABACAVIR/LAMIVUDINE/EFVIRENZ after HIV infection diagnosis in October 2013. She presented with a chronic cough more than 6 months, which did not respond to empiric tuberculosis treatment; progressive severe dyspnea with stridor within 2 weeks prior to admission. The HIV-associated Kaposi's sarcoma was initially diagnosed in December 2013 on left axillary lymph nod biopsy.

At admission, her weight was 23 kg and her height was 130 cm. Her blood pressure was 90/60 mm Hg; her pulse rate was 155 beats per minute, respiratory rate 52 breaths

per minute and oxygen saturation of 85% on room air. The physical examination reveals an acute on chronically ill-looking patient, stunted, severely wasted, pale, lower limbs edema with generalized lymphadenopathy. Skin examination reveals two raised purple lesions suspicious of Kaposi's sarcoma noted on her left forearm as well as the left lower limb. The respiratory examination reveals tachypnea with severe stridor and diffused crackles in bases of both lung fields. The rest of exam was unremarkable except a smooth non-tender hepatomegaly. The investigations done showed: anemia, CD4: 25/mm³, HIV Viral load: log 1,6, hila infiltrates and intra-tracheal mass on Chest x ray (Figure 1), reticulonodular opacities involving bilateral basal regions on chest CT scan (figure 2).

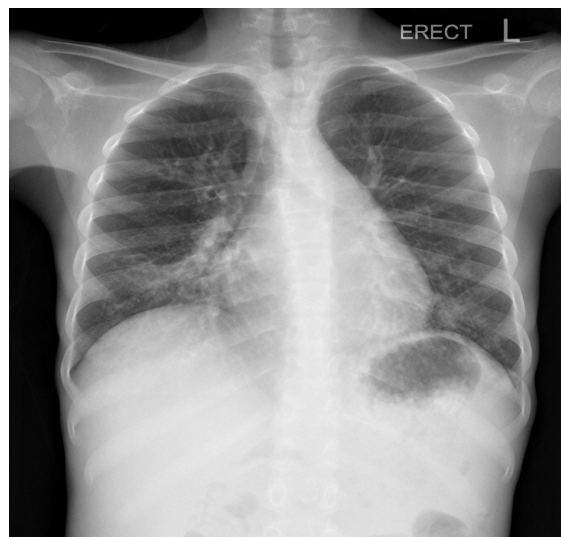


Figure 1: Bilateral reticulonodular interstitial opacities, most prominent in the perihilar distribution of the mid and lower lobe, rounded opacity interfering with normal tracheal clarity and blurring of the right heart border consistent with segmental middle lobe consolidation.

The Bronchoscopy done showed an intraluminal tracheal tumor causing 95% airway obstruction with features consistent with Kaposi sarcoma on histopathology with HHV-8 positive (figure 3). Based on the histopathological features of excisional biopsies, Kaposi's sarcoma was confirmed.

* Correspondence to: A. Kanyamuhunga, MD
Faculty of Health Sciences
Stellenbosch University
South Africa
Email: kanyamuhunga@yahoo.fr



