

Disseminated Kaposi Sarcoma and Lymphoplasmocytic Interstitial Pneumonitis as Initial Presentation of HIV Infection

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INTRODUCTION

- Pulmonary infections are the major cause of morbidity and mortality in persons infected with HIV; however, non infectious disorders of the lung have been identified.
- Kaposi sarcoma (KS), a vascular tumor of low grade malignant potential is associated with Human Herpes Virus 8 (HHV8) in HIV infected and non infected individuals.
- Lymphoplasmocytic interstitial pneumonitis (LIP) is characterized by infiltration of alveolar interstitial spaces by lymphocytes, plasma cells and other lymphoreticular elements.
- We describe an unusual presentation of an HIV infected individual as disseminated KS involving lung, muco-cutaneous sites and bone with concomitant LIP.

HISTORY

A 33 year-old woman, an immigrant from Kenya, presented with neck swelling that increased in size over a period of one month, progressive exertional dyspnea of two months duration, and increasing back pain over four months.

PHYSICAL EXAMINATION

- Tachycardia.
- Lymphadenopathy in the posterior chain of the neck.
- Bronchovesicular breath sounds.
- Nodular skin lesions.
- Tenderness over the thoracic spine.

LABS AND TESTS

• CD4 T-cell count	326 cells/mm ³
• HIV viral load	> 5.9 log
• PFT	Isolated moderate reduction in DLCO
• CT chest	Nodular opacities in upper lobes, ground glass opacities in right middle lobe and lingula. Lytic lesions at T1, T2 and L3 vertebrae
• MRI spine	Abnormal enhancement in multiple vertebrae
• Lung biopsy	Kaposi sarcoma in background of LIP
• Vertebral biopsy	Kaposi sarcoma
• Skin/nasal biopsy	Kaposi sarcoma
• Neck mass biopsy	Kaposi sarcoma
• Echocardiogram	Right to left extra cardiac shunt

CLINICAL COURSE

- The patient was initiated on highly active anti-retroviral therapy (HAART) and subsequently treated with chemotherapy with clinical improvement.

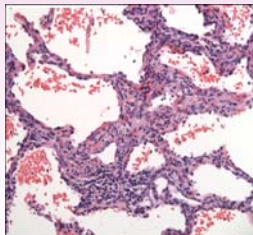


Figure 1. Lung wedge biopsy showing KS and LIP. KS travels alongside vessels and capillaries in the interstitium. LIP occupies both peribronchial and periarterial tissue.

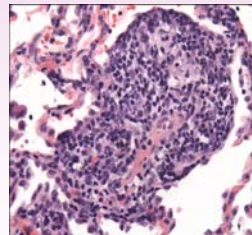


Figure 2. Lung wedge biopsy showing LIP. Infiltrates are composed of lymphocytes, plasma cells, monocytes and epithelioid histiocytes.

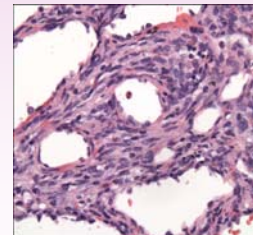


Figure 3. Lung wedge biopsy featuring KS. Histology features bland tightly packed spindle cells, with red cells scattered in between spindle cells.

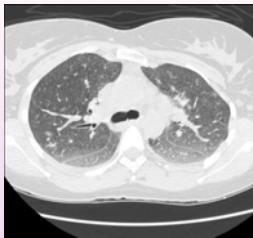


Figure 4. CT chest showing nodular opacities.



Figure 5. MRI spine showing abnormal enhancement in multiple vertebrae.

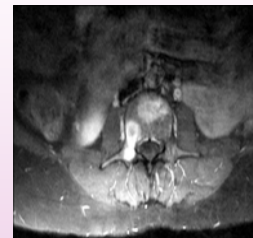


Figure 6. MRI spine showing abnormal enhancement in a vertebra.

DISCUSSION

- Although the incidence of Kaposi sarcoma is declining it still remains an important neoplasm in patients with AIDS.
- Our patient is particularly interesting given the disseminated nature of KS involving visceral organs as well as unusual sites like bone. Isolated osseous KS lesions are usually seen in patients with a CD4 count less than 100 cells/mm³.
- The cause of LIP is unknown. A possible viral etiology for LIP is suggested by it's association with HIV, EBV and HTLV-1. The concomitant LIP probably explains the dyspnea; however, the reduction in DLCO may be partly explained by a right to left extra cardiac shunt.
- It is extremely unusual to see KS and LIP present together in adults.
- It is particularly challenging to treat patients with KS and HIV without immune compromising them especially since steroid treatment is suggested in individuals with LIP of moderate severity.
- Although reported cases of LIP in HIV-infected adults failed to demonstrate improvement with antiviral therapy alone, our patient showed clinical and radiological improvement with initiation of HAART and chemotherapy for KS suggesting a direct role of HIV in causing LIP.

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