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## Bilateral Chest X-Ray Shadowing and Bilateral leg lesions - A case of Pulmonary Kaposi Sarcoma

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# Bilateral Chest X-Ray Shadowing and Bilateral leg lesions - A case of Pulmonary Kaposi Sarcoma

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## Abstract

We report a case of a 30 year old gentleman seen on the respiratory ward with no significant past medical history presenting with a three week history of worsening dyspnoea, productive cough, fever, bilateral leg swelling and bilateral leg swelling. Initial differential diagnosis included community-acquired pneumonia, cellulitis and deep vein thrombosis. After much investigation a diagnosis of AIDS-related kaposi's sarcoma with visceral manifestations was made.

## Introduction

Kaposi sarcoma (KS) is a low-grade vascular tumor associated with kaposi sarcoma herpesvirus/human herpesvirus 8 (KSHV/HHV8) infection. HIV-associated KS is typically seen in patients who have a low CD4 cell count (< 150 cells per cubic millimeter) and a high viral load (>10,000 copies per milliliter).<sup>1</sup>

Kaposi sarcoma lesions predominantly present at mucocutaneous sites, but may involve all organs and anatomic locations, with the gastrointestinal tract and lungs being commonly affected.

## Case Report(s)

A 30 year old gentleman presented to Accident & Emergency with a three week history of worsening dyspnoea, productive cough, fever and bilateral leg swelling. The patient denied having any neurological symptoms, chest pain, abdominal pain, rigors, night sweats, hemoptysis, hematemesis or rectal bleeding.

The patient had no significant medical or sexual history, was not taking any regular medicines, had not donated or received blood transfusions, denied being an intravenous drug user and denied any recent travel abroad.

On admission the patient looked unwell, had tachycardia and a temperature of 40 degrees Celsius. Examination revealed bilateral reduced breath sounds, enlarged cervical lymph nodes, splenomegaly, and

erythematous tender lower limbs with bilateral cutaneous lesions on the legs.

The emergency department started presumptive treatment for bilateral cellulitis with intravenous fluids and antibiotics. Admission laboratory results demonstrated anaemia (haemoglobin of 76 g/L), thrombocytopenia ( $45 \times 10^3/\text{mm}^3$ ) a white blood cell count of  $5.1 \times 10^3/\text{mm}^3$  and a CRP of 61 mg/dL. A chest X-Ray was performed which showed bilateral hilar shadowing and interstitial reticular shadowing in the lower zones. (illustration 1) Due to the patients low haemoglobin the patient received 2 units of red blood cells.

Based on these results differentials included atypical infections, tuberculosis, haematological malignancies and HIV associated pneumocystis pneumonia and kaposi Sarcoma. In order to investigate these a HIV test, haematinics screen, atypical urine antigens, sputum for AAFB and MC+S and blood cultures were obtained.

The patients HIV test came back positive, sputum samples did not detect any organism and atypical pneumonia antigens were negative.

The genitourinary, haematological and dermatological teams were subsequently consulted regarding this patient and following these consultations further investigation took place. The patients CD4 count was 12 count cells/mm<sup>3</sup> and HIV Viral Load was 1420045 copies/ml. The patient also had genotypic testing for HIV drug resistance and antiretroviral therapy with truvada and raltegravir was started. The patient was also started on atovaquone as Pneumocystis pneumonia prophylaxes as the CD4 count was less than 50 count cells/mm<sup>3</sup>.

The haematology team carried out a bone marrow aspirate and a trephine biopsy to investigate for haematological malignancies however the bone marrow aspirate unfortunately clotted, and it was decided that the patient would have a lymph node biopsy instead. The patient's haemoglobin was 66 g/L despite receiving two units of red blood cells three days ago, and the patient received another 2 units of red blood cells.

The dermatology team carried out a punch biopsy which was positive for HHV-8 in keeping with the diagnosis of cutaneous kaposi Sarcoma.

A CT/High-Resolution CT was performed which showed mediastinal lymphadenopathy, lungs nodularity and patchy opacity which might represent part of the kaposi sarcoma and features of superadded infection/atypical infection were also seen. The CT also showed a gross splenomegaly and hepatomegaly. (illustration 2)

Bronchoscopy revealed oral kaposi sarcoma lesions, and patches of kaposi Sarcoma were also seen in the trachea and main lobar bronchi. No biopsy was taken due to the patients low platelets, however bronchial washings for PCP and TB were taken, both of which were negative.

The patient was discharged with a lymph node biopsy booked as outpatient, and with follow-up from the genitourinary team in regards to further treatment after results of lymph node biopsy were available. The patient had the lymph node biopsy five days after discharge, and this showed lymph node was extensively replaced by kaposi sarcoma, marked plasmacytic hyperplasia (polytypic plasmacytic proliferation), with focal EBV positivity. This is entirely consistent with immunodeficiency associated plasmacytic hyperplasia (with EBV positivity). There was no evidence of malignant lymphoma and no opportunistic infection was identified.

The patient developed a severe pneumonia before he could be assessed for chemotherapy in regards to treatment of kaposi Sarcoma. He required admission to the intensive care unit, was ventilated and despite treatment with antiretrovirals, steroids, fluids and antibiotics the patient developed multi-organ failure and died.