Figure 2: Intraluminal tracheal tumor causing 95% airway obstruction

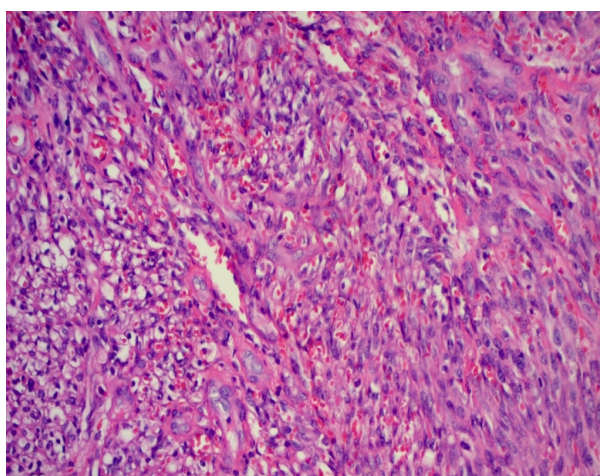
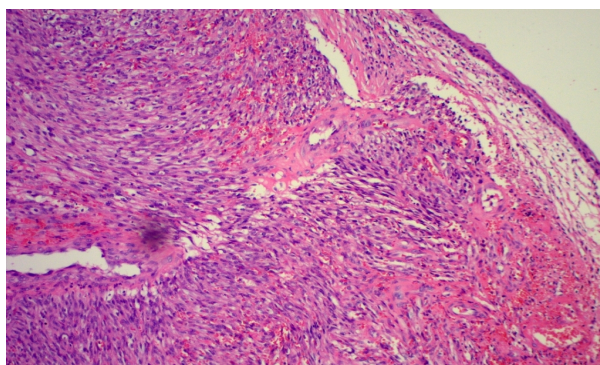


Figure 3: Sections show KS with spindle shaped cell forming slit like vascular channels. The surface shows necrosis and is lined by either respiratory or squamous epithelium

The patient was admitted and she got immediately a tracheostomy with a supportive treatment beside of HAART then systemic cytotoxic chemotherapy was scheduled as follow: Bleomycin 15units/m², Adriamycin 20 mg/m², and Vincristine 1.5 mg/m²; 3 weekly cycle interval. After 3 courses of ABV, the patient was improved and the bronchoscopy showed a markedly shrinkage of intraluminal tracheal mass but the tract was not yet formed thus closure of tracheostomy was scheduled after the 4th course of ABV regimen cytotoxic chemotherapy. The patient was discharged, clinically stable but

unfortunately She died at home due to bleeding from tracheostomy on 13th April 2014; one week prior to the 4th ABV chemotherapy cycle.

DISCUSSION

Dr. Moritz Kaposi first described Kaposi's sarcoma in 1872. It is one of the AIDS defining skin diseases and is strongly linked to oncogenic virus human herpes virus type 8 (HHV-8) [4,5]. KS is characterized by few or widespread multifocal brown violaceous or dark red color patches, papules, plaques and sometimes deep skin nodules. Typically, the lesions are bilateral, symmetrically distributed along the lines of skin cleavage, involving the extremities. However, in advanced AIDS, KS will often disseminate to the oropharynx, larynx, tracheobronchial tree, lungs, and other viscera [3,5]. Thoracic disease is found in approximately 45% of patients with cutaneous AIDS-related KS [6]. Manifestations include parenchymal, tracheal, lymphatic, pleural, and chest wall abnormalities. Common clinical symptoms are chronic cough, dyspnea, fever, and hemoptysis [7]. A typical purplish endoscopic appearance of tracheobronchial KS is the key finding and helps to raise index of suspicion. The AIDS-related pulmonary Kaposi's sarcoma is a fatal condition, about 20% of deaths are related to its complications such as: upper airway obstruction, hemorrhage, or lung parenchymal destruction; but the majority of deaths in these patients are related to concomitant infection. The thoracic involvement by AIDS-related Kaposi's sarcoma is generally seen in the patients with advanced disease stage with a very low CD4 count less than 100 cells/mm³ mostly when not yet treated with HAART [8]. In the present case, the last CD4 before these symptoms was 25cells/mm³.

The pathogenesis of Kaposi's sarcoma is still under investigation. Current studies have focused on the search for causative infectious agent mainly due to its correlation with immune-compromised patients. A large epidemiologic study has strongly linked all KS clinical variants with HHV-8 infection and further work established that HHV-8 infection heads the development of KS lesions [9]. The co-infection with HHV-8 and HIV has synergistic impact on the development of the KS lesions. The exact mechanism by which HIV infection facilitates HHV-8 induced tumor genesis remains unclear, but may relate to enhanced growth stimulated by the HIV tat protein or increased production of cytokines and growth factors, the HHV-8 is deemed necessary but insufficient cause of Kaposi's sarcoma [10].

