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A young male patient presented with dyspnea, hemoptysis, and bilateral pulmonary nodular infiltrations: What is your diagnosis?

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A 26-year-old male patient presented to the outpatient clinic of Infectious Diseases Department with complaints of fever, shortness of breath, and hemoptysis. His complaints started 3 weeks ago, and he applied to another hospital, but his general condition got worse under the given treatment. Then, the patient was admitted to our institution. He recently had complaints of fatigue, weight loss, dyspnea, and anorexia, which got gradually worse.

In his physical examination, his body temperature was 37.2°C, peripheral oxygen saturation was 90% (at room temperature), and he had tachypnea. There was no edema, icterus, or cyanosis. There were purplish plaques measuring 1–3 cm on the upper and lower extremities and back [Figure 1a]. There were palpable cervical lymph nodes. Respiratory sounds were decreased on the right lung basal segments. No abnormality was found in the examinations of other systems. In the chest X-ray, diffuse bilateral nodular opacifications and linear densities were observed.

In the computed tomography of the thorax, there were multiple mediastinal lymph nodes, dense consolidation areas, bilateral multiple pulmonary nodules, and ground-glass opacities. Pleural effusion of maximum 42 mm was observed on the right side [Figure 1b].

The patient was hospitalized for the further evaluation. Blood tests were as follows: C-reactive protein: 6.6 mg/L ($n < 5$), white blood cell: 6600, neutrophil: 5000, lymphocyte: 1500, hemoglobin: 12.3 g/dL, platelet: 179 K/μL, alanine aminotransferase: 16 U/L, aspartate aminotransferase: 15 U/L, albumin: 3.7 g/dL, creatinine: 0.83 mg/dL. Sputum sample was taken and for acid-fast bacillus. Quantiferon test was also negative.

Dermatology consultation for skin lesions and chest disease consultation for pulmonary lesions were planned. After dermatologic consultation, a 4-mm punch biopsy was taken from a lesion on the forearm. For pulmonary lesions, bronchoscopy was performed [Figure 1c and d].

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What is Your Diagnosis?

1. Tuberculosis
2. Sarcoidosis
3. Toxoplasmosis
4. Bacillary angiomatosis
5. Kaposi sarcoma (KS)
6. Atypical pneumonia
7. Fungal infection
8. Multicentric Castleman's disease
9. Pulmonary lymphoma.

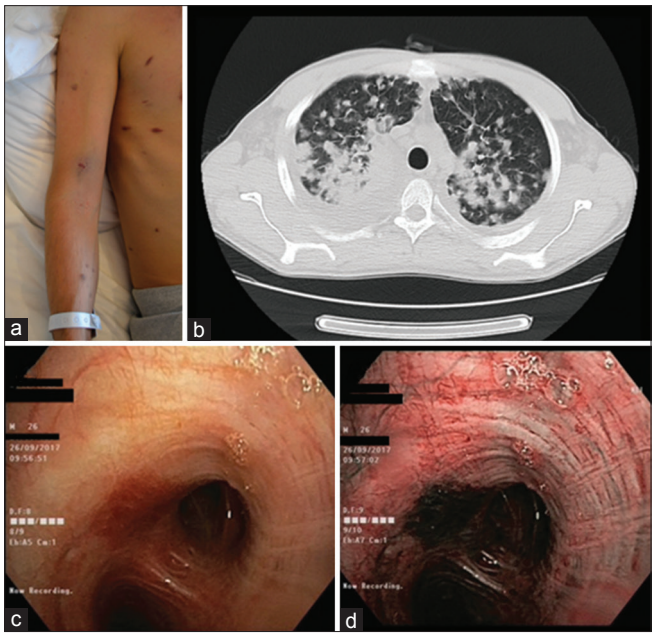

Figure 1: (a) Skin lesions (b) Computed tomography of thorax. Multiple pulmonary nodules and pleural effusion on the right lung were seen in the parenchymal window. (c) Bronchoscopic view of the patient (white-light) (d) Narrow band imaging

