CASE REPORT

Pulmonary manifestations of Kaposi Sarcoma: A Case report and brief review

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ABSTRACT

Infection with Human Immune deficiency Virus (HIV) is relatively uncommon. Such infection predisposes its host to a plethora of infectious, inflammatory, and neoplastic diseases. Kaposi Sarcoma (KS), a vascular malignancy with a mucocutaneous predilection, is the most strongly linked neoplasm to HIV infection. The thorax is among the common victims of KS, with involvement of lung parenchyma, lymph nodes and pleura. This is an instructive case report of the spectrum of thoracic involvement in KS.

Key Words: *HIV; Pulmonary nodules; Kaposi sarcoma; Malignancy*

Recent global estimates indicate that approximately 33 million people are living with human immune deficiency virus (HIV) infection. A disproportionate number live in low- and middle-income countries, with the highest prevalence of HIV infection reported in sub-Saharan Africa (1). Estimates by the World Health Organization and the joint United Nations programme on HIV/AID show that HIV prevalence is low in the Middle East and North Africa region (0.2%) (2).

Pulmonary manifestations of infection with HIV are protean, both infectious and non-infectious. These complications include diseases that are Aquired Immune Deficiency Syndrome (AIDS)-defining (e.g., Pneumocystis pneumonia) or HIV-associated (e.g., bacterial pneumonia), disorders that are not classified as HIV-associated but appear to be more common in those with HIV infection (e.g., lung cancer, pulmonary arterial hypertension, and chronic obstructive pulmonary disease), and conditions whose association with HIV is inconclusive or purely coincidental (e.g., sarcoidosis) (3). The lungs are also common seat for malignancy in patients with HIV. In addition to the increased incidence of bronchogenic carcinoma among HIV patients (after controlling for smoking history), two of the AIDS-defining illnesses; non-Hodgkin's lymphoma and Kaposi Sarcoma (KS), commonly involve the lung (4).

Heitzman et al. reported that one third of patients with AIDS and KS who have required evaluation for pulmonary complaints KS were proved to have thoracic involvement by KS (5). At autopsy, the extent of pulmonary involvement approaches 50% to 75% (3). Most but not all patients with clinically diagnosed pulmonary Kaposi sarcoma have concomitant mucocutaneous disease (3).

The following report describes the case of a homosexual man who acquired HIV infection and later developed Kaposi Sarcoma with the full picture of pulmonary involvement.

CASE HISTORY

A 29-year-old male who works as a wall painter presented to the outpatient clinic of our institute with three-month history of progressive dyspnea and repeated bouts of hemoptysis in the form of blood-tinged sputum. He was a moderate smoker. There was no previous medical history aside from mild occupational asthma. He was single and admitted heterosexual extramarital relationship. At presentation, the patient was in respiratory distress and had central cyanosis. There was mild pyrexia and tachycardia but normal blood pressure. There was diffuse papular rash on the patient's face and extremities that was violaceous in color (Figure 1).

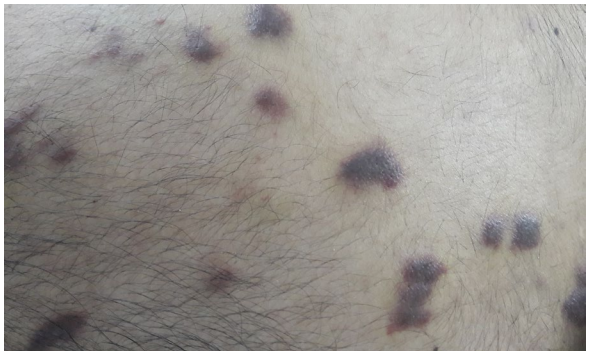

Figure 1) *Violaceous papular rash on the patient's arm*

Similar lesions were found on the back and external genitalia. There was sub-conjuctival hemorrhage in the left eye. Black discoloration was noted on the hard palate. There was no peripheral adenopathy, lower limb edema or finger clubbing. Cardiac examination was normal and lung examination revealed mild wheezes and scattered crepitations bilaterally. Routine laboratory investigations revealed mild anemia and leucocytopenia.

A chest x-ray that the patient presented with revealed multiple bilateral pulmonary nodules in all lung fields. A chest CT with IV contrast was ordered. The study revealed bilateral confluent nodules that showed contrast enhancement (Figures 2A and 2B).