## Discussion

Incidence of kaposi Sarcoma increase during the 1980's and 1990's which was attributed to the increasing number of cases of AIDS. The introduction of highly active antiretroviral therapy (HAART) has helped decreased the incidence of AIDS and KS. Kaposi sarcoma however remains a commonly seen AIDS-defining illness particularly in some African countries where the incidence of untreated HIV remain high.<sup>3</sup>

Kaposi sarcoma usually presents as mucocutaneous lesions which can be red, brown or purple and nodular, papular or blotchy. Lesions typically increase in size and numbers as the disease progresses. Lesions may also involve the internal organs such as the lungs, gastrointestinal tract and lymphatic system. Kaposi Sarcoma involving the respiratory tract has been reported in 6-32% of HIV positive patients with cutaneous disease.<sup>4</sup>

Pulmonary KS can present with constitutional symptoms such fever, malaise, and weight loss, and respiratory symptoms such as dyspnoea, fever, hemoptysis and chest pain, and can involve the trachea, lung parenchyma, pleural spaces, airways and lymph nodes.<sup>5</sup> Studies have shown that approximately 20% of deaths in pulmonary KS are a complication of the disease such as; airway obstruction, haemorrhage and parenchymal destruction. The majority of deaths are related to other factors such as superadded infection.<sup>6,7</sup>

Radiological imaging, particularly Computed Tomography (CT) scans of the thorax and abdomen, plays an important role in the diagnosis of pulmonary KS lesions and potential opportunistic infection. CT scans may demonstrate; reticular changes in the middle and lower zone ;parenchymal nodules that can progress to consolidation with pleural collections and hilar or mediastinal adenopathies.<sup>8</sup> Bronchoscopy is required to confirm the diagnosis of pulmonary KS. Classical red/purple macular KS lesions may be seen during bronchoscopy and the presence of these alongside radiological findings, skin biopsy and positive HIV test allows the presumptive diagnosis of pulmonary KS to be made without a transbronchial biopsy. Biopsy is only recommended when the clinical and radiological findings are not typical, and a definitive diagnosis is required.<sup>9</sup>

The principle aims of treating pulmonary KS are preventing progression of disease and reduction of tumor burden. Prompt initiation of highly active antiretroviral therapy (HAART) is recommended, and some patients with pulmonary KS have adequate treatment response with HAART alone. Other treatment modalities need to be considered alongside HAART in patients with symptomatic pulmonary KS and those with advanced or progressing disease. The National Cancer Institute recommends the use of; surgery, radiotherapy and chemotherapy in conjunction with HAART in these patients after consideration is given to the prognosis of the patient and a case-by-case benefit and risk analysis of using

these modalities.<sup>10</sup>

Liposomal doxorubicin and liposomal danorubicin are anthracyclines which are widely used as first line chemotherapy agents based on studies showing higher efficacies as compared to other chemotherapy agents in the treatment of KS.<sup>11</sup>

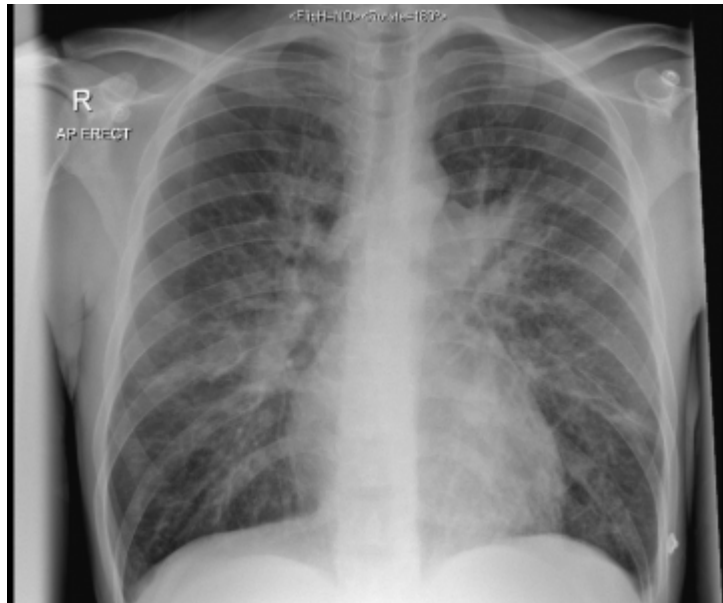
Despite the use of HAART and chemotherapy, pulmonary KS remains a disease associated with high levels of mortality, with median survivals reported as one and a half year. Visceral involvement of KS is a poor prognosis factor, with a large study reporting five-year survival for patients with pulmonary KS was 49% as compared to 82% for those with classic KS.<sup>12</sup>

Kaposi sarcoma is a common AIDS defining disease and pulmonary KS is a potentially life-threatening disease. HIV patients and patients presenting with atypical infections/imaging presenting with worsening respiratory symptoms should have the differential of pulmonary KS considered, particularly if cutaneous lesions are also seen. Early HIV testing, skin biopsy, radiological imaging and bronchoscopy are recommended so that prompt diagnosis can be made and early HAART and chemotherapy can be initiated.

## Illustrations

### Illustration 1

Chest X-Ray of patient showing hilar enlargement and focal enlargement of the left region Superiorly which has mass like appearance.



### Illustration 2

Computed tomography scan showing grossly enlarged spleen up to 22 cm and an enlarged liver.