Pulmonary Kaposi Sarcoma

The patient was human immunodeficiency virus (HIV)-positive with chronic hepatitis B infection. HIV diagnosis was made in February 2017. At that time, the patient began his treatment with elvitegravir-cobicistat, emtricitabine-tenofovir disoproxil fumarate, but the drug was changed because of the side effects. He has been currently treated with tenofovir, emtricitabine, dolutegravir, and trimethoprim + sulfamethoxazole. CD4 count was 64 cells/μL, and HIV RNA was 47 k/ml. Toxoplasma immunoglobulin G was also checked and was negative. He was started on piperacillin-tazobactam, linezolid, clarithromycin, and metronidazole treatment. Piperacillin-tazobactam was started with preliminary diagnosis of pneumonia and pneumocystis pneumonia (PCP) but stopped due to an allergic reaction to the drug. Patient's complaints of dyspnea continued and saturations lowered to 82%. Prednol 40 mg was administered intravenously, and inhaled beta-2 agonists and inhaled steroid were added to his regimen. The patient had cough, sputum, but no fever.

In bronchoscopic examination, there were multiple purplish-reddish plaques on the bronchial mucosa, which were consistent with pulmonary KS. Punch biopsy of the lesion on the skin supported KS diagnosis by immunohistochemical presence of human herpesvirus-8 (HHV-8) on the endothelial cells. Sputum examination was negative for tuberculosis. In cytopathologic examination of the bronchoalveolar lavage, macrophages, lymphocytes, and epithelial cells were seen and no atypical cell was found.

Total abdominal ultrasonography (USG) was performed to look for any abdominal involvement after the patient developed nausea. USG examination did not give any additional information, except pleural effusion in the right lung.

The patient was discharged on the eight day of his admission on his own wish. His treatment was rearranged. It was stated that patient must continue on antiretroviral treatment and use clarithromycin, trimethoprim + sulfamethoxazole, and fluconazole in addition to antiretroviral drugs. Before discharge, the patient was also consulted with the oncology department. The therapy was planned with six cycle doxorubicin. Five days after the discharge, the patient was given the first cycle of doxorubicin treatment. The control complete blood count of the patient was within normal after 1 week of first cycle doxorubicin and saturation was 95%. The skin lesions have faded on arm, leg, and trunk compared to the first presentation.

KS was first described in 1872 by Moritz Kaposi as angioproliferative lesions caused by HHV-8. It is described as an AIDS-defining condition according to the Centers for Disease Control and Prevention and World Health Organization. The KS seen in the late stages (CD4 <100 cells/μL) of HIV infection is usually extensive and rapidly progressive.[1] Pulmonary KS can present as lung parenchyma, pleura, intrathoracic lymph nodes, and airway involvement.

Patients with pulmonary KS usually present with dyspnea, hypoxemia, and dry cough developing over a few weeks. Hemoptysis and fever may also accompany as in our patient.[2]

PCP is the most common pulmonary disease seen in patients with AIDS. Bacterial pneumonia, tuberculosis, fungal infections, and lymphoma should also be considered in the differential diagnosis of pulmonary disease in AIDS patients.[3] Parasitic infections responsible for pulmonary infection in these patients are mainly *Toxoplasma gondii*, *Strongyloides stercoralis*, *Cryptosporidium*, and *Microsporidia*.[4]

Diagnosis of pulmonary KS is based on clinical data, which are KS cutaneous involvement with distinctive appearance, laboratory findings, radiologic appearance, and endobronchial lesions on bronchoscopy and epidemiology. Nuclear scanning can be used for pulmonary KS when lung tissue sampling is not feasible and there is no cutaneous disease. KS is positive on thallium-201 scans but negative on gallium citrate scans.

Analysis of spontaneously expectorated sputum is the best technique in the differential diagnosis of tuberculosis and other pulmonary infections. Biopsy is not necessary for the diagnosis of KS, and visualization of pathognomonic mucosal infiltrations with flexible bronchoscopy is mostly diagnostic.

Today, the main treatment modality in AIDS-related pulmonary KS is antiretroviral therapy (ART). If the patient has asymptomatic pulmonary KS, ART is usually enough. If the patient is symptomatic like our patient, systemic chemotherapy is applied in addition to ART. This treatment modality increased the survival rates and decreased the incidence of the disease.[5]

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Conflicts of interest
There are no conflicts of interest.

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