Management of HIV-associated Kaposi's sarcoma is still a matter of debate. The specific treatment is highly dependent on the nature and extent of lesions and areas of body involvement. It is aimed to reduce intensive skin disease, shrinkage of problematic lesions, pain control and alleviation of symptomatic visceral disease. It involves the use of HAART, radiotherapy and systemic chemotherapy [5]. In our case, the patient was on HAART for 4 months, the systemic chemotherapy was initiated based on ABV regimen protocol with better clinical improvement after the second cycle of chemotherapy. No matter what treatment modality chosen, clinicians should bear in mind that even in its classical form, KS may be a malignant,

rapidly progressing tumor with visceral involvement. Primary obstructive tracheal KS is relatively uncommon disorder in general and very unusual presentation in pediatric patients with AIDS thus we would like to outline the following take-home messages for the primary care clinicians from this patient's case report (i) in patients with KS or even those on HAART, they should be warned of deterioration within the first 3-4 months of treatment; (ii) patients with HIV-related KS with persistent respiratory symptoms despite the tuberculosis treatment course given should be thoroughly examined to exclude the pulmonary Kaposi's sarcoma; (iii) bronchoscopy is highly recommended to an HIV-related KS with chest signs including stridor regardless the age; and (iv) pulmonary Kaposi's sarcoma is fatal and requires systemic cytotoxic chemotherapy.

CONCLUSION

Kaposi's sarcoma is the most common neoplastic disease associated with HIV that, in advanced AIDS, may disseminate to lungs parenchymal. The persistent respiratory signs with intermittent stridor to HIV-related KS should heighten the physician's awareness of existing an obstructive tracheal KS for early diagnosis and treatment. In this paper, we presented unusual intraluminal tracheal KS in 14 year old girl with HIV infection on HAART.

REFERENCES

1. Frederick JS, Ramzi SC. Blood vessels. In: Cotran RS, Kumar V, Collins T Robbins Pathologic Basis of disease. Philadelphia, WB Saunders, 1999: 535-537.
2. Fouret PJ, Touboul JL, Maynaud CD, Akoun GM, Roland J. Pulmonary Kaposi's sarcoma in patients with acquired immune deficiency syndrome: a clinicopathological study. *Thorax* 1987; 42:262-8.
3. Haramati LB, Wong J: Intrathoracic Kaposi's sarcoma in women with AIDS. *Chest* 2000, 117:410-414.
4. Wood NH, Feller L: The malignant potential of HIV-associated Kaposi sarcoma. *Cancer Cell Int* 2008, 8:14.
5. Feller L, Lemmer J, Wood NH, Jadwat Y, Raubenheimer EJ. HIV-associated oral Kaposi's sarcoma and HHV-8. A review. *J Int Acad Periodontol*. 2007; 9(4): 129-136.
6. Restrepo CS, Martinez S, Lemos JA, Carrillo JA, Lemos DF, Ojeda P, Koshy P: Imaging manifestations of Kaposi sarcoma. *Radio- graphics* 2006, 26:1169-1185.
7. Hannon FB, Easterbrook PJ, Padley S, Boag F, Goodall R, Phillips RH:
8. Bronchopulmonary Kaposi's sarcoma in 106 HIV-1 infected patients. *Int J STD AIDS* 1998, 9:518-525.
9. Marchiori E, Muller NL, Soares Souza A Jr, Escuissato DL, Gasparetto EL, Franquet T: Pulmonary disease in patients with AIDS: high- resolution CT and pathologic findings. *AJR Am J Roentgenol* 2005, 184:757-764.
10. Antman K, Chang Y: Kaposi's sarcoma. *N Engl J Med* 2000, 342: 1027-1038.
11. Mbulaiteye SM, Goedert JJ. Transmission of Kaposi's sarcoma-associated herpes virus in Sub-Saharan Africa. *AIDS*. 2008; 22(4):535-537.