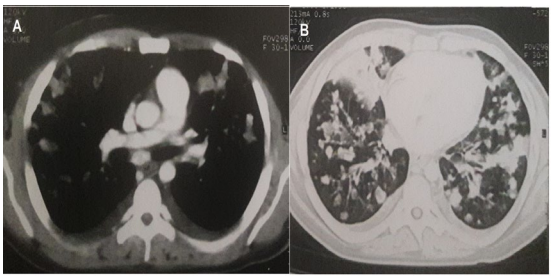

Figure 2) *(A) Bilateral enhancing pulmonary nodules in upper lobes as well as bilateral pleural effusion and very brightly enhancing subcarinal lymph node. (B) Bilateral variable-sized nodules and right lower lobe consolidation*

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Mild bilateral pleural effusion was also noted as well as several enlarged lymph node groups, notably the preaortic and subcarinal groups. Strikingly, the subcarinal lymph nodes exhibited significant contrast uptake (Figure 2A).

Abdominal ultrasound was done and revealed bilateral renal calyceal stones with normal sized liver and spleen with no evidence of ascites or lymphadenopathy. Given the sexual history of the patient and the peculiar morphology of the cutaneous lesions, dermatologist consultation was sought, and serologic test for HIV was ordered.

To our surprise, the patient was already known and diagnosed by colleagues from the dermatology clinic. He previously tested positive for HIV by PCR and the skin lesions were diagnosed as muco-cutaneous KS secondary to HIV infection. Re-asking the patient, he admitted he was previously engaged in homosexual activity.

Given this diagnosis, all radiological abnormalities fit into the range of manifestations reported about thoracic involvement in KS and thus our final diagnosis was metastatic pulmonary KS causing respiratory failure in an HIV positive patient.

Unfortunately, until this point of time the patient avoided seeking medication for HIV due to the stigma of the disease. Only respiratory distress was the driver to seek medical advice again. The patient improved on oxygen therapy and was transferred to Fever Hospital to start anti-retroviral therapy (ART).

DISCUSSION

The most common HIV-associated malignancy is Kaposi sarcoma, although its incidence has decreased dramatically with combination ART [6]. Kaposi sarcoma is an angioproliferative tumor that most commonly presents with mucocutaneous involvement (7).

The incidence of HIV-associated Kaposi sarcoma has always been significantly higher in homosexual men than in other HIV risk groups (90% to 95% of cases) (8). The disease has been causally linked to infection with human herpes virus (HHV8), also known as Kaposi sarcoma-associated herpes virus (KSHV) or KS agent, which was isolated from patient with HIV- and non-HIV-associated KS (3). It became more widely known as one of the AIDS-defining illnesses in the 1980s. Kaposi sarcoma is a systemic disease that can present with cutaneous lesions with or without visceral involvement. In the context of immune suppresion, the erythematous to violaceous cutaneous lesions seen in KS can be pathognomonic (9).

These lesions have several morphologies; macular, patch, plaque, nodular, and exophytic. The cutaneous lesions can be solitary, localized or disseminated. KS can involve the oral cavity, lymph nodes, and viscera. Classic KS tends to be indolent, presenting with erythematous or violaceous patches on the lower extremities. (Figure 1) Dermatologic manifestations of KS can be alarming, but it is the visceral involvement that is most commonly life threatening. Pulmonary KS may be difficult to differentiate from other infectious or neoplastic conditions, yet the distinction is essential, for without treatment, patients with pulmonary KS have a median survival of only a few months (10). Pulmonary Kaposi sarcoma presents at the lower range of CD4+ lymphocyte counts, typically < 200 cells/cm^2 (3).

In the presented case, the patient was referred to us for evaluation of diffuse nodular infiltrates of the lung. Such picture can be encountered in disseminated malignancy, certain infections and vasculitis (11). The very characteristic muco-cutaneous lesions were the driver for the work-up for HIV infection. The patient's CT showed all the reported thoracic manifestations of KS, namely; pulmonary nodules of varying sizes, consolidations, pleural effusion and lymphadenopathy (Figure 2).

CONCLUSION

The final diagnosis of pulmonary KS is ideally done by demonstrating the typical violaceous mucosal lesions in the trachea or main bronchi at bronchoscopy (10). Unfortunately, this was not feasible in our patient due to high oxygen needs of the patients and feared consequences of procedure-induced hypoxia on the patient's safety.

Tumors can regress in size and number in response to ART, and therefore all patients with Kaposi sarcoma should receive combination ART if no other contraindications exist (12). Treatment of systemic disease also includes chemotherapy, with approved agents consisting of doxorubicin, daunorubicin, and paclitaxel; in addition, the antiviral agent cidofovir, although not effective on its own, may have a role as adjunctive therapy (12).

Survival in patients with pulmonary Kaposi sarcoma appears substantially worse when compared to patients without pulmonary involvement (3).

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